| | | |
|---|---|---|
| Beta-HCG | <0.1 ng/ml | <0.1 ng/ml |
| NSE | 13.2 ng/ml | <10.0 ng/ml |
| VMA/Cr | 7.7 mg/gCr | |
| HVA/Cr | 18.9 mg/gCr | |
| CEA | <0.9 ng/ml | <2.5 ng/ml |
| Ferritin | 96.1 ng/ml | 3120 ng/ml |

Laparotomy confirmed a well-encapsulated solid tumor arising from the lesser curvature of the stomach and extending into the lumen and retroperitoneum. The tumor was not adherent to adjacent structures. Resection of the tumor was performed, with a 0.5-cm gastric wall margin taken. The tumor was positive in the surgical margin near the esophagogastric junction (EGJ) by intraoperative pathological assessment. Additional resection was performed for the margin. The tumor was 5.5×4.4×4.0 cm in size and weighed 70 g. Histological examination demonstrated an immature teratoma with a variety of components derived from endoderm, ectoderm, and mesoderm, including mature cartilage, alimentary tract epithelium, and immature central nervous system. No yolk sac histology was observed.

The infant's postoperative course was uneventful. Serum AFP levels decreased to <10 ng/ml by 7 months after the operation and were maintained at 7.3 ng/ml for 12 months. However, at the child's 24-month check-up, AFP had risen to 356.2 ng/ml. CT and magnetic resonance imaging (MRI) revealed an enhanced tumor under the left lobe of the liver and the right side of the spleen (Fig. 2). These findings prompted a second-look laparotomy, which was performed 26 months after the initial surgery. The recurrent tumor originated from the greater curvature of the stomach, locally invading the lumen and more growing toward the retroperitoneum surrounding the stomach. The recurrent tumor origin was near the surgical margin of the first operation. It was adherent to the spleen and the left lateral lobe of the liver. The tumor was removed en bloc with the spleen, the left lateral lobe of the liver, and regional lymph nodes. The pathologic diagnosis was YST without any area of immature teratoma (Fig. 3). By standard histologic microscopic evaluation, no invasion was observed to the spleen or the liver. Likewise, no metastasis was seen to any regional lymph nodes.

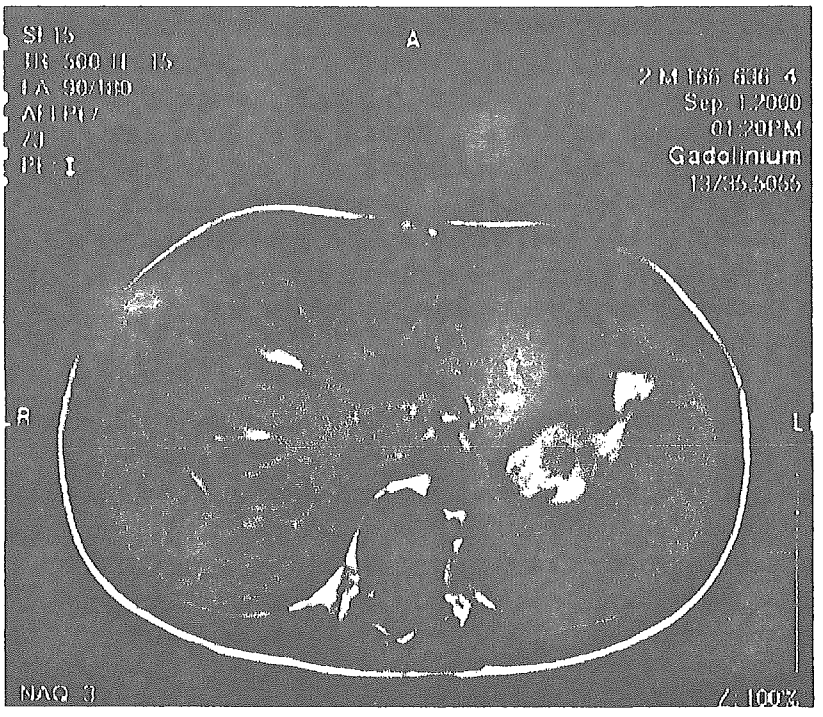


**Fig. 2** Magnetic resonance imaging clearly outlines a tumor of high signal intensity on gadolinium-enhanced T1-weighted images (*T1WI*) 2 years after the first operation

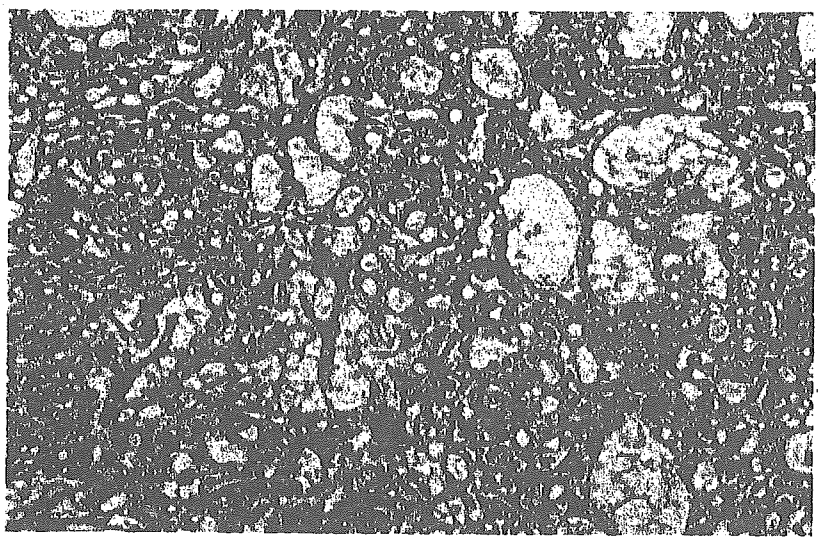

**Fig. 3** Photomicrograph of the tumor resected at the second operation. The pathological diagnosis was pure yolk sac tumor (*YST*) without immature teratoma components (hematoxylin and eosin staining, original magnification ×100)

After the operation, chemotherapy with four courses of PVB regimen (cisplatin, 20 mg/m$^2$ ×5 days; vinblastine, 0.2 mg/kg ×2 days; bleomycin, 15 mg/m$^2$ ×1 day) was administered. No recurrence has been observed 3.5 years after the second operation and 3 years after the end of chemotherapy.

## Discussion

At birth, teratomas present mainly as coccygeal tumors, whereas in the first 6 months teratomas are predominantly localized in the testis. Later, between the ages of 9 and 15 years, ovarian teratomas are seen [*13*]. Gastric teratomas are very rare and exhibit distinguishing characteristics compared with teratomas from other organ sites. For example, most gastric teratoma patients are male (90%) [*2*], whereas the other teratomas predominate in females. Most gastric teratomas described in previous papers were in infants less than 1 year of age. In addition, gastric teratomas have predominantly exhibited benign behavior. Only a few cases of gastric teratomas with malignant components have been reported [*10–12*]. A case of malignant transformation in an adenocarcinoma arising from immature teratoma in an 83-year-old male was reported [*14*]. Gastric teratoma recurring as a malignant lesion has never been reported.

Our patient's teratoma was diagnosed as an immature teratoma. Immature teratomas are the most controversial and least well-characterized germ cell tumors in children [*15*]. The prevalence of microscopic foci of YST is said to be directly related to the grade of the malignant potential and to be the only valid predictor of recurrence in pediatric immature teratoma. Researchers have reported that elevations of serum AFP concentration greater than 100 ng/dl almost always indicated the presence of foci of YST [*15*]. In our case, the concentration of serum AFP was 80,050.0 ng/ml, within the normal range for a 5-day-old patient. Positive AFP immunoreactivity was not found in the tumor resected at the first operation. However, because clusters of YST cells may be very small or associated intimately with immature neural tissue, or both, and because they frequently do not stain positively for AFP, they are easily overlooked [*16*]. In one series, half of relapsing patients showed highly malignant tumor histology at relapse (mainly YST and in a few cases embryonal

carcinoma) [13]. Relapses were observed for patients with mature as well as immature teratomas. Tumors have had foci of YST in immature teratoma. The recurrent YST in our patient originated near the initial surgical margin but was apart from the EGJ that was positive by intraoperative pathological assessment.

The most common germ cell tumors of childhood are sacrococcygeal teratomas, which have been reported to have a malignant potential of 17% [17]. A case of sacrococcygeal teratoma with immature elements was reported with local recurrence as malignant teratoma [18]. Without coccygectomy, local recurrence of sacrococcygeal teratomas has been reported and was postulated to have arisen from the microscopic remnants of the primary tumors [19]. Our observations suggest that this mechanism may have led to relapse in our patient. This is the first time gastric teratoma has been reported to recur locally with a malignant histology.

The serum concentration of AFP was a good indicator for tumor recurrence in our patient. The AFP concentration—which had been 7.3 ng/ml 12 months after the operation, indicating normal range for the patient's age—was abnormally elevated compared with age-appropriate normal controls when found to be 356.2 ng/ml 24 months later, and CT and MRI confirmed tumor recurrence. MRI was useful in that it made the tumor contour stand out against surrounding viscera. MRI clearly indicated a tumor of low T1 but high T2 signal intensity, with gadolinium enhancement (Fig. 2). A gallium and bone scintigram were negative.

For treating the recurrent tumor, we selected en bloc surgical resection together with surrounding viscera firmly adherent to the tumor. Complete surgical resections are recommended as the most effective treatments for teratomas with or without malignant elements [20].

After the surgery, we chose the PVB (cisplatin, vinblastine, and bleomycin) regimen as an antineoplastic chemotherapy. Vincristine, actinomycin D, vinblastine, bleomycin, doxorubicin, cisplatin, and etoposide have proved effective in treating various tumors of germ cell origin [16]. Cisplatin and its incorporation into combination regimens has resulted in a substantial increase in disease-free survival. VP16 may cause secondary leukemia [21], and ifosfamide may cause severe multiorgan toxicity [22]. Thus, we chose PVB, expecting stronger effects than regimens without cisplatin and weaker side effects than regimens with VP16 or ifosfamide. Four courses of the same PVB regimen that we used are the standard therapy for stage 1 YST in Germany, where favorable outcomes are reported [23].

Our patient has been free of recurrence for 3.5 years after the second operation. Although immature gastric teratomas are considered to have benign behavior, a pediatric surgeon should consider the possibility of recurrence, and periodic follow-up checks with AFP tumor marker measurement should be mandatory.

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