研究成果の刊行に関する一覧表

書籍

著作氏名	論文タイトル名	書籍全体の編集者名	書籍名	出版社名	出版地	出版年	ページ
嘉山孝正 櫻田香 毛利渉 齋野真 佐藤慎哉	優位半球頭頂葉腫瘍の手術	山下純宏	脳腫瘍の外科 Biological behavior にのっとった新しい治療戦略	メディカ出版	大坂	2005	24-29
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Nagata S Iwaki T Sasaki T Hayashi K					

Ⅳ. 研究成果の刊行物・別冊

脳腫瘍の外科

—Biological behaviorにのっとった新しい治療戦略—

金沢大学脳神経外科 名誉教授　山下 純宏 編

MC メディカ出版

優位半球頭頂葉腫瘍の手術

Surgery for tumors within dominant parietal lobe

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抄　録

優位側頭頂葉の腫瘍の摘出の際，注意すべき領域はarea 1, 2, 3の後中心回とarea39の角回，area40の縁上回である．後中心回周囲の腫瘍に関しては，錐体路との関係も把握する必要がある．腫瘍が後中心回に限局している場合や，後方より前中心回を圧迫しているものは，ある程度の感覚障害は残存するが，原則として腫瘍全摘出術が可能である．しかしながら，後中心回より深部白質を経て前中心回へ浸潤するような腫瘍は，全摘出は不可能である．一方で角回，縁上回に限局するような腫瘍は基本的には覚醒手術で摘出術を行っている．我々はシルビウス裂を遠位端まで開放し，そこから後方へアプローチすることで腫瘍を摘出している．自験例を提示しつつ我々の方法を紹介する．

Abstract

When we remove the tumors locating dominant parietal lobe, we have to keep in mind to preserve eloquent areas such as postcentral, angular and supramarginal gyri. In addition, a tumor arising from post central gyrus, somteimes compress or invade pre central gyrus or pyramidal tract. So it is also important to preserve motor function during surgery for the tumors locating even in the parietal lobe. Using appropriate monitoring, we could totally remove the tumors locating within post central gyrus or just compressing the pyramidal tract. However, when the tumors are invading pyramidal tracts, they were never totally removed without aggravation of motor function.

Functionl brain mapping by awake surgery could be very useful to remove the tumors surrounding angular or supramarginal gyri. We could successfully remove the tumor adjusting angular gyrus thorough transsylvian approach after functional brain mapping.

Key words: parietal lobe, angular gyrus, monitoring, mapping

序　文

頭頂葉腫瘍に対する手術方針は，前頭葉腫瘍に対する手術方針と異なり一定の見解がないように思われる．どこの領域までをeloquentと考えるかは，ある程度術者や患者の考え方にもよると思われるが，摘出により，明らかなADLの低下をきたす場合は，その領域をeloquentと考えることには異論がないように思われる[1]．今回は優位側頭頂葉においてeloquentな部位と考えられるpost central gyrus（area3, 1, 2）近傍，angular gyrus，supramarginal gyrus（area39, 40）近傍腫瘍に対する，我々の治療方針を紹介する．

Post Central Gyrus近傍腫瘍

Post central gyrus近傍腫瘍は以下の3群に分類される．

1. Post central gyrus内に限局している腫瘍
2. Post central gyrusより発生し，Pre central gyrusを圧排している腫瘍
3. Post central gyrusより発生し，Pre central gyrusあるいは深部白質に浸潤している腫瘍

この鑑別は通常，MRIのみでは困難である．術前検査としてMEG（magnetoencephalography），diffusion tensor image（DTI）やtractographyなどの検査が治療方針決定に有用である[5,6]．また術前の患者の運動機能評価はきわめて重要な指標であり，軽度の麻痺を呈する症例は上記の2の群に含まれることが多い．術中はSEP, MEP, subcortical stimulationなどのモニタリングが必須である[2,3]．

1）Post central gyrus内に限局している腫瘍

原則的に全摘出可能と考えている．Primary sensory cortexに限局する腫瘍の場合は，症状は感覚鈍麻，あるいはしびれ感，しびれ発作といったものが主症状であるが，軽度の麻痺を示している例もあり，錐体路との関係を充分に術前，術中に検討する必要がある．

症例1：23歳，女性，右利き

- 主訴；てんかん発作
- 現病歴；2001年2月に単純部分発作（右下肢）で発症．2002年2月5日 右下肢にはじまり，右半身から全身に広がる全身けいれんを起こし，近医受診．MRI上異常所見を認め，2002年4月11日 当科紹介入院となった．
- 神経学的所見；右下肢のhypesthesia (9/10), dysesthesia，単純部分発作（右下肢）（薬物コントロール不良）．
- 放射線学的所見；MRI上，病変はpost central gyrus内側に限局していると考えられ（Fig. 1A），MEGによりpost central gyrus内に存在することを確認した（Fig. 1B）．
- 手術所見；post central gyrusに存在する

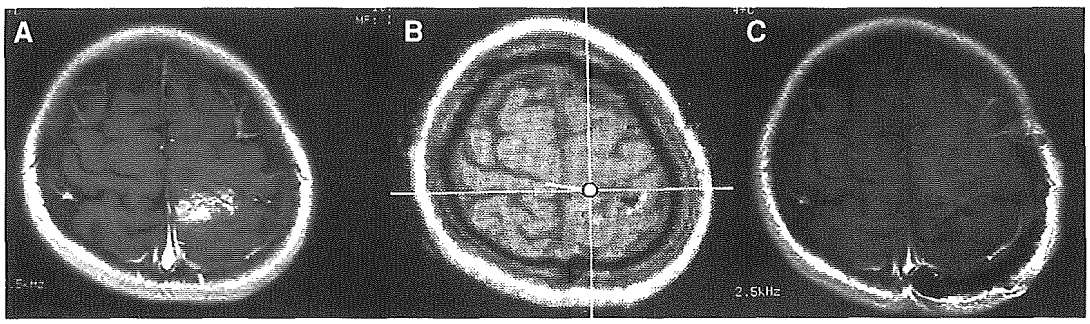

Fig. 1　Case 1

A: Preoperative gadolinium-enhanced axial T1-weighted MR imaging showing the lesion within left post central gyrus.
B: Preoperative magnetoencephalography (MEG) confirming the lesion within left post central gyrus.
C: Postoperative gadolinium-enhanced axial T1-weighted MR imaging showing total removal of the lesion.

cavernous angiomaの診断で，手術を施行した．SEPよりcentral sulcusを同定し，MEPをモニターしつつ腫瘍を摘出した．

• 術後経過；術後の経過は良好であり，MRI上腫瘍は全摘出され（Fig. 1C），明らかな麻痺を認めなかった．下肢のしびれ感は残存するも，てんかん発作は消失した．

2）Post central gyrusより発生し，Pre central gyrusを圧排している腫瘍

Post central gyrusより発生していても，Pre central gyrusあるいはpyramidal tractの圧排のため，中等度の麻痺があることもある．術前に腫瘍と，pyramidal tractとの関係を検討し，術中，適切なモニタリングを行えば麻痺の改善が期待できる．

症例2：64歳，女性，右利き

• 主訴；左上下肢の麻痺

• 現病歴；2002年10月より左下肢の脱力感があったが放置していた．しかし改善しないため，2003年2月24日 近医受診し，MRI上異常所見を認め，2003年3月4日 当科紹介入院となった．

• 神経学的所見；左上下肢麻痺（DeJong 3/4程度）で歩行は不可能であった．

• 放射線学的所見；MRI，MEG上，病変はpost central gyrus後方より発生し，深部はPre central gyrusを前方に圧迫していた（Fig. 2A, B）．tractographyではpyramidal tractは腫瘍により外前方に圧排されていた（Fig. 2C）．

• 手術所見；Post central gyrusに存在するgliomaの診断で，手術を施行した．SEPよりcentral sulcusを同定し，MEPをモニターしつ

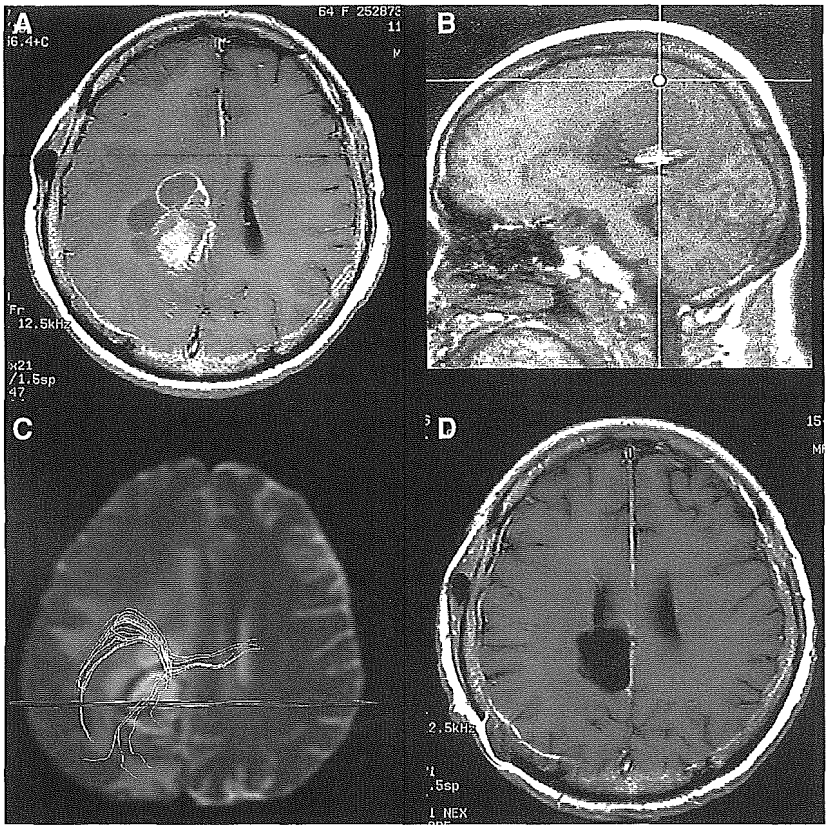

Fig. 2 Case 2

A: Preoperative gadolinium-enhanced axial T1-weighted MR imaging showing the multi-cystic tumor within right parietal lobe.
B: Preoperative magnetoencehalography (MEG) confirming the tumor within left parietal lobe.
C: Preoperative diffusion tensor tractography showing the pyramidal tract shifted by the tumor.
D: Postoperative gadolinium-enhanced axial T1-weighted MR imaging showing total removal of the tumor.

つ腫瘍を摘出した．術中MEPの低下は認められなかった．

• 術後経過；術後の経過は良好であり，MRI上腫瘍は全摘出され（Fig. 2D），麻痺もDeJong 3から4に回復し歩行可能となった．病理組織学的診断はAnaplastic pilocytic astrocytomaであった．術後，放射線化学療法を行い，現在も腫瘍の再発は認めていない．

3）Post centra gyrusより発生し，Pre central gyrusあるいは深部白質に浸潤している腫瘍

原則的には全摘出は不可能である．MEPなどのモニタリングも麻痺が重度な場合（DeJong 2以下）ではモニタリング自体が不可能である[3]．重度麻痺の再発症例等に対しては生命予後を重視し，十分なinformed consentが得られれば全摘出も例外的に許容されるが，その場合，症状の悪化は通常避けられない．

症例3：51歳，女性，右利き

• 主訴；左上下肢の麻痺

• 現病歴；1998年6月より右上下肢の脱力感あり徐々に悪化したため，1998年8月31日 近医受診し，MRI上異常所見あり，1998年9月29日当科紹介入院となった．

• 神経学的所見；右上下肢麻痺（DeJong 4/4程度）を認めた．

• 放射線学的所見；MRI上，病変はcentral sulcusをまたいでpre central，post central gyrus両方に浸潤していた（Fig. 3A）．

• 手術所見；Lt. fronto-parietal gliomaの診断で，手術を施行した．SEPよりcentral sulcusを同定し，MEPをモニターしつつ，post central gyrusに存在する腫瘍のみを部分摘出した．

• 術後経過；術後，MRI上腫瘍は部分摘出された（Fig. 3B）．術後の麻痺の悪化は認めなかったが下肢のしびれを認めた．病理組織学的診断はdiffuse astrocytomaであった．術後放射線化学療法を施行したが6ヶ月後再増大を認めた．

Angular gyrus近傍腫瘍

Angular gyrus（area39），Supramarginal gyrus（area40）は通常，優位半球においては，いわゆるGerstmann syndromeに関係した領域であり，その近傍の腫瘍には注意を要する．areas 39, 40に限局している腫瘍は，頻度は低いと考えられるが，側頭葉腫瘍上後方に位置するもの，前頭葉腫瘍で下後方に位置するもの（シルビウス裂遠位端）は病変がarea39, 40に関係する可能性があり，注意が必要である．

側脳室三角部の腫瘍などはその部位を避け皮質切開を行えば問題ないが，皮質直下の腫瘍に

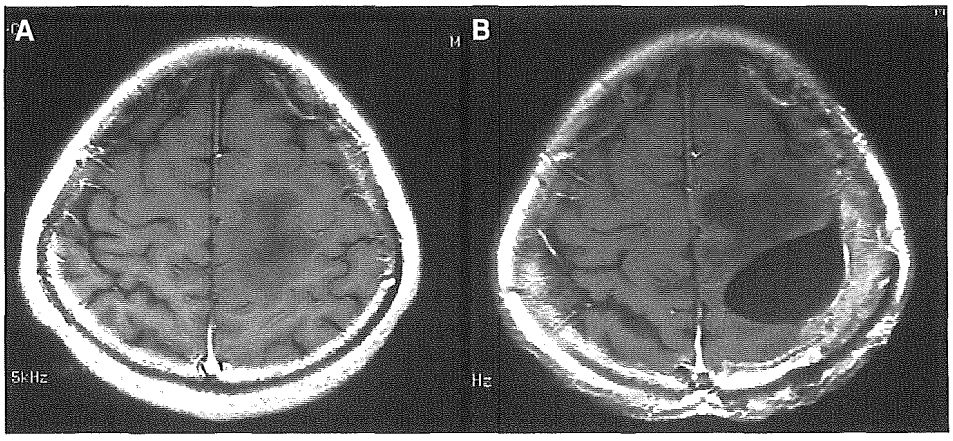

Fig. 3 Case 3

A: Preoperative gadolinium-enhanced axial T1-weighted MR imaging showing the tumor invading beyond the central sulcus.
B: Postoperative gadolinium-enhanced axial T1-weighted MR imaging showing partial removal of the tumor.

対してはそのアプローチには注意が必要である．我々は経シルビウス裂内よりAngular gyrus直下の腫瘍の摘出を試みた．

症例4：44歳，女性，右利き

• 主訴；語尾の間違い

• 現病歴；2002年10月頃より，接客中に言葉がうまく話せないことに気づく．3日間継続し自然軽快したが，その後，精査のためMRIを施行され，異常が認められたため当科紹介となった．

• 神経学的所見；明らかな異常所見は認められなかった．

• 放射線学的所見；MRI上，側頭葉上側頭回からシルビウス裂遠位端を囲むように深部白質に存在していた（Fig. 4A）．

• 手術所見；Lt. T-Pのcavernous angiomaの診断で摘出術を施行した．覚醒手術下に脳機能マッピングを行った．病変はWernicke's area の後方に存在していた（Fig. 4B）．深部白質の線維の温存を企図とし，シルビウス裂遠位端を開放し，内側面に言語機能がないことを確認した上で，内側面に切開を加え腫瘍に到達，摘出した．

• 術後経過；術後経過は良好で，MRI上腫瘍は全摘出され（Fig. 4C），現在まで明らかな神経学的異常所見は認められない．

考　察

頭頂葉腫瘍の手術に際して，注意するポイントは優位，劣位にかかわらず，まず第1に頭頂葉から前頭葉に進展する腫瘍の摘出の際に錐体路を傷害しないよう心がけることである．上述のごとく，術前の評価を適切に行い，手術のプランニングを適切に行わなくてはならない．また術中もモニタリング等を行い，無理な摘出によ

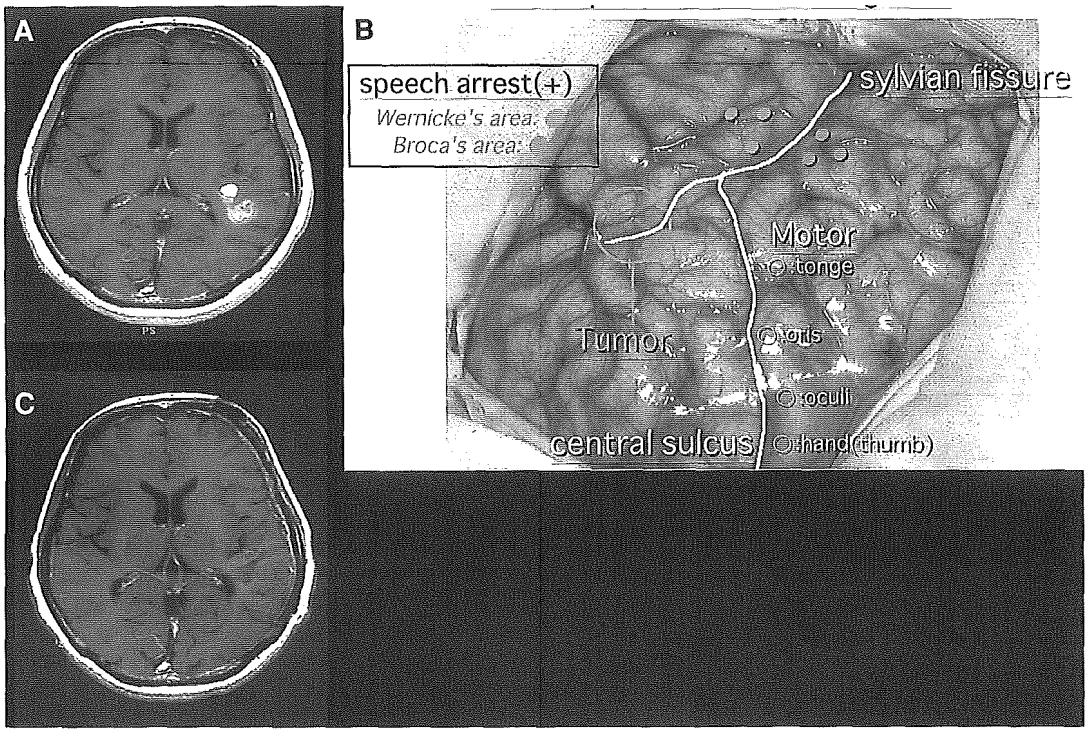

Fig. 4 Case 4

A: Preoperative gadolinium-enhanced axial T1-weighted MR imaging showing the tumor adjusting angular gyrus.
B: The result of functional brain mapping created by awake surgery.
C: Postoperative gadolinium-enhanced axial T1-weighted MR imaging showing partial removal of the tumor by transsylvian approach.

り症状を悪化させないよう心がけるべきである．

次に優位側頭頂葉腫瘍に特異的な部位としてAngular gyrus，Supramarginal gyrusが挙げられる．その近傍に腫瘍が存在する場合は，原則的には覚醒手術を行い，脳機能マッピングを行うべきである．しかしながら覚醒手術は，体位や麻酔の問題から腫瘍が大きい場合，後方に進展している腫瘍の場合は，実施が困難なことが多い．しかし自験例のように比較的小型で側頭葉より後方に伸展している腫瘍は，脳機能マッピングが可能と考える．本例ではシルビウス裂内側よりアプローチし腫瘍を摘出したが，Superior temporal，Inferior frontal，Angular，Supramarginal gyri直下に腫瘍が存在する場合，有用なアプローチと考える[4]．

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SECTION I

CHAPTER 96

GERM CELL TUMORS IN CHILDREN

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The histopathologic entity of primary intracranial germ cell tumors includes a number of subtypes such as germinoma, germinoma with syncytiotrophoblastic giant cells (STGCs), mature teratoma, immature teratoma, yolk sac tumor, choriocarcinoma, and mixed germ cell tumors that include several types of components of germ cell tumors.[11] Even though the symptoms and radiologic appearances are similar, the prognoses and outcome of primary intracranial germ cell tumors are extremely different and depend on tumor type type.[2,6,22]

In this chapter, the characteristic findings of germ cell tumor in children are described. In addition, the prognostic factors for survival and quality of life are presented. Finally, we summarize the most recent data from the literature on primary intracranial germ cell tumors treated with surgery, radiotherapy, and chemotherapy.

EPIDEMIOLOGY

Intracranial germ cell tumors consist of rare and varied histologic types of brain tumors. A higher incidence of germ cell tumors in Asian countries compared with Western countries has been reported. The incidence of pineal region tumors in Asia is higher (3.0%) than in Europe and the United States (0.4 to 1.0%). Of the germ cell tumors limited to the pineal region in Japanese and Korean studies, pure germinoma was found in 70.3% and 80.2%, respectively. In European and U.S. studies, pure germinomas comprise 53.6% and 58.3% of pineal region germ cell tumors, respectively. Age and gender distributions of pineal region germ cell tumors show a high incidence in males and in children. The sex ratio of the occurrence of germ cell tumor in the pineal region shows a marked male predominance, with a male to female ratio of 51:5.5. Males predominate for all the different subtypes of germ cell tumors. As for age distribution, germinoma, embryonal carcinoma, and malignant teratoma have a peak age incidence at 10 to 14 years, whereas choriocarcinoma and teratoma are widely distributed through ages 10 to 20 years.

As for the germ cell tumors in the suprasellar region, the sex ratio between males and females is almost equal. Compared with the pineal region, there is a tendency for suprasellar germ cell tumors to be nongerminomatous germ cell tumors, especially immature teratoma.

LOCATION

As with other extragonadal germ cell tumors, intracranial germ cell tumors tend to arise in the midline. More than 80% of intracranial germ cell tumors arise in the region of the pineal gland, followed by tumors in the suprasellar region. Other sites of origin of intracranial germ cell tumors include intraventricular, basal ganglia, thalamic, cerebral hemispheric, bulbar, intramedullary, and intrasellar. Multifocal or germ cell tumors usually involve the pineal region and suprasellar compartment simultaneously (synchronously) or sequentially (metachronously).

DIAGNOSIS

Clinical Features

The clinical manifestations of germ cell tumors and their duration vary with histologic type and lôcation. Tumors of the pineal region often compress and obstruct the cerebral aqueduct, resulting in progressive hydrocephalus with intracranial hypertension. Also, pineal region tumors may compress and invade the tectal plate, producing a characteristic paralysis of upward gaze and convergence known as Parinaud's syndrome. Suprasellar tumor typically compresses the optic chiasm, causing visual field deficits, and often disrupts the hypothalamus as suggested by the occurrence of diabetes insipidus and manifestations of pituitary failure, which include growth retardation and delayed sexual maturation. Germ cell tumors may also cause precocious puberty by pineal or hypothalamic destruction or by the elaboration of human chorionic gonadotropin (HCG), a stimulant of testosterone production that is secreted by neoplastic syncytiotrophoblasts located in the tumor.

Tumor Markers

Tumor marker levels in the serum or cerebrospinal fluid (CSF) provide quite reliable data for the diagnosis of the germ cell tumor type, even for primary intracranial mass lesions. HCG-β is mostly elevated with the diagnosis of germinoma with STGCs. In cases of choriocarcinoma, the levels of HCG-β sometimes exceed 10,000 ng/mL in the serum, and they correlate well with a poor prognosis. α-fetoprotein (AFP) is another

reliable marker for the diagnosis of yolk sac tumor, and levels greater than 4000 milli-international units per milliliter are indicative of a poor prognosis. Sometimes the serum levels of carcinoembryonic antigen (CEA) are elevated in cases of embryonal carcinoma. Finally, placental alkaline phosphatase (PLAP) is positive at times in immunohistochemical studies of germinoma.

Imaging

The diagnosis of germinoma may be suggested on plain computed tomography (CT) when nodular clusters of calcification in the pineal region are present. Usually, hydrocephalus can be seen because of obstruction of the aqueduct by the mass. Germ cell tumors usually appear as solid masses that are isodense or slightly hyperdense relative to gray matter. Following contrast administration, the germ cell tumors typically show prominent enhancement on CT or magnetic resonance imaging (MRI). Suprasellar germ cell tumors may initially cause only enhancement of the pituitary stalk until a later stage, when the tumor may infiltrate the optic chiasm. In cases of teratoma, cystic regions may also be identified as part of the mass. An intratumoral hemorrhage seen on imaging studies strongly suggests the diagnosis of choriocarcinoma.

Histology

Whereas germinoma and teratoma typically occur as pure tumor subtypes, choriocarcinoma, yolk sac tumor, and embryonal carcinoma occur as mixed germ cell tumor types. Of all the pineal region germ cell tumors in Japan, the most common tumor subtype is germinoma, accounting for 70.3% of tumors, followed by immature teratoma at 14.7%.

TREATMENT

Most malignant germ cell tumors other than germinoma are associated with a poor prognosis; however, recent progress in surgical techniques and effective chemoradiotherapy has improved the prognosis.

Surgery

The surgical procedures that are used for germ cell tumors include a shunt operation or third ventriculostomy for hydrocephalus, tumor biopsy, or aggressive tumor resection. It remains somewhat controversial as to whether surgery of germ cell tumors should be the first line of therapy. For example, in Western countries there is a tendency to select surgery, but in Asian countries radiotherapy or chemotherapy is selected as the first treatment, because radiochemosensitive germinoma is predominantly found for tumors in the pineal region. Certainly the role of radical surgery for germinoma has never been proved. However, for nongerminomatous germ cell tumors, total resection of the mass lesion may improve the prognosis.

Radiation Therapy

Radiation therapy has long been known to be effective for germinoma. After the diagnosis is made, germinomas are treated by radiation therapy that comprises treatment of the whole ventricle or focal boost cranial irradiation. Whole-brain irradiation was used in the past, but this practice has largely disappeared. If spinal seeding has been demonstrated, the spine is incorporated into the radiation fields. Because of some of the deleterious effects of radiation therapy on the central nervous system (CNS), current approaches have explored the possibility of reducing or even eliminating radiation therapy.[15] Recently, it has been recommended that the radiation dose and volume for germinomas be reduced to 24 to 30 Gy in a localized field (including the entire ventricular systems).[3,21] The 5-year cumulative survival rate in patients who received reduced radiotherapy with chemotherapy has been almost equal to that of those who were treated with full-dose irradiation alone. Finally, in cases of other malignant germ cell tumors, the prognosis is still unsatisfactory, even after full-dose irradiation with chemotherapy.

Chemotherapy

In efforts to reduce radiation dose and to improve prognosis, combination chemotherapy with radiation therapy has been generally accepted.[1] Of the chemotherapeutic agents used to treat germ cell tumors, cisplatin, carboplatin, vinblastine, etoposide, bleomycin, or CCNU are generally effective. Several drug combinations such as PVB (cisplatin, vinblastine, bleomycin), PE (cisplatin, etoposide), and ICE (ifosfamide, carboplatin, etoposide) have been shown to have particular promise. Today, PE therapy may be the chemotherapy treatment of choice for germ cell tumors. Interestingly, chemotherapy alone without radiotherapy is currently not recommended because many patient recurrences have been reported.[13] 2

OUTCOME—SURVIVAL

Outcome for germ cell tumors varies widely among differing histologies. Of the cumulative survival rates for various types of germ cell tumors, germinoma shows the highest survival rate at 89.2%. In the case of germinoma, it is known that diffuse dissemination of disease at diagnosis is not a risk factor for poor prognosis. On the other hand, embryonal carcinoma, yolk sac tumor, and choriocarcinoma have worse survival rates than the other types of germ cell tumors (Table 96-1).[12] 3

FUNCTIONAL OUTCOME

Functional outcome after treatment for germ cell tumors can be assessed by examining performance status, neurologic deficits, hormonal dysfunction, and brain injury after radiation therapy.

Performance Status

Yoshida et al analyzed performance status after radiochemotherapy for germ cell tumors. They showed a performance status score of 75% for pure germinoma, 75% for germinoma with STGCs, and 57% for nongerminomatous germ cell tumors. In this report, the response rate for all germ cell tumors was 67%.[25] In long-term survivors, if patients had received radiation therapy with a total dose of more than 30 Gy, cognitive dysfunction was found in some cases. In general, intellectual decline correlates directly with total radiation dose.

Follow-up MRI studies in patients who have received cranial irradiation will typically show cerebral atrophy. In one study that examined patients treated for germ cell tumors with a mean follow-up of 99 months, only 13% of patients maintained a performance status of 100%.[20]

Though not included as an assessment item for performance status, sterility caused by cisplatin therapy, which is an indispensable chemotherapeutic agent for germ cell tumors, should be seen as a major problem for long-term survivors. In addition, etoposide, which is one of the major drugs used in the treatment for germ cell tumors, is known to induce secondary neoplasms, albeit at a low rate.

Neurologic Deficits

After neurosurgical treatment for tumor excision or insertion of a CSF shunt, hydrocephalus is usually well controlled. For tumors originating around the optic pathway, there may be a high frequency of visual impairments affecting acuity and fields. Some patients will demonstrate limitations in upward gaze palsy as a result of involvement of tumor with the quadrigeminal plate. High-tone hearing loss has been observed after cisplatin therapy, which can also cause peripheral neuropathy.

4

Hormonal Disorder

For patients with suprasellar lesions, hormone replacement therapy is a common requirement after treatment, affecting almost 80% of patients. Diabetes insipidus or anterior pituitary dysfunction is common. Treatment of the former with DDAVP is usually long term. Saki et al emphasized that pituitary dysfunction present before treatment persisted or even worsened after patients went into remission, except for patients with small and localized masses on admission.[17]

5

Cerebral Injury after Radiation Therapy

Cerebral injury from radiation therapy is usually seen following whole-brain irradiation of 30 Gy or more. On MRI, generalized brain atrophy, multiple cerebral lacunae infarction, and high signal intensity areas in the white matter representing demyelination are generally observed. Associated with cerebral atrophy are mental and physical deterioration, hypothalamic or endocrinologic dysfunction, and impaired quality of life. A feared complication of radiation therapy is a radiation-induced secondary neoplasms. Sawamura et al also described 4 patients from a total of 84 intracranial germ cell tumor patients (4.7%) who developed radiation-induced neoplasms, including two glioblastomas and two meningiomas.[20] Moreover, there is another report describing a 12% occurrence of secondary neoplasms over a 20-year period.[13]

As an illustrative case, we describe a 14-year-old boy with a solid mass in the suprasellar area who underwent biopsy and proved to have an immature teratoma. Chemoradiation therapy was given, and a total radiation dose of 60 Gy was administered. After these treatments, MRI showed complete remission of tumor. The patient did well in school until 6 years after the initial treatment when he complained of headache and depression. A follow-up MRI study revealed a low-intensity lesion in the right basal ganglia with mild brain atrophy. After 6 months, a repeat MRI scan showed progressive brain atrophy (Figure 96-1). This case is typical for delayed brain injury induced by radiotherapy.

SUBCLASSIFICATION OF INTRACRANIAL GERM CELL TUMORS

According to the World Health Organization (WHO) classification of intracranial germ cell tumors, germinoma, embryonal carcinoma, yolk sac tumor (endodermal sinus tumor), choriocarcinoma, mature teratoma, immature teratoma, teratoma with malignant transformation, and mixed germ cell tumors are the main types.[11]

Germinomas

Pure Germinoma

Pure germinoma may be cured with a better than 90% 5-year survival rate using radiotherapy alone. In the report of Japan's Brain Tumor Registry in 2000, a remarkable improvement in cumulative survival for germinoma by radiation therapy was shown.[4] Several issues concerning quality of life, however, are known to be induced by radiotherapy. Therefore combined chemoradiotherapy has been used to reduce the total radiation dose to the brain. Nowadays, it is generally accepted that patients with germinoma can be cured by preirradiation chemotherapy followed by reduced doses of irradiation.

According to the analyses by the Brain Tumor Registry in 2000, relative survival rates for germinoma at 1-year, 2-year, and 5-year intervals were 96.6%, 94.3%, and 91.2%, respectively.[4] Brandes et al showed a 5-year survival rate of 96%.[3] Recently, Sano also showed 5- and 10-year survival rates of 96% and 93%, respectively.[19]

Germinoma with Syncytiotrophoblastic Giant Cells

Approximately 13% of germinomas contain syncytiotrophoblastic giant cells (STGC) positive for HCG-β.[21,22] This type of tumor has been shown to have a different response to chemoradiotherapy. Although the response rate is high, the tumor mass tends to regress much more slowly, and a complete response rate is generally lower than for pure germinoma. Also, germinomas with elevated HCG-β levels in serum or CSF are considered to have a higher risk of recurrence.[9] One report of patients with germinoma with elevated CSF HCG-β levels showed a 40% recurrence rate even after complete conventional radiotherapy.

To illustrate this point, a 15-year-old male who complained of headache received a MRI scan with gadolinium that showed an enhanced solid mass in the pineal lesion. CSF and serum HCG-β levels were elevated. After treatment with chemotherapy and focal radiotherapy, the MRI revealed complete remission. Four years after treatment, this patient had cerebellar ataxia. Repeat MRI showed complete remission of the original pineal lesion but diffuse dissemination of disease in the brain and spinal cord (Figure 96-2).