Peripheral-type primitive neuroectodermal tumor arising in the tentorium

Case report

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The authors report the case of a peripheral primitive neuroectodermal tumor (PNET) arising in the tentorium in a 5-year-old boy who presented with frequent vomiting and mild palsy of the left abducent nerve. Following complete surgical excision of the tumor via a transtentorial approach, the patient has thus far been disease free for 7 years. The tumor tissue was composed of small cells with uniform round nuclei and minimal identifiable cytoplasm. Homer–Wright rosettes were frequently observed. Immunohistochemical studies demonstrated a positive reaction to HBA-71, which recognizes the cell surface glycoprotein p30/32, a product of the MIC2 gene. Both the clinical and immunohistochemical characteristics of this tumor are consistent with a diagnosis of peripheral PNET, which is genetically distinct from the more common intracranial PNET.

KEY WORDS • brain neoplasm • Ewing’s sarcoma • primitive neuroectodermal tumor • tentorium

Both peripheral primitive neuroectodermal tumors (PNETs) and Ewing’s sarcomas are small cell tumors that arise in peripheral soft tissue or bone. They constitute a family of tumors with a common cytogenetic translocation t(11; 22)(q24; q12)21,23 and the same highly consistent pattern of protooncogene expression.17 Immunoreactivity to HBA-71, which recognizes the cell surface glycoprotein p30/32 (a product of the MIC2 gene), is a highly sensitive histochemical marker of this family of tumors.17 Intracranial PNETs, which were originally defined by Hart and Earle,10 represent another well-known group of small cell tumors arising in the brain.6,14,20 In this group of tumors, a loss of heterozygosity on 17p is the most common finding2,16,19 and immunoreactivity to HBA-71 is usually absent,13 indicating that intracranial PNETs are genetically distinct from those of the peripheral type.

Because peripheral PNETs and Ewing’s sarcomas originate in the soft tissue or bone at various locations in the body, it seems theoretically possible that the same tumors could also arise within the intracranial cavity. We previously reviewed 10 cases of Ewing’s sarcoma of the cranial bone as reported in the literature and by us;5 to the best of our knowledge, however, no report of this type of tumor originating in the intracranial soft tissue has yet been published. In this paper we report the case of an intracranial tumor arising in the tentorium, which on light microscopy and immunohistochemical analysis demonstrated characteristics consistent with a peripheral PNET. We describe a peripheral PNET originating at a very unusual location and discuss the biological behavior of this rare type of tumor, which appears to warrant aggressive surgical intervention even when it arises intracranially.

Case Report

History. This 5-year-old boy had been in excellent health until he presented to another clinic with frequent vomiting. After a computerized tomography (CT) scan revealed a large intracranial mass lesion, he was referred to our service.

Examination. On admission the patient was alert and fully oriented. Mild palsy of the left abducent nerve was noted. No destruction of the bone structures was detected
on plain craniograms or bone window CT scans. The mass appeared on the CT scans as a well-circumscribed tumor that encompassed the left tentorium and extended into the middle as well as the posterior fossae. The tumor consisted mostly of an isodense mass, which was strongly enhanced after intravenous injection of contrast medium, together with a cystic component and high-density spots. Magnetic resonance (MR) imaging demonstrated the tumor to be hypodense on both T1- and T2-weighted images and enhanced by intravenous infusion of gadolinium–diethylenetriamine pentaacetic acid (DTPA) (Fig. 1). The tentorium was also enhanced in the region encompassed by the tumor. No perifocal edema was noted. The left temporal lobe, brainstem, and cerebellum were markedly compressed and deformed (Fig. 1 and Fig. 2 left). Angiograms of the internal and external carotid arteries revealed only displacement of the normal intracranial vessels. No evidence of tumor was detected in any other organ.

Operation. The patient underwent surgical removal of the tumor via a transpetrosal approach. The mass was found to be located wholly within the intracranial cavity and attached to the tentorium. There was no bone involvement. The tumor was readily separated from the temporal lobe, brainstem, and cerebellum and was totally removed by piecemeal excision, leaving the arachnoid membrane intact. The portion of the tentorium to which the tumor was attached was resected by using a wide surgical margin.

Postoperative Course. The patient’s postoperative course was uneventful. Magnetic resonance imaging yielded no evidence of a residual tumor mass (Fig. 2 right). A cerebrospinal fluid puncture procedure that was repeated several times before and after surgery revealed no evidence of leptomeningeal dissemination. The patient underwent whole-brain and local radiation therapy (50 Gy in total). Intrathecal injections of methotrexate were given weekly for 2 weeks and then monthly for 6 months. Further chemotherapy was not administered because the patient’s family refused consent. When last seen 7 years postsurgery, the patient was in excellent health. A complete battery of neuroimaging studies revealed no evidence of recurrence or leptomeningeal dissemination.

Pathological Findings. On conventional light microscopy examination the tumor tissue was found to be composed of small cells containing uniform round nuclei with minimal identifiable cytoplasm (Fig. 3 left). Tumor cells were also found within the resected tentorium. Homer–Wright rosettes were frequently observed. Immunohistochemical studies demonstrated a positive reaction to HBA-71 (Fig. 3 right) but no reaction to vimentin, neuron-specific enolase, S-100 protein, synaptophysin, or glial fibrillar acidic protein.

Discussion

The extracerebral location and immunoreactivity to HBA-71 of the tumor in our case strongly suggested that it belonged to the peripheral-type PNET family. These tumors range in form from the least differentiated (Ewing’s sarcoma) to the more differentiated (for example, peripheral PNETs).4,9,11,18 In a new classification scheme proposed by Schmidt, et al.,18 immunoreactivity to at least two neural markers and/or the presence of Homer–Wright rosettes are required for a diagnosis of PNET. Based on the presence of these rosettes, we reached a diagnosis of peripheral PNET in the present case.

In our literature review we found four cases of intracranial PNETs that were located within the extracerebral space.3,8,12,15 However, most of these cases presented primarily with leptomeningeal dissemination, presumably from an intracranial PNET. No examinations for immunoreactivity to HBA-71 were performed in these cases. The present case thus appears to represent the first unequivocal example of a peripheral PNET arising within intracranial soft tissue.

The distinction between PNETs of the peripheral type and intracranial PNETs is important clinically because these two types of tumors carry different prognoses.4,9,10,14,18 Among patients with the latter type, long-term survival is extremely rare and many die within 1 year of diagnosis.4,10,14 For example, Dirks, et al.,7 recently reported that the 5-year survival rate in their patients with intracranial PNETs was only 18% despite combined surgery, radiation therapy, and chemotherapy. In contrast, many patients with peripheral PNETs achieve long-term survival.
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Although disease-free survival \(9,11,18\) despite the fact that these tumors also display relatively aggressive biological behavior. Schmidt, et al.,\(^{18}\) reported that the disease-free survival rates at 7.5 years in patients with peripheral PNETs and Ewing’s sarcoma are 45% and 60%, respectively. Patients with Ewing’s sarcoma of the cranial bone have also survived free of disease for many years. The patient with Ewing’s sarcoma of the cranial bone previously reported by us\(^{22}\) has thus far achieved a disease-free survival time of 9 years following complete surgical excision of this tumor. The favorable clinical course observed in the present case is consistent with the features of peripheral rather than intracranial PNETs.

In addition to its biological characteristics, the location of the tumor in the case reported here may have played an important role in securing a good outcome for our patient. Complete excision was possible because of the sharply circumscribed margin of the tumor and the absence of brain involvement or leptomeningeal dissemination. Although further accumulation of cases will be necessary before definite conclusions can be reached, it should be emphasized that lengthy disease-free survival times can be attained in cases of peripheral PNETs arising in the intracranial soft tissue when complete surgical excision is achieved. In contrast to intracranial PNETs, this type of tumor clearly warrants aggressive surgical intervention.

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References


Fig. 3. Photomicrographs showing the tumor tissue to be composed of cells with uniform round nuclei containing minimal cytoplasm. H & E (left) and immunohistochemical staining for HBA-71 (right). Dark reaction products in the cytoplasm represent immunoreactivity to HBA-71. Bar = 50 μm.

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