Primary embryonal-cell carcinoma of the parietal lobe

Case report

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A case of primary embryonal-cell carcinoma of the parietal lobe is reported. The unusually chronic presentation of such a malignant tumor is described. The atypical computerized tomography and magnetic resonance imaging characteristics of this lesion are presented. Review of the literature yielded no previous reports of a lobar embryonal-cell carcinoma. The rarity of intracranial germ-cell tumors presenting off the midline is discussed.

Key Words • brain neoplasm • embryonal-cell carcinoma • germ-cell tumor • parietal lobe

Primary intracranial embryonal-cell carcinomas are rare brain tumors that tend to occur along the midline. As a group, they constitute less than 0.5% of all intracranial neoplasms.5 Of the primary intracranial germ-cell tumors, only about 5% are embryonal-cell carcinomas; the majority are germinomas. When Jennings, et al.,5 reviewed the literature on intracranial germ-cell tumors in 1985, they found that fully 95% arose in the midline. We describe a primary intracranial embryonal-cell carcinoma that arose in the right parietal lobe of a 7-year-old boy.

Case Report

This 7-year-old right-handed boy first presented at the age of 5 years with a focal motor seizure involving the left arm. His neurological examination was normal. A computerized tomography (CT) scan at that time showed a well-demarcated, serpiginous, low-density lesion in the white matter of the right parietal lobe. There was no enhancement (Fig. 1). The patient was treated with phenobarbital and had no further seizures. For 18 months, serial CT scans showed no change in the lesion; then left focal seizures recurred, this time with both motor and sensory involvement. Repeat CT (Fig. 2) showed that the lesion had enlarged and there was enhancement at its superior extent. There was no surrounding edema. The T1-weighted magnetic resonance (MR) images (Fig. 3) showed a well-circumscribed low-intensity lesion in the posterosuperior portion of the right parietal lobe. The lesion exhibited high intensity on the T2-weighted images; only subtle mass effect was present and, again, there was no edema. After the administration of gadolinium, T1-weighted MR images demonstrated an enhancing nodule. The patient was referred for neurosurgical evaluation.

Examination. Physical examination showed normal fundi. There was no testicular mass. Neurological examination was normal except for mildly decreased graphesthesia and stereognosis on the left side.

Operation. At right parietal craniotomy the surface of the brain appeared normal. A paramedian corticectomy was made and at a depth of approximately 1 cm a soft, grayish, nonencapsulated glassy intra-axial mass was encountered with the gross appearance of a low-grade astrocytoma. There was no cyst or nodule. A moderately good plane between the tumor and white matter was found and all apparent tumor was removed.

Pathological Examination. Pathological examination revealed a malignant germ-cell tumor (Fig. 4). Some areas had the appearance of a germinoma, others of an embryonal-cell carcinoma. The mitotic rate was high in all areas. Stains for human chorionic gonadotropin and alpha-fetoprotein were negative. Placental alkaline phosphatase was present in some cells.

Postoperative Course. Postoperatively, the patient had a mild left-sided hemiparesis and a parietal sensory deficit, both of which resolved over 1 month. A CT
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**Fig. 1.** Contrast-enhanced computerized tomography scan performed when the patient first presented with a focal seizure. There is a nonenhancing low-attenuation lesion in the right parietal region.

**Fig. 2.** Noncontrast-enhanced (left) and contrast-enhanced (right) computerized tomography scans obtained prior to the operation, 19 months after the scan shown in Fig. 1. The right parietal low-attenuation lesion has increased in size and assumed a more globular shape. There is enhancement of the rim.

**Fig. 3.** Magnetic resonance images obtained just prior to the operation. Left: T₁-weighted image (TR 650 msec, TE 20 msec) showing a well-circumscribed low-intensity lesion in the superior aspect of the right posterior parietal lobe. Center: T₁-weighted image (TR 650 msec, TE 20 msec) after administration of gadolinium-diethylenetriamine penta-acetic acid showing enhancement in the lateral posterior portion of the lesion. Right: T₂-weighted image (TR 3000 msec, TE 90 msec) showing a brightly intense signal within the lesion.

**Fig. 4.** Photomicrographs of the lesion showing sheets and cords of embryonal carcinoma cells with prominent nucleoli, as well as a lymphocytic infiltrate. H & E, × 190 (left); × 300 (right).
scan showed no evidence of residual tumor. Chest x-ray studies and total myelography were normal. Cerebrospinal fluid (CSF) cytological testing was negative for malignant cells. Serum and CSF samples were negative for alpha-fetoprotein, human chorionic gonadotropin, and carcinoembryonic antigen. Preirradiation chemotherapy was administered consisting of a regimen of cisplatin and etoposide alternating with cyclophosphamide and etoposide every 3 weeks for four courses. This was followed by external beam irradiation with sequential field reductions such that 5000 cGy was delivered to the tumor bed and 2340 cGy to the whole brain. At 2 years following surgery, the patient is neurologically intact and active in school and sports.

Discussion

Classification of Germ-Cell Tumors

Germ-cell tumors include germinomas, teratomas, embryonal-cell carcinomas, endodermal sinus tumors, and choriocarcinomas. Some reports use the terms "embryonal-cell carcinoma" and "endodermal sinus tumors" interchangeably. Russell and Rubinstein proposed that they should be differentiated according to the dominant cell type: tumors with mostly solid sheets of cells are embryonal-cell carcinomas, and those with embryoid bodies, alpha-fetoprotein droplets, and Schiller-Duval bodies are endodermal sinus tumors. Because mixed nongerminomatous germ-cell tumors are frequent, Jennings, et al., classified them according to the most malignant cell type present. Whether one uses the pathological classification of Russell and Rubinstein or Jennings, et al., the lesion reported here is properly classified as embryonal-cell carcinoma.

Literature Review of Embryonal-Cell Carcinoma

Review of the literature yielded no previous reports of lobar embryonal-cell carcinoma. These tumors tend to occur in the midline, either in the pineal or the suprasellar region. Jellinger reviewed 17 primary intracranial germ-cell tumors; all three of his embryonal-cell carcinomas were found in the midline. Packer, et al., reported six embryonal-cell carcinomas; five located in the pineal gland and one in the suprasellar region. Björnsson, et al., reviewed 70 intracranial germ-cell tumors and found two embryonal-cell carcinomas. The location of one was not known, and the other was located in the "temporo-occipital region." Jennings, et al., reviewed the 389 published cases of primary intracranial germ-cell tumors, and found that 21 (5%) were embryonal-cell carcinomas; none of these was located off the midline. In addition, all the cases described by Russell and Rubinstein occurred in the diencephalopineal region. Ono, et al., reported an embryonal-cell carcinoma of the basal ganglia.

Thus, it appears that our case may be unique. Although the possibility exists that the intracranial tumor represents a metastasis from an occult primary site, after a follow-up period of more than 21 months no extracranial tumor has become evident.

Other Germ-Cell Tumors

The other varieties of intracranial germ-cell tumors do, on rare occasions, occur off the midline. However, most of these examples are in the basal ganglia or associated with the ventricular system. Germinomas have infrequently been found in the basal ganglia or in the lateral ventricle, and parietal lobe have been reported. Finally, almost all cases of teratoma also arise in the midline.

Clinical Features

Our patient was 5 years old when his tumor first became symptomatic. The majority of embryonal-cell carcinomas develop in males between the ages of 10 and 15 years, but the appearance of an embryonal-cell carcinoma in a 6-year-old boy is not unusual. What is unusual, however, is the long interval prior to diagnosis in this case (19 months). Typically, germ-cell tumors are diagnosed within 6 months after symptoms begin because of rapid growth.

Another unusual feature of this case is the radiographic appearance of the lesion. Embryonal-cell carcinoma, like most of the other germ-cell tumors, tends to appear denser than surrounding brain on the precontrast CT scan and enhances brightly, homogeneously more often than heterogeneously. In our case, the tumor was initially an entirely low-density mass, even after administration of contrast material. Only much later did a small area of enhancement develop. It should be noted that germinomas located in the basal ganglia and thalamus are more likely to have an atypical CT appearance. Cysts, calcification, and heterogeneous enhancement may be seen.

Embryology

Germ-cell tumors are thought to derive from primordial germ cells. These cells can first be identified in the endoderm of the fetal yolk sac. From there, they migrate to the urogenital ridges, which are paramedian structures. Although most of the primordial germ cells reach their targets within the gonadal ridges, others disseminate widely along the midline and then persist in the diencephalopineal and mediastinal regions. It is not known why extragonadal germ cells tend to persist there.

Evidence for the hypothesis that germ-cell tumors derive from primordial germ cells was summarized by Gonzalez-Crusi. First, germ-cell tumors are most common in the gonads, where most of the germ cells are located. Second, extragonadal germ-cell tumors,
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whether sacrococcygeal, mediastinal, or dienephaloneural, tend to occur in the midline, which corresponds to the migration of the primordial germ cells in the embryo. Third, primordial germ cells of fetal mice and embryonal carcinoma cells from adult mice are morphologically identical. If one accepts the "germ-cell theory" on the origin of germ-cell tumors, then it can be postulated that the embryonal-cell carcinoma reported here arose from germ cells that strayed off the midline. Such a developmental error would represent only a very short deviation from the normal migratory pattern in the embryo. Jennings, et al., suggested that the changes occurring in the diencephalopineal region at the time of puberty might induce malignant behavior in the germ cells residing there. Thus, gonadotropins would act as transforming agents. This is consistent with the sharply defined age range of these tumors. If a germ-cell tumor develops in a prepuberal boy, in the present case, then perhaps the precursor cells required only low levels of exposure to gonadotropins for transformation. These considerations must remain speculative, however.

Conclusions

Our patient has done well after resection, combination chemotherapy (similar to the regimen employed by Kobayashi, et al., for recurrent intracranial germ-cell tumor), and radiation therapy. He remains alive and well 2 years after diagnosis, exceeding the expected period of survival. Our experience suggests that primary intracranial embryonal-cell carcinoma should be included in the differential diagnosis of a cerebral hemisphere lesion in the pediatric patient.

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References


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