Meningiomas in Children: MR and Histopathologic Findings

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PURPOSE: To present the MR and histopathologic findings in five children with meningiomas. **METHODS:** Five children aged 3 months to 16 years with pathologically proved meningiomas underwent preoperative contrast enhanced MR. Tissue in four patients was sent for chromosomal analysis in addition to routine histopathologic studies. **RESULTS:** All five tumors were extra-axial, two supratentorial and three in the posterior fossa. MR showed variable signal intensity on precontrast T1-weighted images. All of the tumors were hyperintense on proton density- and T2-weighted images and showed intense contrast enhancement. Histopathologic analysis showed two meningotheliomatous, one transitional, one chordoid, and one hemangiopericytic variant of meningioma. Chromosomal analysis showed deletions involving chromosome 22 in two of four tumors studied. **CONCLUSION:** Meningiomas in children have a higher incidence of posterior fossa location and different histologic types than seen in adults. MR showed the tumors in our patients to be extra-axial, hyperintense on proton density- and T2-weighted images with intense enhancement on postcontrast T1-weighted images. Chromosomal aberrations were noted in two patients.

Index terms: Brain neoplasms, in infants and children; Meninges, neoplasms; Meninges, magnetic resonance

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Although meningiomas account for approximately 15% of brain tumors in adults, they are uncommon in children, comprising 1%–3% of pediatric brain tumors (1–5). Previously described characteristics of childhood meningiomas include equal sex incidence as well as propensity for location in unusual sites such as the lateral ventricles, posterior fossa, and within brain parenchyma (6). Although recent authors have reviewed the clinical and histologic findings in childhood meningiomas (1–6), there is a paucity of information concerning magnetic resonance (MR) findings. We wish to report the MR and clinical findings in five children with meningiomas.

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Patients and Methods

Five children ranging in age from 3 months to 16 years had surgical and histopathologic diagnosis of meningioma at our pediatric hospital between 1989 and 1991 (Table 1). All patients had preoperative contrast-enhanced MR and two had contrast computed tomography. MR scans were performed on a 1.5-T scanner. T1-, proton density-, and T2-weighted scans were performed before the administration of intravenous Gd-DTPA (.2 mL/kg) in four patients and T1-weighted scans were performed after contrast injection in all five. Case 4, with previous resection of cervical spinal cord tumor, did not have precontrast T2-weighted scans of the head because the examination was performed after injection of intravenous contrast for examination of the cervical spine. Histopathologic analysis was performed on surgically excised tissue in all five children and chromosomal analysis was performed in four.

Results

Clinical, histopathologic, and radiographic data are summarized in Table 1. One 16-year-old boy had type 2 neurofibromatosis and previous surgical treatment of a cervical cord ependymoma. A 15-year-old girl had growth hormone deficiency as well as resection of a Wilms tumor at age 2, with subsequent chemotherapy and radiation

TABLE 1: Meningiomas in children

Case No.	Age/Sex	Neurologic Symptoms	Location	MR Findings[a]	Pathologic Type	Chromosomal Analysis
1	3 mo/M	None	Sphenoid wing	Hyperintense T1, T2 PD, + enhancement	Meningotheliomatous meningioma	Not performed
2	10 yr/M	Headache, hearing loss, hemianopsia	Tentorium	Isointense T1, hyperintense PD, T2, + enhancement; large vessels in tumor	Hemangiopericytic variant of angioblastic meningioma	Normal male
3	15 yr/F	Headache, dizziness, diplopia	Tentorium	Hypointense T1, hyperintense PD, T2, + enhancement	Chordoid meningioma with microcystic and myxomatous components	Chromosome 22 deletion
4	16 yr/M	None related to meningioma	Parasagittal	No precontrast scans; + enhancement	Transitional meningioma	Multiple complex abnormalities
5	16 yr/F	Quadriparesis	Foramen magnum	Isointense T1, hyperintense PD, T2, + enhancement	Meningotheliomatous meningioma	Normal female

[a] PD, proton density.

therapy to the abdomen. A 3-month-old infant had sagittal synostosis as well as multiple cutaneous and hepatic hemangiomas. The other two children had no other preexistent medical problems prior to onset of neurologic symptoms and MR and neurosurgical diagnosis of meningioma. All patients were treated by primary surgical resection of the tumor. Gross residual tumor was present after surgery in three patients, a 16-year-old boy whose tumor invaded and partially obstructed the superior sagittal sinus, a 16-year-old girl with a large lesion at the foramen magnum encasing the left vertebral artery, extending into the left posterior fossa, and in a 3-month-old infant with a sphenoid wing lesion. The foramen magnum lesion has subsequently been treated with radiation therapy and is stable on follow-up MR. The tumor involving the superior sagittal sinus is stable without further therapy. The other three patients show no residual or recurrent tumor on postsurgical MR.

Discussion

In contrast to adults in whom meningioma is the most common benign brain tumor, in children meningiomas are rare. Only 1% of 1283 intracranial tumors seen at the Hospital for Sick Children in Toronto between 1934 and 1985 were meningiomas (1). Several authors have reported differences between adults and children with meningiomas. In adults, meningiomas occur two times more commonly in women than in men, whereas, in children, an equal sex incidence has been reported (4). In our series, there were three boys and two girls.

Some authors have reported that children have an increased incidence of posterior fossa meningiomas, intraventricular lesions, and tumors without dural attachment (5, 6). In our patients, all five tumors were extraaxial and three of five originated in the posterior fossa. None of the tumors were intraventricular or intraparenchymal.

An increased incidence of malignant meningiomas in children has been reported (7). One series found a very high (30%) incidence of meningeal sarcomas in children (3), and, in the Mayo Clinic series, there was an increased incidence (10%) of papillary tumors that correlated with a very poor (40%) 5-year survival (4). Besides hemangiopericytic meningioma, which is considered malignant, classical meningiomas may also show malignant changes. Occasionally the malignant meningiomas show sarcomatous changes, particularly when they recur. These sarcomatous meningiomas retain some morphologic attributes of meningioma. There is another group of malignant tumors of the dura, meningeal sarcomas, that more commonly occur in infants and children. Most of these sarcomas are fibrosarcomas of the dura, and morphologically akin to soft-tissue fibrosarcomas. Many of them arise postirradiation. These fibrosarcomas are considered a distinct group from malignant meningiomas (7).

Two of the children in this series had unusual concommitant conditions. An infant initially underwent neuroimaging because of sagittal synostosis and because of multiple cutaneous and hepatic hemangiomas (Fig. 1). A 15-year-old girl with a chordoid meningioma had previously undergone resection of a Wilms tumor with subsequent abdominal irradiation (Fig. 2). This patient is also undergoing treatment for growth hormone deficiency.

The association of meningiomas with neurofibromatosis is well known (8, 9). About 25% of meningiomas in children are associated with neu-

rofibromatosis, especially type 2, as in one of our patients (Fig. 3). Cranial irradiation has also been associated with an increased incidence of meningioma (10).

Chromosomal abnormalities, particularly deletions involving chromosome 22, have been described in meningiomas. It has been hypothesized that loss of tumor suppressor genes may be a causative factor in the development of brain tumors and that tumors may be treated in the future by replacement of defective genes (10). Two of four tumors in our patients showed abnormalities of chromosome 22. In addition, two patients had normal chromosomal analysis of tumor tissue.

Initial reports of MR findings in meningiomas indicated that many of these tumors were isointense to brain on T1- and T2-weighted images, especially at low or mid field strength (11). More recent papers have shown that most meningiomas are iso- to hyopintense on T1-weighted scans and are frequently hyperintense to brain on T2-weighted images (12, 13). With the addition of contrast-enhanced MR, even small asymptomatic lesions can be detected. Elster et al, correlated MR signal changes with histopathology and found that low signal changes on T2-weighted images correlated with fibrous or transitional tumors presumably due to the presence of fibrous tissue and psammoma bodies and that high signal on T2-weighted images were seen in syncytial and angioblastic lesions perhaps due to microcystic changes, dilated blood vessels, and high cellularity seen in these lesions (13). However, Spagnoli et al, found no correlation between signal changes on MR and histopathology (12). All of the lesions in our patients were easily identified on MR and all enhanced markedly after the administration of intravenous contrast material. All four tumors examined with precontrast proton density- and T2-weighted scans showed hyperintensity compared to adjacent brain, although the hyperintensity was somewhat less than that usually seen with glial tumors. The sphenoid wing tumor in the infant was focally hyperintense on T1-weighted scans and showed some magnetic susceptibility effect on gradient-echo scans, perhaps due to some internal paramagnetic substance

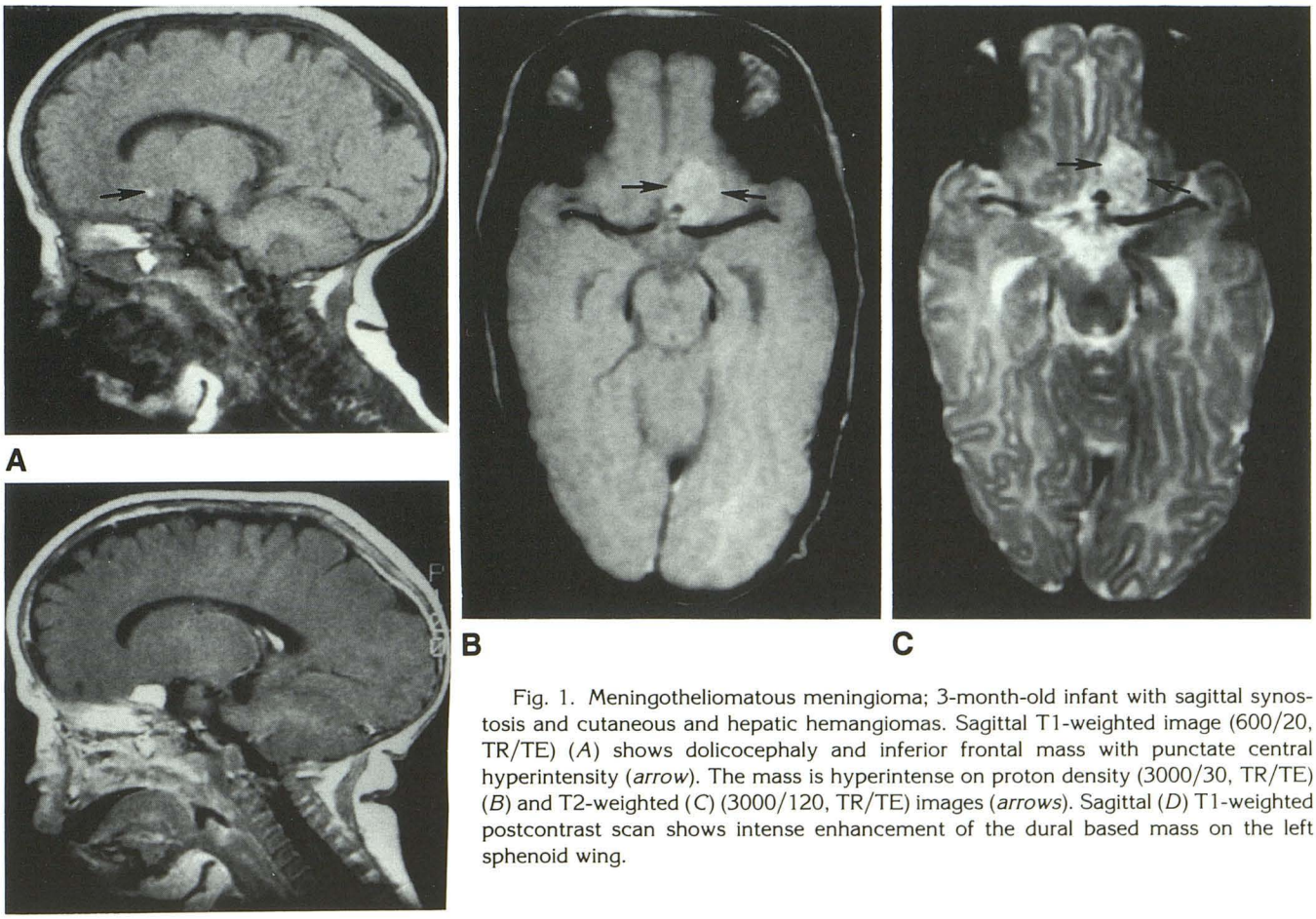

Fig. 1. Meningotheliomatous meningioma; 3-month-old infant with sagittal synostosis and cutaneous and hepatic hemangiomas. Sagittal T1-weighted image (600/20, TR/TE) (*A*) shows dolicocephaly and inferior frontal mass with punctate central hyperintensity (*arrow*). The mass is hyperintense on proton density (3000/30, TR/TE) (*B*) and T2-weighted (*C*) (3000/120, TR/TE) images (*arrows*). Sagittal (*D*) T1-weighted postcontrast scan shows intense enhancement of the dural based mass on the left sphenoid wing.

such as calcium or melanin. The child with the hemangiopericytic variant of angioblastic meningioma showed large flow voids throughout the tumor and a highly vascular tumor was found at surgery.

Histopathologic analysis of the tumors in our patients showed two meningotheliomatous and one transitional meningioma, tumors more frequently found in adults. The tumor in the 3-month-old infant was a typical meningotheliomatous lesion. Only one aggressive lesion (hemangiopericytic variant of angioblastic meningioma) was seen. This tumor is a controversial entity as its true cell of origin is not known. Some authors prefer to exclude this tumor from the meningioma group altogether and consider it as a vascular tumor (5); however, most authors are of the opinion that it be considered a variant of meningioma. One unusual tumor, a chordoid meningioma, was present. This variant of meningioma, which morphologically appears similar to chordoma, is associated with lymphohistiocytic infiltrates and is sometimes associated with Castleman syndrome (14). No evidence of Castleman syndrome was present in our patient.

Meningiomas in children are less common than in adults and are often located in the posterior fossa (4–6). As in our patients, unusual histopathologic findings are often present (3, 4). All five tumors in our series were extraaxial in location and showed intense contrast enhancement. In contrast to previous reports in adult patients (11), meningiomas in our patients were hyperintense to adjacent brain on proton density- and T2-

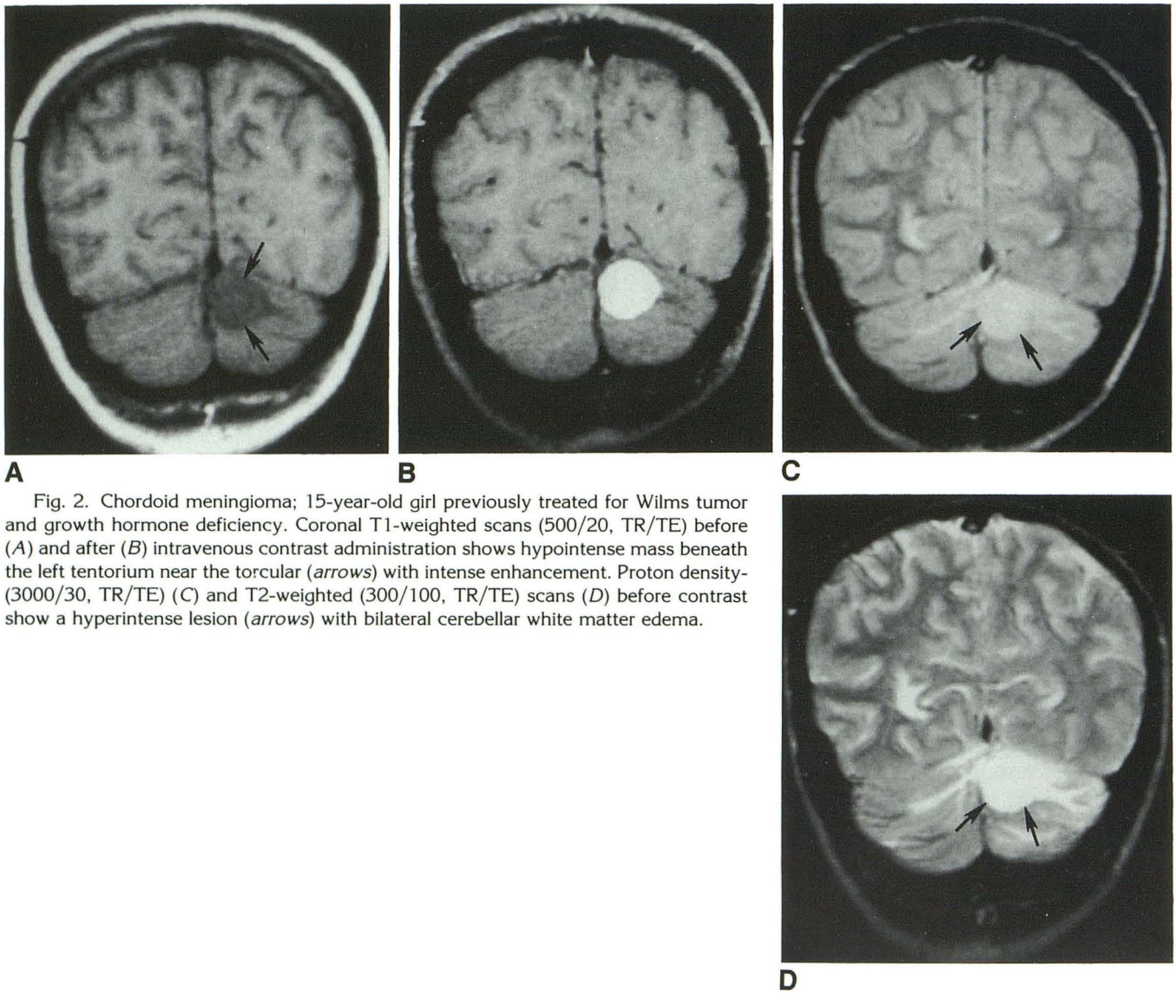

Fig. 2. Chordoid meningioma; 15-year-old girl previously treated for Wilms tumor and growth hormone deficiency. Coronal T1-weighted scans (500/20, TR/TE) before (*A*) and after (*B*) intravenous contrast administration shows hypointense mass beneath the left tentorium near the torcular (*arrows*) with intense enhancement. Proton density- (3000/30, TR/TE) (*C*) and T2-weighted (300/100, TR/TE) scans (*D*) before contrast show a hyperintense lesion (*arrows*) with bilateral cerebellar white matter edema.

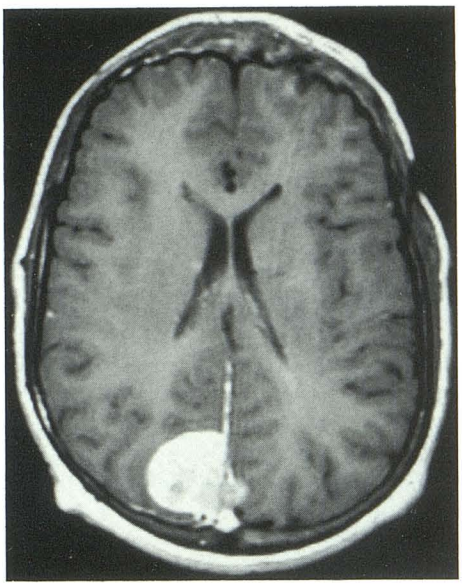

Fig. 3. Transitional meningioma; 16-year-old boy with Type 2 neurofibromatosis. Contrast-enhanced axial T1-weighted (600/20, TR, TE) MR shows bilobed, enhancing dural-based mass extending on both sides of the superior sagittal sinus posteriorly.

weighted images, generally allowing visualization of the lesion before the administration of intravenous contrast.

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