MR of intracranial neuroblastoma with dural sinus invasion and distant metastases.

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Primary cerebral neuroblastoma is a rare entity that has been classified within the broader category of primitive neuroectodermal tumors (PNET). We present a case of primary cerebral neuroblastoma that had invaded the dural sinuses and metastasized to the lungs at the time of diagnosis. MR imaging provided an excellent means of localizing the extent of the primary tumor as well as identifying cases at risk for systemic metastases.

Case Report

A 3-year-old girl presented with a 2-month history of rapidly progressive loss of vision and crossed eyes. The parents reported that the child was irritable and lacked coordination. There was no history of seizures, nausea, or emesis. Physical examination was significant for mild developmental delay, marked irritability, bilateral papilledema, and a right esotropia. Pupils were sluggish, but reactive, bilaterally. The remainder of the neurologic examination was normal. Visual acuity was not properly assessed because of poor patient cooperation.

An unenhanced CT scan obtained at another hospital revealed a large left occipital mass containing irregular areas of high attenuation interpreted as calcification. MR imaging was performed at our institution on a 1.5-T unit. We obtained T1-weighted spin-echo images without and with gadopentetate dimeglumine, proton-density-weighted, and T2-weighted spin-echo images, which all confirmed a left parietooccipital mass measuring 8.5 cm. Unenhanced T1-weighted and T2-weighted images revealed a mass that was predominantly isointense with gray matter with large irregular areas of high signal consistent with hemorrhage (Fig. 1A). Abnormal soft tissue, isointense with the tumor on all pulse sequences, was identified in the torcula and both proximal transverse sinuses. A small signal void indicating flowing blood was seen adjacent to the intrasinus mass. Encasement of the straight sinus was present with tumor invasion through the tentorium into the superior vermis (Figs. 1B and 1C). T2-weighted axial images revealed invasion through the falx as well as high signal areas flanking the main tumor mass representing cyst formation or tumor necrosis (Fig. 1D). There was minimal peritumoral edema. Contrast-enhanced T1-weighted sagittal images demonstrated marked irregular enhancement of the tumor and intrasinus mass (Fig. 1E). Preoperative differential diagnosis included PNET and cerebral or meningeal sarcoma.

A subtotal resection of the left occipital lobe mass was performed and a central venous line was placed for chemotherapy. The immediate postoperative chest radiograph showed a right upper lobe mass. No preoperative chest radiograph had been obtained. Subsequent chest CT scans confirmed the right upper lobe parenchymal mass (completely surrounded by air-containing lung) and revealed numerous other pulmonary mass lesions (Fig. 1F). No paraspinal mass was present. Total spine MR imaging with contrast enhancement, bone scan, and nuclear meta-iodobenzylguanidine scan were performed in the postoperative period and no additional metastases were identified.

Tissue samples were routinely processed for light and electron microscopic studies. On light microscopy, the neoplasm was composed of sheets of closely packed round small cells containing scanty cytoplasm. The nuclei were hyperchromatic. Four to 10 mitotic figures were seen at 400 x. Karyorrhexis was a common finding. In focal regions the tumor cells were larger with vesicular nuclei and globoid, pale cytoplasm. These cells were suspended in a highly fibrillar matrix. A few perivascular pseudorosettes, Homer Wright rosettes, and Flexner rosettes were seen. Staining with an antibody against glial fibrillar acidic protein showed foci of astrocytic differentiation.

On electron microscopy, most of the cells corresponded to undifferentiated elements with scanty cytoplasm containing large numbers of ribosomal rosettes. Occasionally, there were clusters of cells with more abundant finely fibrillar cytoplasm, moderate numbers of microtubules, numerous lysosomes, and well-developed Golgi apparatus. These cells were joined by desmosomal junctions. A few of the cells contained microvilli. No cilia were encountered. In some fields the tissue was composed of numerous cell processes. They contained empty and dense core vesicles, microtubules, and filaments (Fig. 1G). Synapticleike contacts were occasionally encountered (Fig. 1H). The electron microscopic findings indicated neuronal differentiation, supporting the diagnosis of neuroblastoma.

Following surgery, the patient began the Children’s Cancer Study Group’s brain tumor protocol of extensive chemotherapy and delayed radiation therapy. Over a 6-month period, she received six courses of systemic chemotherapy followed by external-beam radiation therapy to the occipital mass. Serial MR studies have shown enlargement of the occipital mass and decreased contrast enhancement. Chest CT has shown resolution of the pulmonary masses.

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Fig. 1.—3-year-old girl with 2-month history of rapidly progressive loss of vision and crossed eyes.
A, Left parasagittal T1-weighted (700/20/1) MR image shows irregular area of high signal intensity consistent with hemorrhage within the occipital tumor.
B, Sagittal T1-weighted (700/20/1) MR image shows occipital tumor traversing the tentorium with encasement of the straight sinus (arrow). Note that soft-tissue mass is isointense with tumor in the torcula (arrowhead).
C, Right parasagittal T2-weighted (2500/80/1) MR image shows invasion of tumor into right occipital lobe. Note that abnormal signal is isointense with tumor in right transverse sinus (arrow).
D, Axial T2-weighted (2500/80/1) MR image shows tumor invading the falx, spreading from left to right occipital lobe. The tumor is flanked by high-signal cystic or necrotic areas (arrows).
E, Left parasagittal T1-weighted (700/20/1) postcontrast MR image. The parenchymal and dural sinus components of the tumor show marked contrast enhancement.
F, Axial CT scan through lower chest reveals multiple pulmonary metastatic lesions.
G, Electron micrograph shows several cell processes filled with microtubules (asterisk). The process in the center contains empty and dense core vesicles (arrows) ranging in external diameter from 160 to 250 nm. (x24,000)
H, Electron micrograph shows numerous cell processes. A focal membrane condensation between two cell processes (arrow) is encountered. This suggests a synaptic contact and indicates the neuronal origin of this tumor. (x10,300)
Discussion

Primary cerebral neuroblastoma is a rare tumor that in the past—along with medulloblastoma, medullopithelioma, spongioblastoma, ependymoblastoma, pineoblastoma, and retinoblastoma—has been placed within the group of neoplasms categorized as primitive neuroectodermal tumors [1]. Controversy surrounds the precise histologic classification of cerebral neuroblastoma [2-5]. The classification of the tumor in the present case as neuroblastoma is best supported by electron microscopic findings, which showed cell processes containing dense core vesicles and synaptic contacts. These structures are a clear indication of neuronal differentiation [6].

Primary cerebral neuroblastoma occurs predominantly in children and young adults, with approximately 80% of cases occurring in the first decade of life. No sex predominance has been appreciated. Bennett and Rubinstein [7] cite a 5-year postoperative survival rate of 30%.

Neuroblastoma may occur in any region of the CNS as an intraparenchymal, intraventricular, or spinal cord mass. There is some predilection for the parietal and occipital lobes [8-10]. Just et al. [11] and Davis et al. [12] have recently reported on the MR findings of cerebral neuroblastoma, the latter having also compared these findings with CT. They described tumors that sometimes attained large size, frequently contained calcification and/or hemorrhage, and had cystic areas. Peritumoral edema was often absent or minimal. Dissemination throughout the subarachnoid space was seen occasionally. Enhancement with iodinated contrast medium or gadopentetate dimeglumine was frequent. MR examination showed mass lesions that were inhomogeneous but predominantly of low signal intensity on short TR images and of mixed low and high signal on long TR images. Our case demonstrated many of the above features, including large size, hemorrhage, lack of edema, cystic areas, marked enhancement, and similar signal intensities on MR images. Dural sinus invasion was encountered in the present case, although it was not seen in the series by Davis et al. [12]. Presumably, the sinus invasion allowed access by the tumor to distant sites, such as the lungs. This has been proposed previously in a report by Rubinstein [13], in which a nonoperated malignant astrocytoma invaded the superior sagittal sinus and metastasized to vertebral bone marrow and abdominal and pelvic lymph nodes. Systemic metastases to lung, liver, bone marrow, pericardium, cervical lymph nodes, and skin have been reported with primary cerebral neuroblastoma [10, 14]. The detection of dural sinus invasion should alert the radiologist and clinician to the possibility of distant metastases, which may alter treatment.

REFERENCES