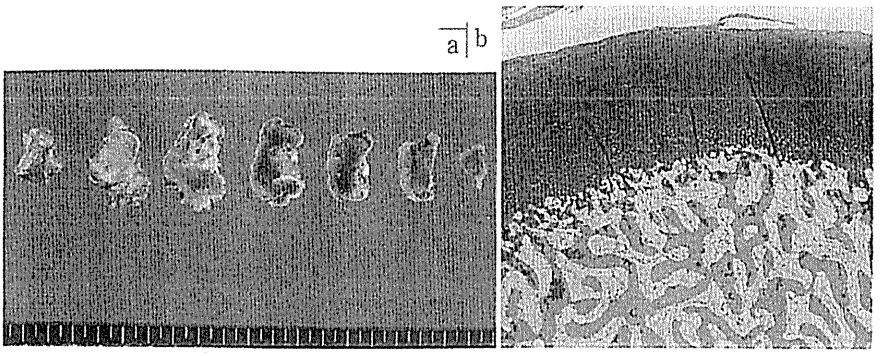

図 5. 病理組織学的所見
a：縦割すると，軟骨帽様組織が連続して観察された．
b：軟骨組織は正常骨髄になだらかに移行している像を示した(×20)．

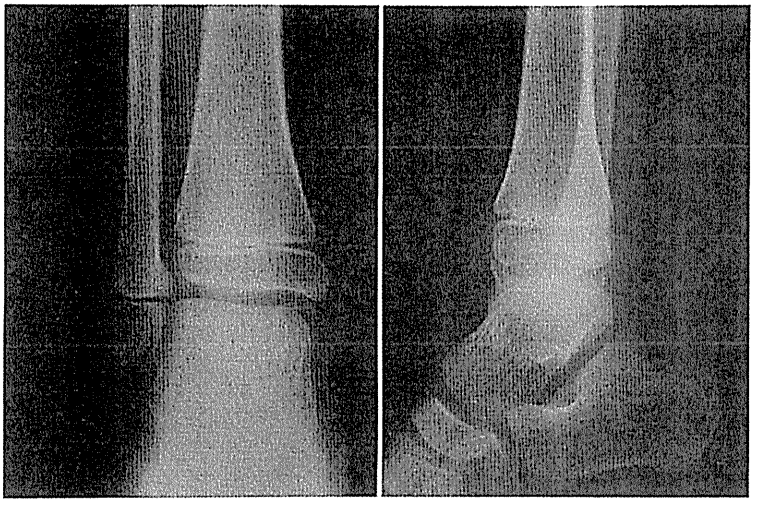

図 6.
術後単純 X 線像
a，b：術後 1 年 7 か月．明らかな再発は認めていない．

足関節発生例であり，年齢，性別，発生部位はいずれも典型的である．

DEH は，肉眼的には骨軟骨種を思わせる軟骨帽を有しており，病理組織学的所見では両者の鑑別は困難である．また軟骨組織を含み関節近傍に発生する滑膜性骨軟骨症との鑑別も必要である．

DEH と骨軟骨腫の違いは，発育方向が DEH では骨端方向に向かうのに対して，骨軟骨腫では骨幹端から骨幹方向に向かう点で両者は異なる[5]．また多発例では遺伝性がある．骨軟骨腫にみられる EXT-1，EXT-2 遺伝子の活性の低下は，DEH においては見られない[3]などの点で両者は鑑別される．

滑膜性骨軟骨症は滑膜組織由来であるといわれており，滑膜内や関節内に局在する．好発年齢は DEH よりも高く，病理組織学的には滑膜との連続性を認める[4]．遊離体の存在なども鑑別に有用である．

自験例は遊離体や腫瘤と滑膜との連続性はなく，滑膜性骨軟骨腫は容易に否定できたが，骨軟骨腫との鑑別は極めて困難であった．しかし，発生部位と腫瘤の伸展方向が DEH とは異なり過去の報告例と臨床像が一致したことから DEH と診断した．

DEH の主な症状は腫瘤，運動時痛，関節可動域制限などであり[6]，これらの改善を目的として切除術を適応されることが多い．手術は，関節機能の温存を目的に，関節変形が発生する前に行うことが一般に推奨されている[1]．切除範囲に関しては，完全切除が困難な関節近傍に病変がある場合は，完全切除でなくても時間の経過とともに腫瘤は消失することもある[7]．自験例は腫瘤の増大に伴い関節可動域制限が進行して転倒しやすくなったため，腫瘤の切除術を施行した．また放置しておくと機能障害は進行し，関節変形をきたす可能性が高いとも考えた．

術後1年の現在，術前と比べて足関節の背屈は10°と正常域に達し，転倒もほとんどしなくなった．画像所見では現在再発は認めていない．

関節内に発生した症例のため，変形性関節症の発症の有無について，術後の長期経過観察が必要と考えている[2]．

謝　辞：御指導，御校閲を頂いた，石井良章名誉教授，里見和彦教授，病理学教室　藤岡保範教授に深謝いたします．

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Abstract

Dysplasia Epiplasia Hemimelica in the Ankle Joint：A Case Report

Takayuki Aoyagi, M. D., et al.
Department of Orthopaedic Surgery, Kyorin University

We report a case of dysplasia epiplasia hemimelica in the ankle joint of a 6-year-old boy. On initial examination, he presented an enlarged bony mass 5 cm in diameter and limited range of motion in the right ankle joint. The mother reported first noticing symptoms at two years earlier, and the boy continued to suffer frequent falls while walking. Radographs and CT showed two bony lesions on the talus and distal portion of the tibia. Resection was performed to obtain a histopathological diagnosis and to improve the range of motion. The histopathological findings suggested an osteochondral lesion similar to osteochondroma, but dysplasia epiplasia hemimelica was the final diagnosis since the lesion stemmed from the talus and distal epiphysis of the tibia. After resection, the range of motion was improved with improved gait and relief from pain.

Capillary Hemangioma in a Rib Presenting as Large Pleural Effusion

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Hisashi Tanaka, MD, PhD, Eiichi Morii, MD, PhD,
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Departments of Orthopedic Surgery, Radiology, Pathology, and General Thoracic Surgery, Osaka University Graduate School of Medicine, Osaka, Japan

Intraosseous hemangioma in a rib is extremely rare, and most of the few reported cases are of the cavernous subtype. First we describe a capillary hemangioma arising from a rib in a 64-year-old woman that developed into a large, one-sided pleural effusion during the course of a 3-year follow-up. In addition to the life-threatening condition, the tumor demonstrated malignant imaging features such as a sunburst-like appearance or cortical disruption on plain roentgenogram and computed tomography. This case report adds to the literature on a serious complication and also discusses the diagnosis and management of this rare disease.

(Ann Thorac Surg 2011;91:e59–61)

Primary tumors in the ribs are rare, accounting for 6% to 10% of primary bone tumors, and approximately half of these are malignant [1, 2]. Among benign tumors, cartilagenous tumor and fibrous dysplasia are common, whereas hemangioma is exceedingly rare with only a few cases reported in the literature. Some of the reports have described these hemangiomas as the cavernous subtype on histopathologic examination. Herein, we believe that our report is the first case of a patient with capillary hemangioma of the rib and complications of rapidly accumulating pleural effusion after 3-year follow-up. No reports have described costal hemangioma presenting with these life-threatening conditions.

In November 2006, a 64-year-old woman was referred to our hospital with a bone tumor in the left fifth rib. She had no major past history before the mass was accidentally identified during a medical check-up.

The patient was asymptomatic on presentation with no pain or evident mass. Neither tenderness nor swelling was observed. A roentgenogram of the rib demonstrated increased radio-density of the affected bone compared to adjacent ribs, and the rib was flat and widened on palpation. The mass was expansile with irregular cortex and coarse trabeculae with a "honeycomb" appearance (Fig 1). This mass was accompanied by an extraosseous mass showing a sunburst-like appearance. These findings strongly suggested hemangioma of the bone, and fibrous dysplasia or low-grade chondrosarcoma, which was also considered as a differential diagnosis.

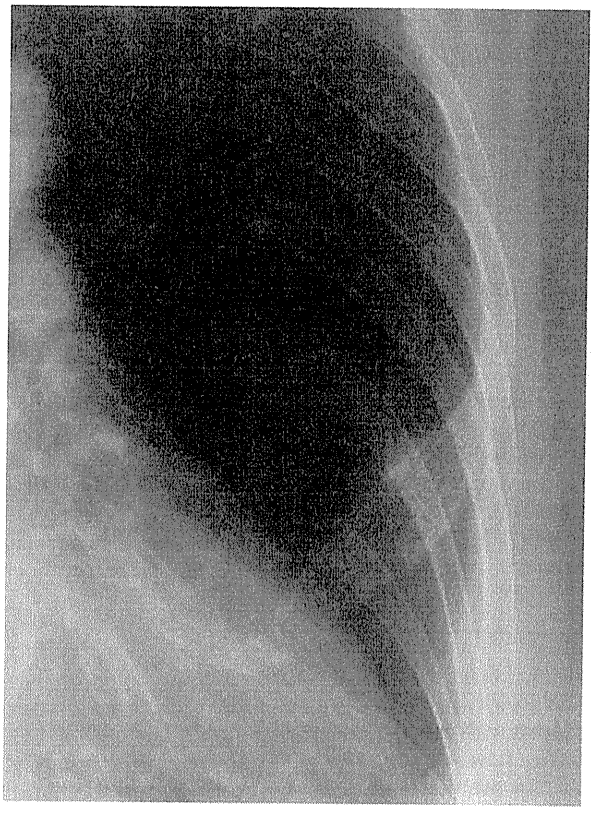

Fig 1. A chest roentgenogram showing an expansile lesion of the left fifth rib with sunburst-like appearance of extraosseous mass.

Due to the absence of symptoms or clinical findings suggesting rapid progression, the patient was followed-up carefully. Increases in tumor size remained minimal until January 2009. A roentgenogram, in January 2010, showed slight enlargement of the tumor along with evidence of a small amount of pleural effusion, although this had not been noticed until the patient began to feel progressive, exertional dyspnea without chest pain, and she was urgently hospitalized in March. At the time of admission, the roentgenogram showed a large, left-sided, pleural effusion.

No abnormality was found in the laboratory studies including tumor markers, except for a subtle increase in C-reactive protein level to 0.39 mg/dL. A computed tomographic scan demonstrated a costal mass with partial cortical destruction and extraosseous extension along with massive pleural effusion (Fig 2). However, neither pleural nor pulmonary tumors were found. A thoracentesis yielded clear, yellowish transudate without significant changes in levels of total protein (4.5 g/dL), glucose (125 mg/dL), amylase (42 U/L), lactate dehydrogenase (114 U/L), adenosine deaminase (5.40 IU/L), or hyaluronic acid (938 ng/mL). Both cytologic and culture examinations showed no abnormalities.

Fluid accumulation continued despite repeated thoracentesis, administration of diuretics, and fluid restriction, suggesting the possibility of malignant pleurisy. Resection of the rib along with the parietal pleura was therefore performed.

Accepted for publication Nov 23, 2010.

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Published by Elsevier Inc

0003-4975/$36.00
doi:10.1016/j.athoracsur.2010.11.074

Macroscopically, a tumor measuring 8 × 4 × 2.5 cm was identified bulging from the inner aspect of the fifth rib toward the thoracic cavity. The surface of the tumor was covered with a smooth pleura and no tumor extension beyond the membrane was observed. Microscopically, the tumor comprised a conglomerate of capillary vessels with few dilated channels. The lining endothelial cells showed no significant atypia or mitosis (Fig 3). Based on these findings, capillary hemangioma of the bone was diagnosed. Pleural tissues adjacent to the tumor showed markedly increased thickness and contained scattered blood cells, suggesting invasion of capillary vessels from the tumor, but no accumulation of inflammatory cells was evident. The patient was discharged after an uneventful postoperative course. No further accumulation of pleural effusion has been observed.

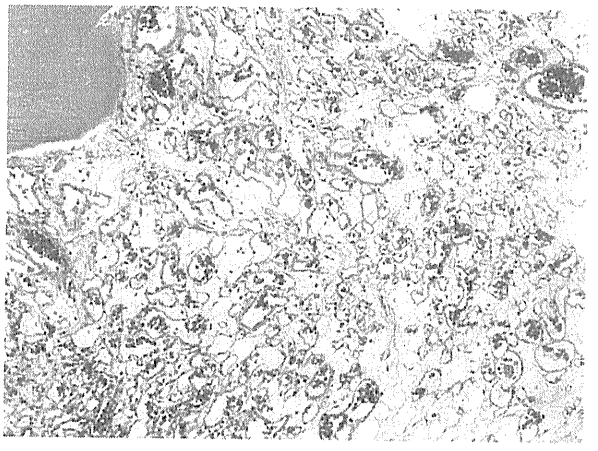

Fig 3. Microscopically, the tumor was composed of capillary-sized vessels. The lining endothelial cells showed no significant atypia. (Hematoxylin-eosin staining; ×100.)

Comment

Hemangioma is a common benign vascular tumor that arises in a variety of organs and sites. The bone is also involved with low incidence, accounting for 1% to 6% of all bone tumors [3, 4]. These lesions are most commonly found in the vertebral column and skull, and the hemangioma involving a rib is exceedingly rare. Some of the reports have described these hemangiomas as cavernous subtype on histologic examination. This is the first report of a capillary hemangioma occurring in a rib.

In addition, this case presented with rapid accumulation of one-sided pleural effusion mimicking malignant pleurisy. Most previous reports describe costal hemangiomas as being asymptomatic, and the diagnosis is often made incidentally on routine chest roentgenograms. There are a few reports of the lesion being symptomatic, producing localized pain; however, accumulation of pleural effusion has not been reported. Although the precise underlying mechanisms and relationships with capillary subtype remain unclear, hypertrophic pleural tissues and capillary tumor vessels facing the thoracic cavity might play a causative role through pleural inflammatory processes or increased microvascular permeability.

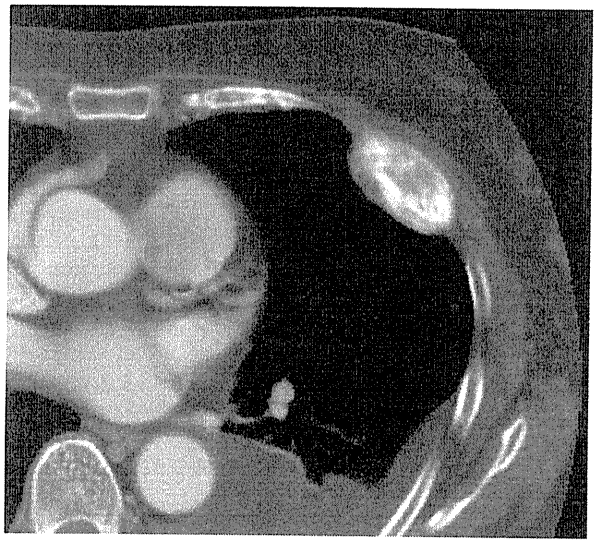

Fig 2. Computed tomographic image showing cortical disruption and "honeycomb" appearance of trabeculae, along with a left-sided, large pleural effusion.

The radiographic appearances have been described in several literatures. The authors have reported that costal hemangiomas showed a sunburst appearance or disruption of the bony cortex with extraosseous extension, or both, although these are generally malignant findings [5, 6]. Honeycomb appearance of the medulla is also common to intraosseous hemangiomas [6]. These findings are helpful for making an imaging diagnosis. For the actual diagnosis, however, histologic examination is essential.

The necessity of preoperative biopsy is controversial. Some authors have recommended one-step resection surgery because biopsy of a hemangioma can result in significant bleeding [7]. This may also cause pathologic misleading due to insufficient sampling. On the other hand, if a tumor is responsive to chemotherapy, such as osteosarcoma, or if Ewing sarcoma is suspected, total resection will make the effectiveness of subsequent chemotherapy impossible to evaluate. Complete resection of the rib lesion should be indicated when hemangioma is strongly suspected.

Taken together, the tumor in the present case showed several malignant features in terms of both symptomatology and diagnostics, despite its benign nature. This case serves to remind clinicians that costal hemangioma can take on this unique, clinical behavior. In cases showing development of expansile costal tumor, even if the growth is gradual, and especially when the tumor is accompanied by malignant findings, such as a sunburst-like appearance or cortical disruption, a surgical resection should be considered.

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Japanese Journal of Clinical Oncology Advance Access published January 19, 2011

JJCO Japanese Journal of Clinical Oncology

Jpn J Clin Oncol 2011
doi:10.1093/jjco/hyq246

Original Article

Eleven Cases of Cardiac Metastases from Soft-tissue Sarcomas

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Received November 17, 2010; accepted December 23, 2010

Objective: Cardiac metastasis is a highly life-threatening condition because it leads to cardiac failure. However, it is difficult to diagnose because its precise clinical features are unknown. Here, we report 11 cases of cardiac metastasis from soft-tissue sarcoma, and discuss its diagnosis and treatment.

Methods: Of 641 patients with soft-tissue sarcoma treated in our institute between 1996 and 2009, we retrospectively reviewed the medical records of 11 patients whose cardiac metastases were diagnosed while they were alive.

Results: The most common primary tumor was leiomyosarcoma ($n = 5$), followed by clear cell sarcoma ($n = 2$). In all cases, metastases to other organs, including lungs ($n = 10$), soft tissues ($n = 5$) and bones ($n = 4$) were found along with cardiac metastases. Cardiac metastasis was diagnosed by echocardiography in six cases and by computed tomography in four cases. In four patients, cardiac metastasis was not detected by chest computed tomography as follow-up to lung metastases and echocardiography was required to make the diagnosis. Although five patients complained of exertional dyspnea, four were asymptomatic. Seven cases were treated with radiotherapy. No patient had surgery for their cardiac metastasis. The median survival of patients who received radiation therapy was 10.5 months; that of those who did not was 3.5 months.

Conclusions: Cardiac metastasis is often asymptomatic. Echocardiography is better than computed tomography for diagnosing cardiac metastasis, and should be considered in all patients presenting with soft-tissue metastases. Owing to the highly life-threatening nature of cardiac metastases and the possibility of soft-tissue dissemination, treatment with radiation therapy is recommended immediately on diagnosis.

Key words: cardiac metastasis – sarcoma – radiation

INTRODUCTION

Cardiac metastases are the immediate cause of death in as many as one-third of cases in which they occur (1). They may cause cardiac failure, arrhythmia and pericardial effusion, with cardiac failure being the main cause of death as a result of restrictive myocardial disease, ventricular inflow and outflow tract obstruction and pericardial constrictive disease. According to autopsy reports, cardiac metastases have been found in 25% of patients with soft-tissue sarcoma (1). The incidence of cardiac metastasis has increased during recent decades because better diagnostic tools and aggressive treatment of localized malignant tumors have led to longer

survival of patients (2). Despite their frequency, cardiac metastases are seldom diagnosed in living patients because other metastatic symptoms prevail. We reviewed 11 cases of cardiac metastasis from soft-tissue sarcoma, which had been diagnosed while the patients were alive, to discuss how to detect and treat them.

PATIENTS AND METHODS

We retrospectively reviewed the medical records of 641 patients with soft-tissue sarcoma treated at Osaka Medical Center for Cancer and Cardiovascular Diseases between 1996 and 2009. The 11 cases that had antemortem diagnoses of myocardial or pericardial metastases were further evaluated.

ILLUSTRATIVE CASES

Patient No. 1

A 40-year-old woman with a history of mesenterium leiomyosarcoma treated with surgery three times, for the primary lesion and two local recurrences, showed lung metastases (treated with radiotherapy and chemotherapy), a right-buttock metastasis (treated with surgery), a left chest wall metastasis (treated with radiotherapy) and a left pelvic metastasis (treated with radiotherapy). Six years after the initial operation, she began to experience dyspnea and general fatigue. An echocardiogram revealed a 53 mm left atrial mass that blocked the left ventricular inflow tract (Fig. 1). Radiation therapy was impossible due to her inability to stay in position. She died on the day the cardiac metastasis was detected.

Patient No. 2

A 55-year-old woman with a history of a right-buttock leiomyosarcoma treated with surgery and chemotherapy showed several metastases to her lung, left back, right forearm, left thigh and right buttock, all of which were surgically resected. Three years after the initial surgery, she presented with general fatigue. An echocardiogram revealed a 30 mm right atrial mass and cardiac tamponade resulting from a large pericardial effusion (Fig. 2). She received continuous drainage of pericardial effusion and radiation therapy for cardiac metastasis, receiving a total dose of 25 Gy in five fractions of 5 Gy. Twenty months after the radiation for cardiac metastasis, re-growth was observed, which was refractory to 25 Gy re-radiation. She died 3 months after the second radiation.

RESULTS

Metastatic disease to the heart was found in 11 cases of soft-tissue sarcoma, including the myocardium in 8 cases and the pericardium in 3 cases. The clinical features of these patients are shown in Table 1. The median age at initial diagnosis was 33 years. The median interval from initial diagnosis to cardiac metastasis was 36 months (range 8–108 months). The most common primary tumor was leiomyosarcoma ($n = 5$), followed by clear cell sarcoma, rhabdomyosarcoma, alveolar soft-part sarcoma, synovial sarcoma and malignant fibrous histiocytoma. Myocardial metastases favored the left over the right side of the heart, with the left ventricle predominating. In all cases, metastatic disease was found in other organs before or concurrently with the cardiac metastasis. The lungs were involved in 10 cases (91%), soft tissues in 5 cases (45%), bone in 4 cases (36%), pancreas in 2 cases (18%), kidneys in 2 cases (18%) and liver in 2 cases (18%).

Echocardiography provided the definitive diagnosis in six cases of this series, with four cases being diagnosed using computed tomography (CT). In one case, pericardium metastasis was found accidentally during surgery of lung metastasis. Clinical symptoms at diagnosis of cardiac metastasis included dyspnea in five cases, and general fatigue in two cases. Four patients were asymptomatic, which is more than might be expected.

Three patients required continuous drainage of pericardial fluid. In two of these three patients, pericardial effusion increased during chemotherapy. Seven cases of cardiac metastasis were treated with radiotherapy. No patient in this series had surgery for their cardiac metastasis. Survival from diagnosis of cardiac metastasis, excluding the one surviving case, was 8 months on average. Median survival of patients who received radiation therapy was 10.5 months, compared with 3.5 months for patients who did not receive radiation.

DISCUSSION

Hallahan et al. (1) reported that cardiac metastases were present in 25% of consecutive autopsies of patients with soft-tissue sarcoma, which is higher than recognized clinically, suggesting that most cases are probably missed. Cardiac metastasis often causes immediate death of a patient, so establishment of appropriate management is very important. However, little is known in terms of its precise clinical features.

The most common symptom of cardiac metastasis in this series was dyspnea, but, as many of the patients also had lung metastases, this is not clearly indicative of cardiac metastasis. It was not unusual for patients with cardiac metastasis to be asymptomatic. Furthermore, there were four cases whose cardiac metastases were not detected on chest CT as follow-up for lung metastases and were later found by echocardiography. It is supposed that chest CT failed to detect cardiac metastases even though tumors existed in hearts because chest CT showed mitral valve calcification and heart enlargement in case 1 and a small amount of pericardial effusion in case 2 but not cardiac metastases. In two cases (cases 6 and 8), patients received echocardiography despite being without symptom in order to investigate

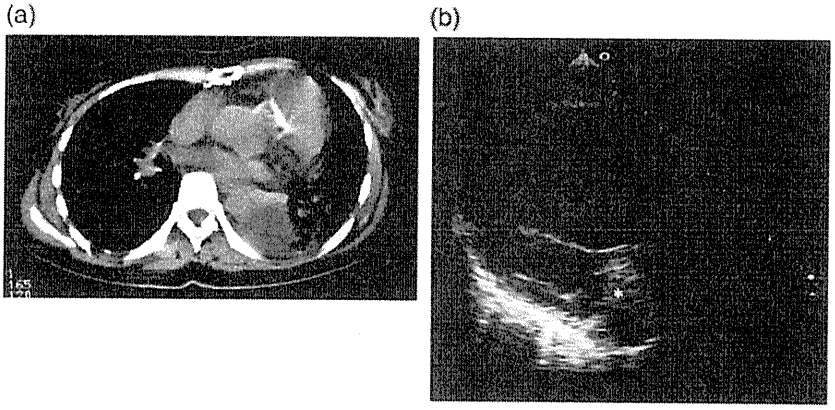

Figure 1. Chest computed tomography (CT) and echocardiogram of Patient 1. (a) and (b) Chest CT showed mitral valve calcification, heart enlargement and multiple lung metastases, but not cardiac metastasis. Two months later, her dyspnea and general fatigue had worsened. An echocardiogram revealed a 53 mm left atrial mass (*) that blocked the left ventricular inflow tract.

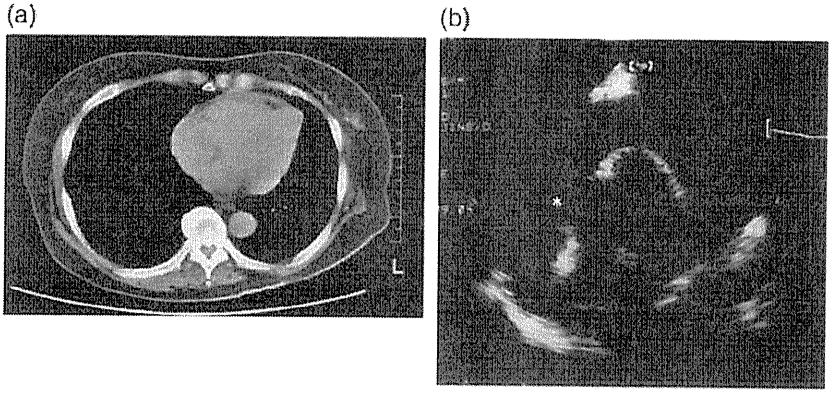

Figure 2. Chest CT and echocardiogram of Patient 2. (a) and (b) Chest CT showed a small amount of pericardial effusion and lung metastases, but not cardiac metastasis. Three months later, she presented with general fatigue. An echocardiogram revealed a 30 mm right atrial mass (*) and cardiac tamponade resulting from a large pericardial effusion.

whether she had myocardial damage by doxorubicin (case 6) and whether he had cardiac function to tolerate surgery for bone metastasis (case 8). Their cardiac metastases were found accidentally by those echocardiograms. In this case series, myocardium metastasis was often accompanied by soft-tissue metastases, possibly as a result of the intracardial metastasis sending malignant tumor cells into the circulation. We recommend that echocardiography should be performed on all patients with soft-tissue metastases.

While theory suggests that the filtration of systemic venous return by the pulmonary circulation would effectively shield the left ventricle compared with the right ventricle, previous autopsy studies have concluded that the left ventricular myocardium is most often the focus of metastatic disease, purportedly due to its level of vascularity (3). In this study, metastases of the heart were observed more often on the left than the right side. Left-side metastasis of the heart is considered to arise from coexistant lung metastases.

In this study, the most common histological type of cardiac metastasis was leiomyosarcoma. The histological types of cardiac metastases have not been included in autopsy reports (1). Leiomyosarcoma is the predominant sarcoma arising from large blood vessels, and it has been reported to be as common as angiosarcoma in primary cardiac sarcoma (4). These facts suggest that leiomyosarcoma can grow easily in the heart and vessels.

Hughes et al. (5) reported that cardiac metastasis correlated positively with pericardial effusion in pediatric sarcoma patients. Instillation of tetracyclines (6), radioactive phosphorus (7) or cisplatin (8) was reported to be a useful palliative procedure for malignant pericardial effusion. Radiation therapy prolongs life in patients with pericardial effusion, compared with repeated pericardiocentesis alone (9). In this series, continuous drainage of pericardial effusion in three cases and emergency pericardiotomy in one case were required for cardiac tamponade. Pericardial effusion increased during chemotherapy in two cases due to hydration of chemotherapy. After radiation therapy for their cardiac metastasis, the need for continuous drainage of cardiac effusion ceased. Therefore, radiation therapy for cardiac metastasis should be performed prior to chemotherapy.

Table 1. The clinical features of 11 cases of cardiac metastases from soft-tissue sarcomas

Pt. no.	Age	Sex	Diagnosis	Primary site	Primary size (cm)	Depth	Histological grade	AJCC staging	Time to initial met (month)	Time to heart met (month)	Heart met site	Other metastases	Chief symptom	Diag. modality	Therapy	Survival from heart met (month)
1	40	F	Leiomyosarcoma	Mesenterium	N/A	Deep	N/A	N/A	32	65	LA	Lung, soft tissue, bone	Dyspnea	Echo	–	0
2	55	F	Leiomyosarcoma	Rt buttock	8	Deep	2	II B	8	36	RV	Lung, soft tissue	Fitigue	Echo	RT (25 Gy/5Fr)/DCE	24
3	63	F	Leiomyosarcoma	Uterus	N/A	Deep	N/A	N/A	24	108	LV	Lung, soft tissue, liver, stomach, bladder, renal	Asymptom	CT	RT (45 Gy/15Fr)	13
4	47	M	Leiomyosarcoma	Chest wall	12	Deep	2	IV	0	30	LV	Lung, soft tissue, bone	Dyspnea	Echo	RT (50 Gy/25Fr)	7
5	26	F	Leiomyosarcoma	Rt groin	7	Deep	2	II B	33	51	LV	Lung	Asymptom	CT	RT (60 Gy/30Fr)	18 (AWD)
6	30	F	Clear cell sarcoma	Rt knee	7	Deep	3	IV	0	31	LV	Lung, soft tissue, bone, lymph	Asymptom	Echo	Immunotherapy	4
7	33	F	Clear cell sarcoma	Rt foot	3	Deep	3	IV	0	64	Pericardium	Lung	Asymptom	OP	–	5
8	23	M	ASPS	Lt shoulder	8	Deep	2	IV	0	27	LV	Lung, bone, pancreas	Asymptom	Echo	RT (60 Gy/30Fr)/Chemotherapy	8
9	69	F	Undifferentiated pleomorphic sarcoma	Rt thigh	10	Deep	3	III	11	13	RA	Pancreas	Dyspnea	CT	–	5
10	16	M	Rhabdomyo sarcoma	Lt buttock	8	Deep	3	III	8	8	Pericardium	Lung	Fatigue	Echo	RT (40 Gy/20Fr)/ Chemotherapy	6
11	13	F	Synovial sarcoma	Lt buttock	12	Deep	3	IV	0	57	Pericardium	Lung, liver, renal	Dyspnea	CT	RT (32 Gy/16Fr)/ DCE/Pericardiotomy/ Chemotherapy	5

Histological grade: according to Fédération Nationale des Centres de Lutte Contre le Cancer grading system for leiomyosarcoma, undifferentiated pleomorphic sarcoma and synovial sarcoma, but National Cancer Institute grading system for others.
AJCC staging: American Joint Committee on Cancer (AJCC) staging at the first presentation according to AJCC staging manual seventh edition.
OP: pericardium metastasis was found accidentally during operation of lung metastasis.
CT, computed tomography; RT, radiation therapy; DCE, drainage of cardiac effusion; N/A, not available; ASPS, alveolar soft part sarcoma; LA, left atrium; RV, right ventricle; RA, right atrium.

The seven patients who received radiation therapy tended to survive longer than those who did not. The tolerance of the heart to radiation therapy has been reported previously (10). Al-mamgani et al. (11) suggested that a total dose of 45 Gy, in 25 fractions of 1.8 Gy, would be appropriate for cardiac metastasis in a curative setting; an additional dose of 10–15 Gy may be delivered through small portals when indicated. In this study, cardiac metastasis treated with 25 Gy of radiation therapy showed re-growth only after 20 months, when re-irradiation was required.

There was no patient who underwent biopsy or surgery for tumor in the heart in this series. Although the possibility remains that tumors in the heart are primary tumors, we decide they are metastases from soft-tissue sarcoma from clinical course. In some cases, we performed resection of the tumors in the lung and/or soft tissue and confirmed they were metastases from soft-tissue sarcoma. While there have been some reports of long-term survival after resection of cardiac metastasis, the perioperative mortality rate remains high (40%) (12). Patients with diffuse cardiac metastases, massive intracavitary disease or uncontrolled metastatic disease have been reported to be poor-risk patients for surgical resection (1).

In conclusion, we suggest that echocardiography should be considered in cases of sarcoma presenting with soft-tissue metastases, and, because of the highly life-threatening nature of the condition and the possibility of soft-tissue dissemination, cardiac metastases should be treated by radiation therapy, a total dose of more than 45 Gy, immediately and prior to chemotherapy.

Funding

This study was supported by a grant from the National Institute of Biomedical Innovation of Japan (07-13).

Conflict of interest statement

None declared.

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The Foot 21 (2011) 157–161

Contents lists available at ScienceDirect

The Foot

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Case report

Prosthetic reconstruction for tumors of the distal tibia. Report of two cases

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ARTICLE INFO

Article history:
Received 16 September 2010
Received in revised form 20 January 2011
Accepted 20 January 2011

Keywords:
Malignant bone tumor
Distal tibia
Custom made prosthesis

ABSTRACT

Prosthetic reconstruction in two patients with malignant bone tumors of the distal tibia was conducted. The diagnoses were metastatic bone tumor in one patient and low grade central osteosarcoma in another. The mean duration of follow-up was 5.5 years (3 and 8 years). Reconstruction was achieved using custom-made prosthesis (JMM, Japan Medical Materials), which replaced the distal tibia. In the patient with metastasis, local recurrence occurred 8 months after the primary surgery and the recurrent tumor was resected. Both patients were free from neoplastic disease at the latest follow-up. The average functional scores according to the system of the Musculoskeletal Tumor Society were 25 and 23. Custom-made prostheses allow an early return to functional weight-bearing without major complications. This technique provides a safe and effective method of stabilization for properly selected malignant tumors of the distal tibia.

1. Introduction

Malignant bone tumors of the distal tibia are very rare [1]. Below knee amputation has been a surgical treatment of choice for the local tumor with satisfactory functional results [1,2]. The subcutaneous location and proximity of the distal tibia to the neurovascular bundle and tendons make adequate excision with wide margins of malignant tumors of the distal tibia difficult to achieve [2]. Therefore, there have been only few publications on prosthetic replacement of the distal tibia [1,2].

In this study, custom-made prosthetic reconstruction without talar surface replacement in two patients with malignant tumors of the distal tibia was performed. Hence, the clinical and functional results of this method are being presented.

2. Preoperative planning and custom-made prosthesis

Before operation, the patients were assessed to determine the extent of the local disease and presence of metastases by clinical assessment, plain radiography, and chest tomography (CT). Magnetic resonance imaging (MRI) was also performed to define the extent of the tumor, involvement of the soft tissues, particularly the neurovascular bundle, and level of bone resection.

The prosthetic system (Japan Medical Materials Ltd., Kyoto, Japan) is custom-made based on the anticipated level of resection of tibia for distal tibial bone tumors (Fig. 1). The prosthesis takes approximately 6 weeks to prepare.

3. Operative technique

Surgical approach was determined by the position of the tumor. Meticulous resection was carried out to possibly preserve a wide margin of tissue. The tumor was excised en-bloc (Fig. 2A and B). The proximal intramedullary canal was reamed and the stem was secured with bone cement, with a clinical outcome of appropriate rotational alignment in mind (Fig. 2C). The joint elements made with ultrahigh molecular weight polyethylene (UMWP) were manufactured to fit the talar joint surface. The range of motion and stability of the ankle were evaluated before the skin was closed.

4. Post operative rehabilitation

The patient was mobilized from bed to chair within the first 48 h postoperatively. A short-leg splint was applied after four weeks and then passive and active movements were commenced (Fig. 2D). Partial weight-bearing was allowed at 4 weeks, which progressed to full weight-bearing at about 6 weeks.

5. Functional assessment

Functional outcome was assessed using the Musculoskeletal Tumor Society (MSTS) functional evaluation system [3]. The MSTS score is composed of pain, function, emotional acceptance, walking ability, gait, and use of walking aids, with a higher score indicating better functional outcome.

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doi:10.1016/j.foot.2011.01.014

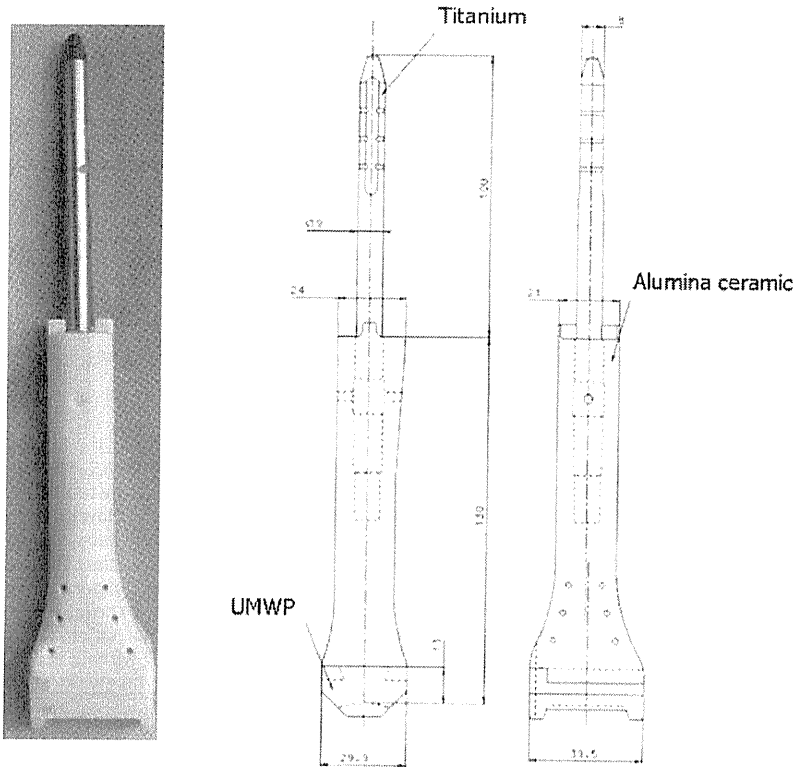

Fig. 1. Custom-made distal tibial prosthesis (Japan Medical Materials). The component is made from titanium (Ti–6Al–4V) intramedullary stem and alumina ceramic body. The articular surface of the joint element is covered with ultrahigh molecular weight polyethylene (UMWP) in order to achieve contact smoothly with the talar surface.

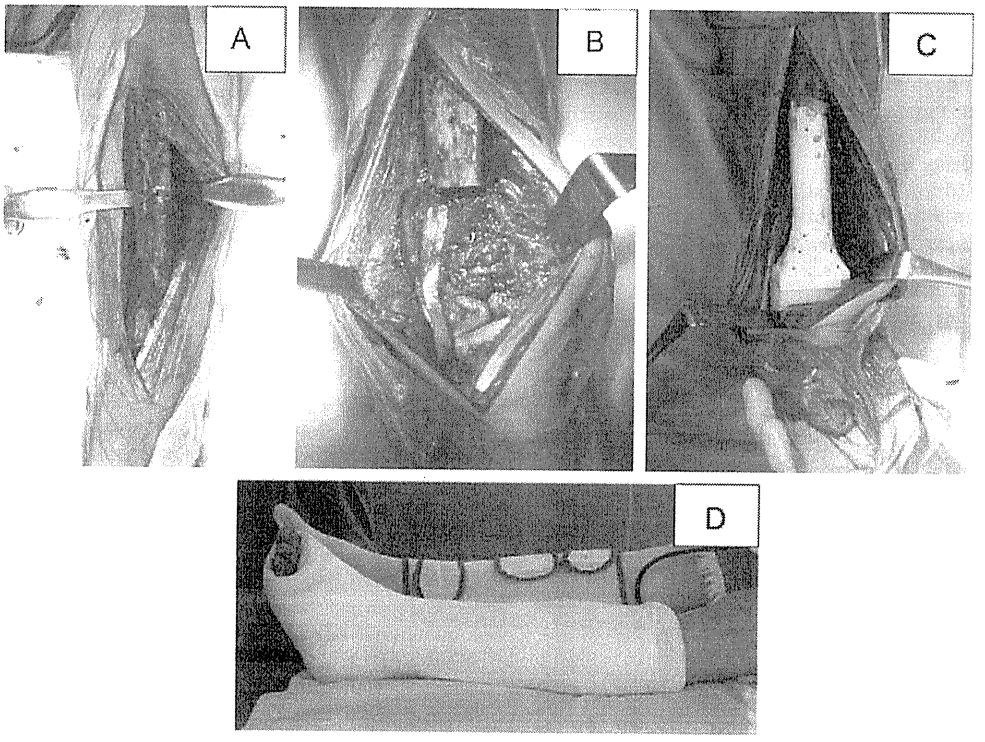

Fig. 2. (A, B) The bone was divided 5 cm above the upper margin of the tumor and resected en bloc. (C) The proximal intramedullary canal was reamed to the appropriate size. The stem of the tibial component was inserted into the medulla of the proximal portion of the tibia and fixed with bone cement, with the clinical outcome of appropriate rotational alignment in mind. (D) A short leg splint was applied for four weeks.

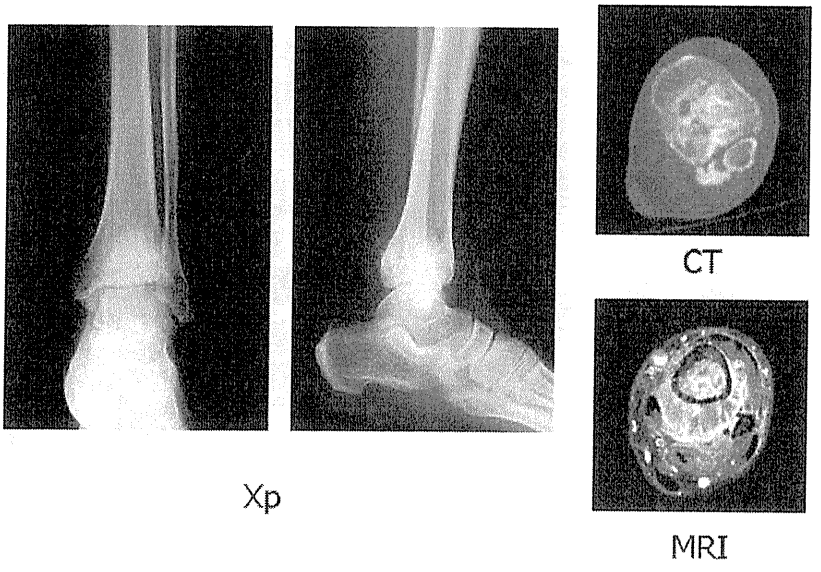

Fig. 3. A 52-year-old man with metastatic bone tumor from colon cancer. Anteroposterior, lateral radiograph, and CT showed mixed lytic and sclerotic lesions involving the distal tibia. Axial T-1 weighted image (Gd-enhanced) MRI showed extensive destruction of the distal tibia and extra-osseous soft tissue extension.

6. Case reports

6.1. Case 1

A 52-year-old male was treated at age 46 for adenocarcinoma of the colon with low anterior resection and chemotherapy. Four years after operation, the patient developed left-sided ankle pain. A needle biopsy led to the diagnosis of bone metastasis from colon cancer. Radiofrequency ablation (RFA) to this lesion was performed in a previous hospital; however, the treatment was not effective. Radiation therapy was additionally performed; however, after eight months, local recurrence occurred. Radiograph of the lower extremity showed mixed lytic and sclerotic lesions in the distal tibia (Fig. 3). The patient refused amputation for psychological and social reasons when informed of the recommended treatment. Instead, tumor resection with marginal resection and prosthetic reconstruction were carried out (Fig. 4). Although the fibular artery, flexor digitorum longus, and fibularis longus muscle were resected, the tibial nerve and posterior tibial artery were preserved. Intraoperative complication did not occur. Subsequently, the patient had local

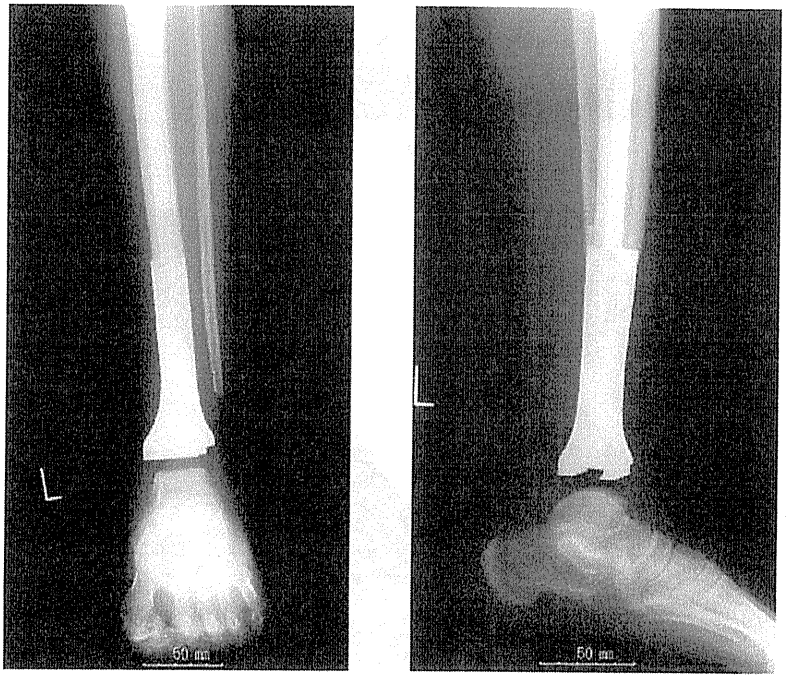

Fig. 4. Postoperative radiographs.

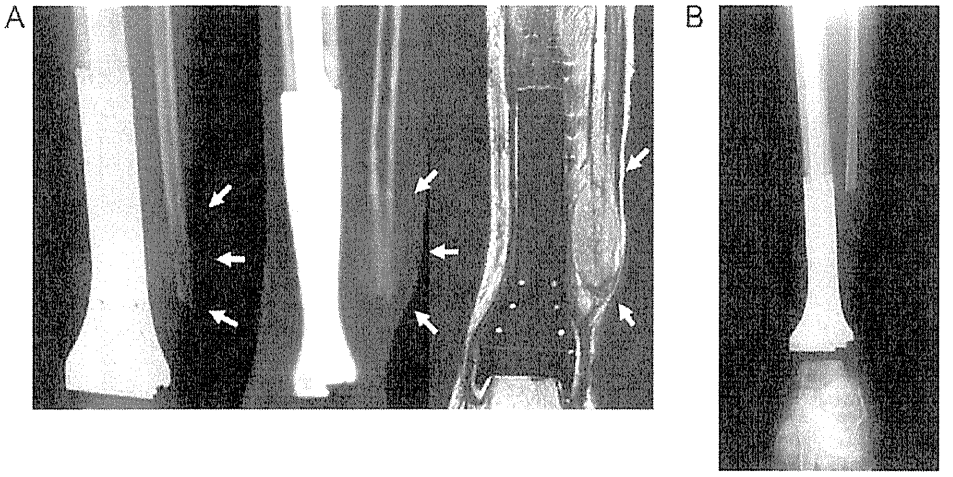

Fig. 5. (A) Local recurrence occurred in the distal end of the fibula eight months after primary surgery (arrow). (B) Second operation was performed and the recurrent lesion was resected.

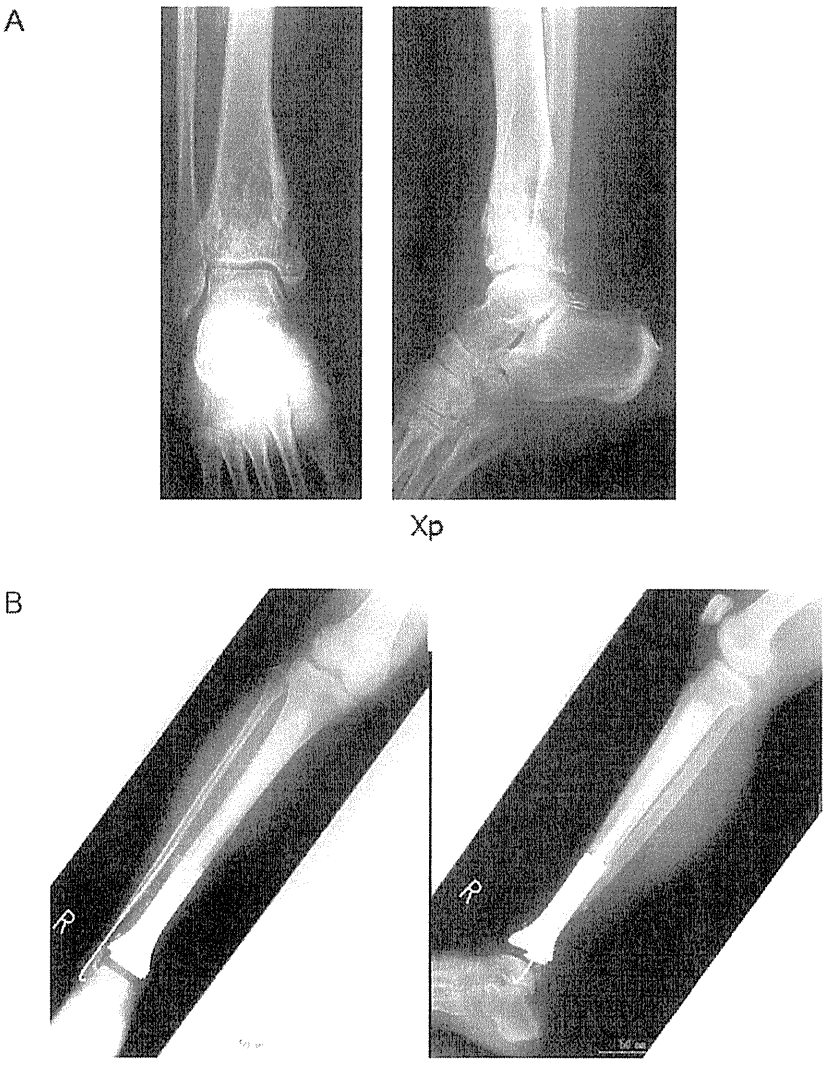

Fig. 6. (A) 52-Year-old woman with low grade central osteosarcoma. Anteroposterior and lateral radiographs showed destructive and osteolytic changes in the right distal tibia (A). Postoperative radiograph (B). Intraoperative fibular fracture occurred.

recurrence in the distal fibula (Fig. 5A). Resection was performed 8 months after the primary surgery (Fig. 5B). The patient remained free from disease at the latest follow up and showed neither radiological loosening nor subsidence of the talar component for three years. The range of ankle doriflexion was 10° and plantar flexion was 30°. The mean MSTS functional score was 25 (pain 5, function 3, emotional acceptance 5, walking ability 4, gait 4, and use of walking aids 4), which indicated 83.3% normal function.

6.2. Case 2

A 51-year-old female presented with a three-year history of swelling and slight tenderness on the right ankle. Pain slightly increased one month ago. A tumor in the distal tibia was pointed out in the previous hospital and the patient was referred to our hospital. Radiograph showed moth-eaten pattern of bone destruction in the right distal tibia (Fig. 6A).

Open biopsy was performed and histopathological diagnosis was low grade central osteosarcoma. The tumor with part of the tibial posterior muscle and flexor hallucis longus muscle were resected and custom-made prosthesis replaced the defect. The posterior tibial artery and nerve were not resected.

Fibular fracture occurred intraoperatively and the fracture was fixed intramedullary with *k*-wire (Fig. 6B). The range of ankle doriflexion after operation was −5° and plantar flexion was 30°. The patient remained free from disease at the latest follow-up and showed neither radiological loosening nor subsidence of the talar component for eight years. The mean MSTS functional score was 23 (pain 5, function 3, emotional acceptance 5, walking ability 3, gait 3, and use of walking aids 4).

7. Discussion

The distal tibia is an uncommon site for primary malignant bone tumors [1,2]. Abudu et al. [2] reported that of 190 patients with primary bone sarcoma of the tibia treated at one center for over 25 years, only 21 (11%) had the distal tibia as the location of the tumor. The most common of the primary bone tumors are the osteosarcomas and Ewing sarcoma [4].

Metastasis below the knee is also uncommon [5]. Lesson et al. [6] reported an approximately only 4% incidence below the knee.

There are limited indications for limb-preserving surgery in patients with malignant tumors of the distal tibia [2]. Most of the patients with malignant tumors of the distal tibia were treated by below knee amputation [1,2,7]. The principal concerns in the choice of limb-salvage surgery are the extent of soft-tissue invasion and the involvement of the distal blood vessels [1]. If neither the posterior tibial artery nor the dorsalis pedis artery proved to be salvageable, amputation should be carried out [1].

The many options of reconstruction, such as autograft, allograft, arthrodesis, implanted prostheses, and composites had been reported [1–3,8–11]. Casadei et al. [8] reported good functional and oncological results in patients with malignant bone tumors of the distal tibia, which were treated by resection and arthrodesis with autogenous bone graft. However, arthrodesis of the ankle has several limitations, including the loss of joint movement, long period of recovery, unpredictable outcome, possibility of non-union, need for multiple operations to achieve arthrodesis, and loss of ankle motion [1,2,12]. Although Bishop et al. [13] reported good results in a reconstruction using a vascular-free fibular graft in patients with malignant bone tumors of the distal tibia, allograft reconstruction is time-consuming, associated with significant risk of graft infection, prone to fractures, related to prolonged immobilization, and the outcome is unpredictable [2].

Ankle replacement arthroplasty has been occasionally carried out in patients with advanced arthritis [9–11]. Kofoed and Sørensen [10] observed a 73% rate of survival for their prosthesis at 14 years in arthritic patients; wherein, they emphasised accurate alignment and congruency of the joint. In this study, resection and prosthetic replacement for malignant bone tumor of the distal tibia were performed in two patients. Both patients were able to achieve rapid relief from pain and early return to maximum function of the limbs without major complications. Good early function, excellent stability of ankles, and long term local control of the tumor at the latest follow-up were observed. The patients still achieved 80% of the pre-morbid functional capacity at the time of last follow up and remained satisfied. Lee et al. [1] reported that talar collapse progressed until the talar component reached to the level of the subtalar joint; however, replacement of the talar surface in this study is not necessary to prevent talar collapse. Disadvantages are inevitable since the custom-made prosthesis takes 6 weeks to manufacture and there is risk for late failure and subsequent need for surgery.

Limb-salvage surgery using tumor prosthesis has an acceptable functional outcome in the medium-term analysis of two patients. This technique provides a safe and effective method of stabilization for properly selected malignant bone tumors of the distal tibia. To avoid amputation, it is important that patients with a suspected malignancy should be referred to a specialist center.

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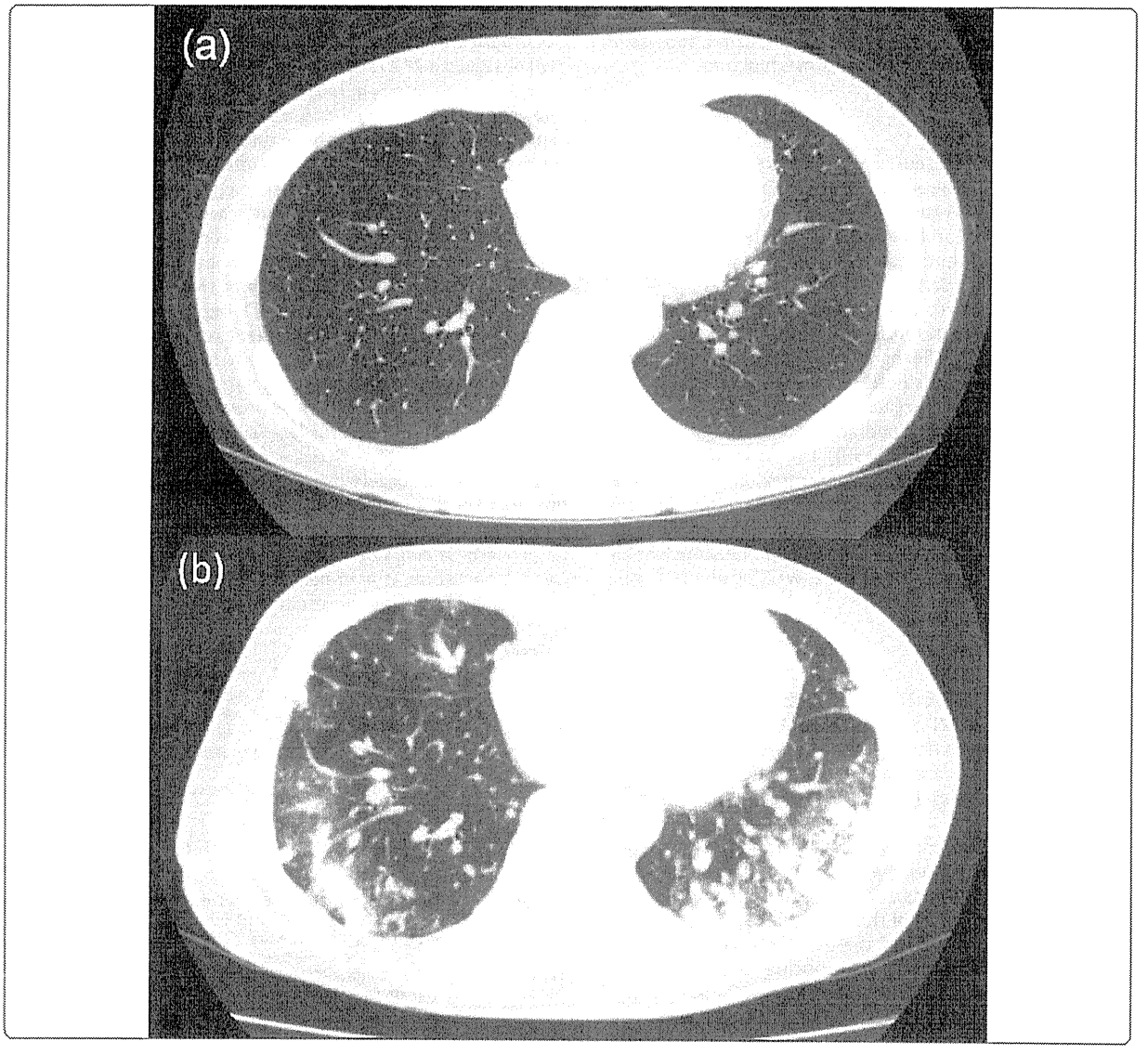

Case of an unusual clinical and radiological presentation of pulmonary metastasis from a costal chondrosarcoma after wide surgical resection: A transbronchial biopsy is recommended

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CASE REPORT Open Access

Case of an unusual clinical and radiological presentation of pulmonary metastasis from a costal chondrosarcoma after wide surgical resection: A transbronchial biopsy is recommended

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Abstract

Chondrosarcomas are the most frequently occurring primary malignant chest wall tumors. Furthermore, the lungs serve as the most frequent sites for metastases. Pulmonary metastases from sarcomas usually appear as round nodules of varying sizes on roentgenograms. Here, we report an unusual clinical and radiographic presentation of pulmonary metastasis from a costal chondrosarcoma. Bilateral pulmonary metastases developed soon after wide surgical resection. Thoracic computed tomography revealed unusual radiological findings: consolidation accompanied with ground-glass opacity. To confirm the metastasis, we recommend a transbronchial biopsy in cases where unusual pulmonary findings are detected.

Background

Chondrosarcomas are the second most frequent primary malignant bone tumors, after osteosarcomas [1,2]. They are also the most common primary malignant chest wall tumors: 5-15% of chondrosarcomas are located in the thoracic wall [3]. Since radiotherapy and chemotherapy are generally ineffective against chondrosarcomas, surgery is the only curative treatment, and the quality of the surgery is an essential prognostic factor [2]. Enneking et al. classified surgical margins into wide, marginal, and intralesional [4]. A wide resection is accomplished by a procedure in which the lesion, its pseudocapsule and/or reactive zone, and a surrounding cuff of normal tissue are taken as a single block. Therefore, resection for chest wall chondrosarcoma should be wide, taking intact pleura internally, intact muscle fascia externally, and transverse rib resection > 2 cm from the tumor on both directions [4,5]. Clinically, the involved rib en bloc should be resected along with the 2 intercostal spaces above and below the tumor.

On roentgenograms, pulmonary metastases usually appear as multiple peripheral, round nodules of varying sizes. Here, we describe an atypical presentation of pulmonary metastasis occurring soon after wide surgical resection of a costal chondrosarcoma. In this case, a thoracic computed tomography (CT) scan showed consolidation, predominantly in both the lower lobes, surrounded by ground-glass opacities and air bronchograms, mimicking serious pneumonia.

Case presentation

A 62-year-old woman was admitted to our hospital because of a mass that grew gradually in the right lateral chest wall for 1 year. Physical examination revealed a tumor (5 × 3.5 cm) in the right eighth rib. The mass was hard with an unclear border, no mobility, redness, or local heat, but it was tender. An X-ray revealed a mass with coarse calcification located on the right eighth rib, expanding beyond the irregular cortex. Thoracic CT revealed a 70 × 60 × 30 mm low-density mass

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(CT value, +18 HU) along the right eighth rib; it arose at the bone-cartilage border and destroyed these tissues (Figure 1a-c). No pulmonary metastasis was observed (Figure 2a). Other metastatic workup, including PET scan, was negative. The physical examination and imaging findings strongly indicated primary chondrosarcoma. Therefore, wide surgical resection was performed without performing a biopsy; the tumor was resected together with the right seventh, eighth, and ninth ribs. Transverse rib resection was performed >4 cm from the tumor in both directions. The chest wall was reconstructed using a Dexon mesh® (US Surgical, Connecticut, USA). Histological examination revealed a grade II chondrosarcoma with increased cellularity and myxoid stroma (Figure 3). All resected surgical margins were wide. The postoperative course was uneventful, and the patient was discharged 2 weeks after the operation.

However, 7 weeks after the definitive surgery, she presented with a slight fever, dyspnea, persistent dry cough, and purulent nasal discharge of 1-week duration. The white blood cell count (WBC)and C-reactive protein (CRP) level were 8.2 × 10^9 cells/L (neutrophils, 75%; lymphocytes, 16%; monocytes, 4.7%) and 3.7 mg/dL (normal: <0.30 mg/dL) respectively. Findings of other biochemical and serologic tests were normal. The chest roentgenogram showed air-space consolidation accompanied with an air bronchogram in the right upper and left lower lung fields (Figure 4) - a finding highly suggestive of bacterial pneumonia. Antibiotics (tazobactam/piperacillin [TAZ/PIPC]) administered for 7 days showed no results. Thoracic CT revealed pulmonary non-segmental consolidation, predominantly in the peripheral lung field, surrounded by ground-glass opacities; bronchovascular bundle thickness and interlobular septal thickness were absent (Figure 2b). Bronchoscopy and consequent transbronchial biopsy revealed blood vessel proliferation in the bronchial wall. Therefore, we considered this as a case of interstitial pneumonia such as cryptogenic organizing pneumonia, and initiated glucocorticoid therapy without waiting for the biopsy results. However, 3 days after the onset of the treatment, transbronchial biopsy sample through the left S8 bronchus confirmed the same histological features as the primary tumor in the peritumoral lumen structure, which was negative for CD34 and D2-40 (Figure 5a, b). The bronchoalveolar lavage fluid culture was negative. The patient died 12 weeks after the definitive surgery.

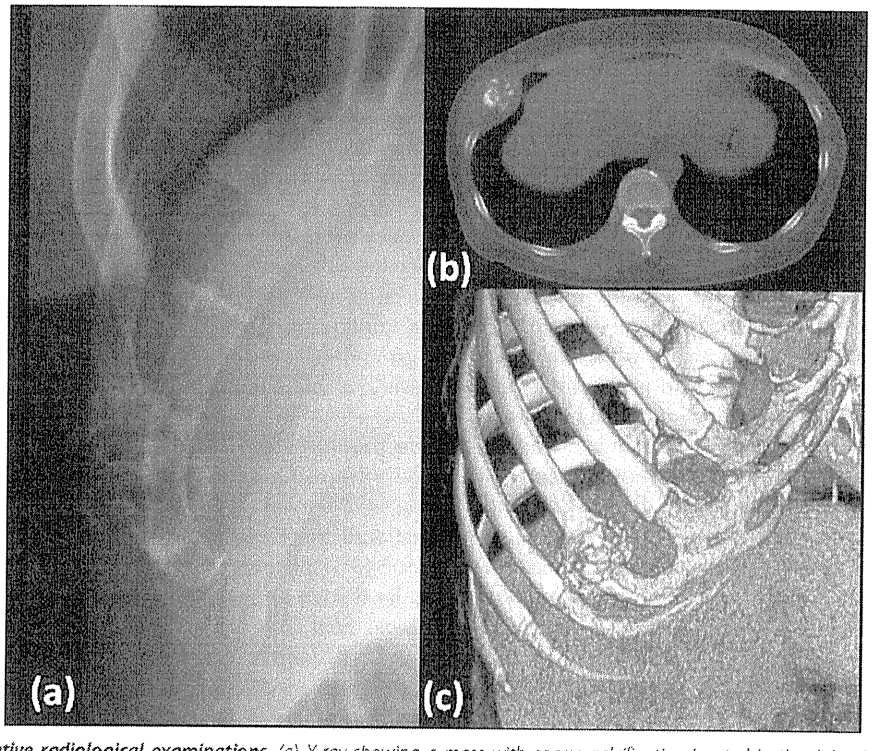

Figure 1 ***Preoperative radiological examinations.*** *(a) X-ray showing a mass with coarse calcification located in the right eighth rib, expanding beyond the irregular cortex. (b) CT scan showing a low -density mass with coarse calcification along the right eighth rib; the mass arose at the bone-cartilage border. (c) 3D-CT scan showing destruction of bone and cartilage destruction, with expansive growth of the tumor at the right eighth rib.*

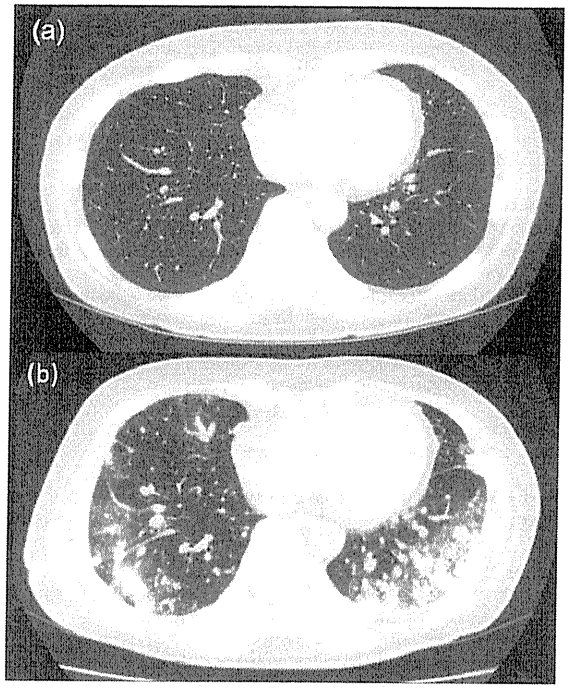

Figure 2 *Chest CT scan*. *(a) Preoperative CT scan showing no pulmonary metastasis. (b) Postoperative CT scan showing pulmonary non-segmental consolidation, predominantly in the peripheral lung field, with surrounding ground-glass opacities; no bronchovascular bundle thickness or interlobular septal thickness was observed.* The tumor was resected together with the right seventh, eighth, and ninth ribs.

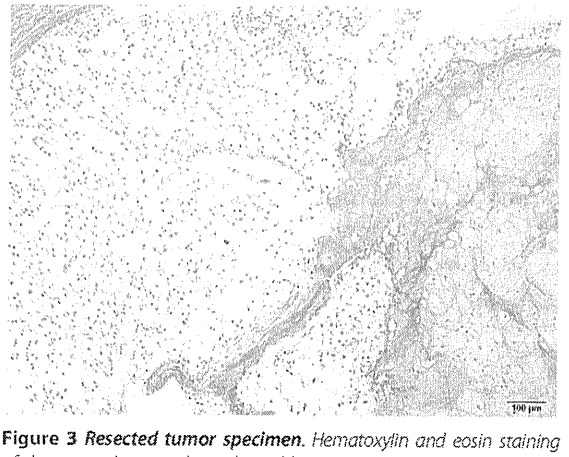

Figure 3 *Resected tumor specimen*. *Hematoxylin and eosin staining of the resected tumor showed a mild increase in cellularity and nuclear atypia. Doubly nucleated cells were seen in the field.*

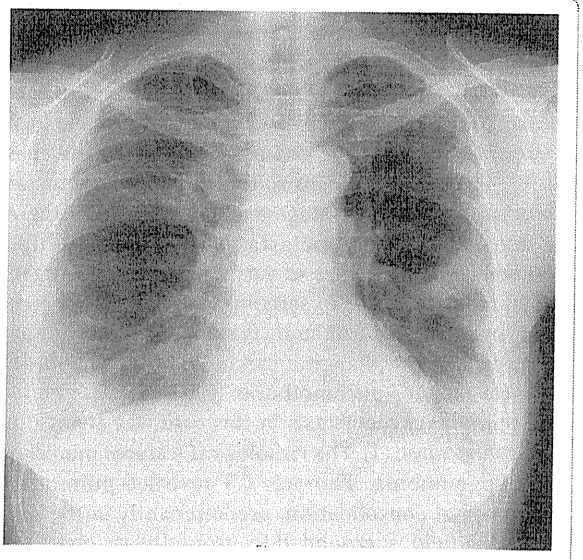

Figure 4 *Chest X-ray*. *Chest roentgenogram showed air-space consolidation with an air bronchogram, predominantly in the right upper and left lower lung fields.*

Discussion

Chondrosarcomas are classified on the basis of their aggressiveness into 3 grades according to their cellular density, degree of anisokaryosis, and nuclear hyperchromatism [6]. The histologic grades of chondrosarcoma correlate well with prognosis, especially for metastases [6]. The most frequent site of metastasis is the lungs; other sites include the bones, brain, regional lymph nodes, and liver [5]. The metastasis rates for grades I, II,

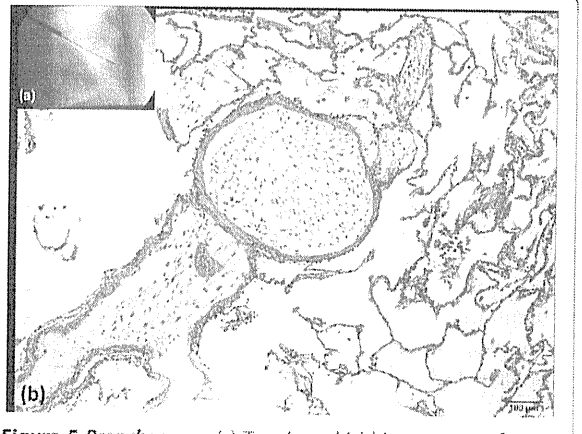

Figure 5 *Bronchoscopy*. (a) Transbronchial biopsy was performed through the left S8 bronchus. *(b) Hematoxylin and eosin staining of the biopsy sample showed a bone tumor in the lumen structure, with the same histological features as the primary bone tumor.*

and III tumors were 0, 13, and 23%, respectively [5]. The incidence of pulmonary metastases varies with the primary tumor and stage of disease. Bone tumors such as osteosarcomas and Ewing's sarcoma show a high incidence of pulmonary metastases. Pulmonary metastasis develops from 20% of the chondrosarcomas of the chest wall [5]. The most common route for pulmonary metastasis of sarcomas is hematogenous dissemination; therefore, most pulmonary metastases appear as multiple peripheral, round nodules of varying sizes on roentgenograms. However, certain sarcomas such as osteosarcomas present with unusual features of pulmonary metastasis, i.e., lymphangitic carcinomatosis, endobronchial metastasis, or pneumothorax [7,8].

The pulmonary metastasis in this case was atypical in the following ways: (1) The radiological features mimicked those of pneumonia. Thoracic CT revealed pulmonary non-segmental consolidation, predominantly in the peripheral lung field, surrounded by ground-glass opacities. This indicated interstitial pneumonia such as cryptogenic organizing pneumonia. (2) Although the operation involved only the right side, bilateral pulmonary metastases developed after the resection. Time taken for metastasis to develop has been reported to be an average of 20 months [2]. In this case, bilateral pulmonary lesions rapidly developed into metastases. Thus, histologic examination was needed in order to confirm the diagnosis.

Transbronchial biopsy, endobronchial biopsy, or surgical lung biopsy can be performed to obtain tissue specimens. Surgical lung biopsy includes video-assisted thoracic surgery (VATS) and open lung biopsy. The procedure chosen is based on clinical judgment, which entails weighing the yield versus the risk to the patient. In particular, transbronchial biopsy is usually the procedure of choice for the initial examination due to its high yield and relatively low risk [9], and therefore, we chose this approach. The transbronchial biopsy revealed pulmonary metastasis from costal chondrosarcoma although the mechanism underlying the pulmonary metastasis remains unknown. The possibility of lymphangitic carcinomatosis was eliminated because of the absence interlobular septal thickness.

Soon after the curative surgery is performed, to confirm the pulmonary metastasis, we recommend that transbronchial biopsy should be performed in cases where unusual clinical and radiological pulmonary findings are detected.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-chief of this journal.

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Authors' contributions

ME: assisted in the writing of the manuscript and in the orthopedic workup of the patient; KH: assisted in the drafting of the manuscript and in the orthopedic workup of the patient; TK: assisted in the writing of the manuscript and performed the radiological evaluation; KN: performed the radiological evaluation; YT: performed the pathological evaluation; NN: assisted in the orthopedic workup of the patient; NA: evaluated critically the manuscript and gave final approval for the manuscript to be published. All authors read and approved the final manuscript.

Competing interests

The authors declare that they have no competing interests.

Received: 8 February 2011 Accepted: 16 May 2011
Published: 16 May 2011

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doi:10.1186/1477-7819-9-50
Cite this article as: Emori *et al.*: Case of an unusual clinical and radiological presentation of pulmonary metastasis from a costal chondrosarcoma after wide surgical resection: A transbronchial biopsy is recommended. *World Journal of Surgical Oncology* 2011 9:50.