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AJNR Am J Neuroradiol 1992, 13 (5) 1353-1364
http://www.ajnr.org/content/13/5/1353

Primary Meningeal Tumors in Children: Correlation of Clinical and CT Findings with Histologic Type and Prognosis

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PURPOSE: To identify the radiologic features that might help in preoperative differentiation of the meningiomas from the remaining primary meningeal tumors, in particular the malignant tumors. **METHODS:** The clinical and computed tomographic features of 21 children with histologically proved primary meningeal tumors were analyzed. **FINDINGS:** Benign tumors (meningiomas) are more likely to occur in older children, to have longer symptom duration, and to have CT appearances similar to the "typical" adult meningioma. Atypical CT features suggest a malignant meningeal tumor, such as meningeal sarcoma, melanoma, or meningeal primitive neuroectodermal tumor. The recent identification of a new subtype of meningioma (a "sclerosing" group) is discussed. This is common in children and the CT and clinical features are similar to those seen in other meningiomas. It is frequently mistaken histologically for an intraaxial tumor, or for an atypical or malignant meningioma. These sclerosing meningiomas may also show brain invasion but despite this, in the short term, the prognosis is no different from other meningiomas. **CONCLUSION:** The bad reputation previously ascribed to childhood primary meningeal tumors should be confined to that small group that are malignant. Meningiomas have a more favorable outlook.

Index terms: Meninges, neoplasms; Computed tomography, in infants and children; Pediatric neuroradiology

AJNR 13:1353–1364, Sep/Oct 1992

Primary meningeal tumors are uncommon in children, representing 1.4%–3% of intracranial tumors (1–6). They have a reputation as large, rapidly growing tumors with a tendency to become malignant and with poor outcome (2, 3, 7). In part, this may be due to the inclusion of meningeal sarcoma with meningioma under the general title of meningioma of childhood (3, 5, 6). However, by considering meningioma as an entity, separate from the remaining primary meningeal tumors, Drake et al (8) and Herz et al (9) have noted that the prognosis for this specific tumor may be more optimistic than previously reported.

Thus, it becomes important to differentiate radiologically the meningiomas as a group from the remaining primary meningeal tumors, particularly the malignant tumors. Review of the literature on childhood meningeal tumors (1–14) disclosed a paucity of information on computed tomography (CT) findings (6, 8, 9, 11, 13, 14). Accordingly, we decided to review the computed tomographic and clinical findings in our primary meningeal tumors.

Subjects and Methods

A review of the radiologic and neuropathologic records between 1976 and 1988 revealed 21 children less than 17 years of age with a histologically proven diagnosis of primary meningeal tumor, in whom CT had been performed preoperatively. Two other patients with neurofibromatosis and with a clinical and radiologic diagnosis of meningioma lacked histologic proof and were excluded. One child, case 13 (Table 1), had been treated with surgery and radiotherapy 13 years previously for a medulloblastoma.

Received August 2, 1991; accepted and revision requested August 19; revision received December 9.

Presented in part at the 26th Annual Meeting of the American Society of Neuroradiology, Chicago, May 14–19, 1988.

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AJNR 13:1353–1364, Sep/Oct 1992 0195-6108/92/1305-1353

Microscopic sections were stained with standard histologic techniques (hematoxylineosin, reticulin, Masson Trichrome) and with the immunoperoxidase technique for glial fibrillary acidic protein (GFAP) and vimentin. Where suitable fixed tissue was available, electron microscopic examination was also performed. Cases of sarcoma, melanoma, and meningeal primitive neuroectodermal tumor (PNET) were diagnosed on histologic and electron microscope grounds only. Meningiomas were further assessed for histologic pattern, cellularity, mitotic rate, pleomorphism, amount of necrosis, and brain invasion.

Eighteen patients had both unenhanced and enhanced CT scans. Two had enhanced scans and one an unenhanced scan only. The presence or absence on CT of discrete calcification, hemorrhage, hyperostosis, bone destruction (with or without extracranial extension), intra- or peritumoral cyst formation, and hydrocephalus was noted. Tumor density on unenhanced scans was graded as hypo-, iso-, or hyperdense relative to gray matter, and, following contrast injection, tumor density was noted as homogenous or heterogenous, and tumor margins as 1) well defined and smooth or lobulated, or 2) irregular and fringe-like. Edema was graded as absent, moderate, or severe, as established by Vassilouthis et al (15).

Results

Incidence and Histology

Histologic diagnoses are shown in Table 1. Twenty-one patients with primary meningeal tumors seen since 1976 represented 3% of 690 histologically proven primary intracranial tumours. Fifteen patients' tumors (71.5%) were benign, all meningiomas. The six malignant tumors (28.5%) included three meningeal sarcomas, two malignant melanomas, and one PNET (arising apparently within meninges and without evidence of intraaxial tumor).

Of the 15 meningiomas, four were of the meningothelial type and four were transitional. Seven tumors fell into a recently described (16) histologic subtype designated as "sclerosing meningioma" in Table 1. These "sclerosing" lesions all had a very similar histologic appearance, with the bulk of the lesions consisting of collagen bundles with a very small population of spindly cells (Fig. 1). Some of the more cellular areas showed perinuclear artifactual halos, giving a "fried egg" appearance, and the cytoplasm lay in delicate wisps, resembling the processes of astrocytes (Fig. 2); however immunoperoxidase stains were positive for vimentin and negative for GFAP in these small strands of cytoplasm. The diagnosis of meningioma was made possible by finding whorls of tumor cells, wrapping around each other and around blood vessels, and it was confirmed by finding intracellular vimentin and an absence of GFAP. Gradual fibrosis and cell loss in these whorls led to the formation of an almost acellular mass in which the whorling pattern of the collagen remained as an indication of its meningiomatous origin (Fig. 3).

Tumor invasion of the brain was seen in four of these seven sclerosing lesions and in none of the other meningiomas.

Clinical Findings

Mean age at presentation was 9 years 4 months (range, 4 months–16 years 8 months) for all the tumors; 11 years 4 months (4 months–16 years 8 months), for the meningiomas, and 4 years 5 months (11 months–9 years 4 months) for the malignant tumors. There was little difference in patient age between those with sclerosing (10 years 4 months) and those with the remaining meningiomas (12 years 3 months). Figure 4 shows the distribution in three age groupings of all meningeal tumors by histology. Overall, there were 14 boys and 7 girls. Meningiomas showed a male predominance of 10:5.

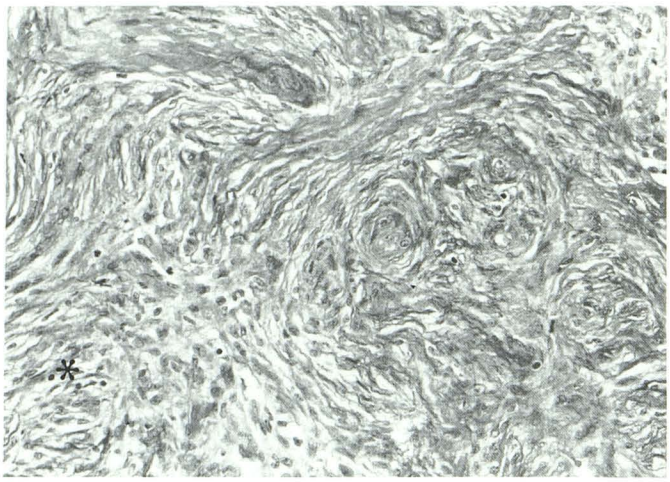
The predominant symptom complex and its duration is listed in Table 1. The benign tumors, all meningiomas, presented with seizures in five children, visual disturbances in four, headache in three, and in one case each, a palpable mass, ataxia, and raised intracranial pressure (details were incomplete in one case). Children with benign meningiomas were symptomatic for longer periods, average 19.6 months (range, 1–72 months). Of the six children with malignant tumors, symptoms of raised intracranial pressure were predominant in five and symptom duration averaged 0.6 months (range, 12 hours–3 months). Five of these six children were symptomatic for less than 2 weeks, and two cases, both melanomas, presented in coma, one on the basis of tumor associated hemorrhage (Fig. 5) and the other, secondary to acute hydrocephalus (Fig. 6). In contrast, nine of 15 children with meningiomas were symptomatic for 6 or more months.

Prognosis was better in those with benign tumors in whom 14/15 were alive at follow-ups ranging from 1 week to 7 years (average 34 months). In contrast, three children with malignant tumors were dead (at 10 days, 3 months, and 9 months) and one showed massive regrowth of tumor 2 months after surgery, despite adjuvant chemotherapy. Two remaining children with ma-

TABLE 1: Findings in 21 children with primary meningeal tumor

Case	Age	Sex	Tumor Size (3 largest dimensions in cm)	Location	Original Histology	Final Histology	Predominant Symptom	Symptom Duration	Follow-up	Comment
1	14	F	4 × 3 × 3	Cerebellopontine angle	Astrocytoma	*Sclerosing meningioma*	Not available	Not available	Alive 3 yr	Post-op radiotherapy
2	16.66	F	4 × 4 × 3	Frontal convexity	Malignant meningioma (frozen section astrocytoma)	*Sclerosing meningioma*	Seizure	4 yr	Alive 4 yr	Brain invasion
3	5.83	M	4 × 4 × 3	Middle cranial fossa	Atypical meningioma	*Sclerosing meningioma*	Seizure	1 yr	1 yr**	Thoracic astrocytoma
4	11.5	M	2 × 2 × 1	Middle cranial fossa	Transitional meningioma	*Sclerosing meningioma*	Seizure	6 yr	Alive 1 wk	Brain invasion
5	0.33	M	3 × 2 × 2	Frontoparietal convexity	Meningeal sarcoma	*Sclerosing meningioma*	Palpable mass	4 mo	Alive 4 yr	Brain invasion; post-op radiotherapy
6	10.58	M	9 × 8 × 8	Anterior cranial fossa	Atypical meningioma	*Sclerosing meningioma*	Seizure	4 yr	Alive 7 yr	Brain invasion
7	13.25	F	2.5 × 2.5 × 2	L lateral ventricle	*Sclerosing meningioma*	*Sclerosing meningioma*	Headache	6 mo	Alive 1 wk	
8	2.5	M	6 × 6 × 5	Sylvian fissure	Transitional meningioma	Same	↑ICP	1 mo	Alive 3 yr	
9	14.75	F	4 × 3 × 4	Parasagittal	Transitional meningioma	Same	Seizure	1 mo	Alive 10 mo	
10	16.17	M	5 × 4 × 4	Inferior tentorium	Meningothelial meningioma	Same	Visual disturbances/ataxia	4 mo	Alive 2 yr	
11	16.33	M	6 × 6 × 5	Lateral ventricle	Transitional meningioma	Same	Headache	2 yr	Alive 3 yr	
12	11.33	M	4 × 3 × 2	Sphenoid wing	Meningothelial meningioma	Same	Visual disturbances	Not available	Alive 2 wk	
13	15.33	M	8 × 7 × 4	Frontoparietal convexity	Transitional meningioma	Same	Headache	6 mo	Alive 4 yr	Radiotherapy for medulloblastoma 13 yr before
14	10.66	F	#, 0.2 × 0.2 × 0.2, and 2 × 0.5 × 0.5	Optic canal (R); optic canal and orbit (L)	Meningothelial meningioma	Same	Visual disturbances	1 yr	Alive 7 yr	
15	10.75	M	#, 0.2 × 0.2 × 0.2 (L & R)	Optic canal (L & R)	Meningothelial meningioma	Same	Visual disturbances	3 yr	Alive 2 yr	
16	0.92	M	4 × 4 × 3	Cerebellopontine angle	Meningeal sarcoma	Same	↑ICP	24 hr	10 days†	
17	4	F	5 × 8 × 5	Parieto-occipital convexity	Meningeal sarcoma	Same	Palpable mass	3 mo	9 mo†	
18	1.42	M	7 × 8 × 6	Temporoparietal convexity	Meningeal sarcoma	Same	↑ICP	2 wk	Alive 2 mo	Deteriorating clinically
19	3.25	M	Diffuse	Posterior cranial fossa	Malignant melanoma	Same	↑ICP (coma)	24 hr	3 mo†	
20	9.33	M	6 × 5 × 3	Parietal convexity	Malignant melanoma	Same	↑ICP (coma)	12 hr	Alive 4 mo	
21	7.75	F	Diffuse	Sylvian, basal cisterns	PNET; meninges	Same	↑ICP	1 wk	Alive 14 mo	

Note.— #, Bilateral optic nerve meningiomas; **, presumed dead after 1 year—massive regrowth tumor without further follow up after 1 year; †, died; ↑ICP, raised intracranial pressure; L, left; R, right.



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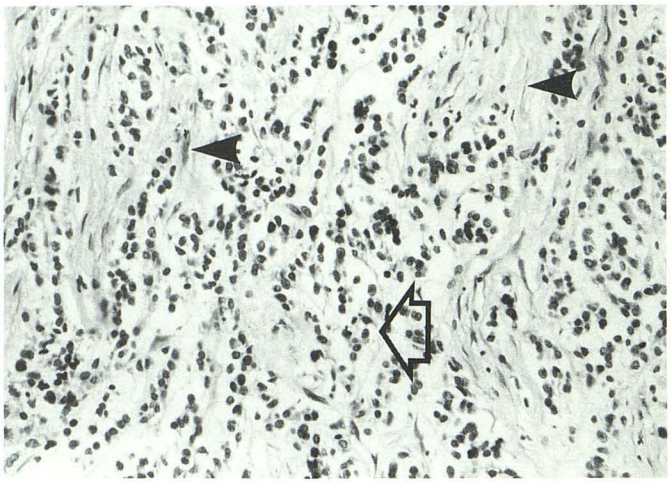

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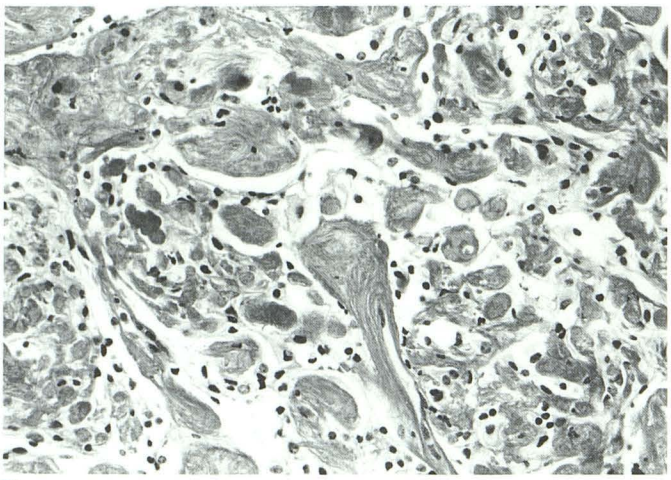

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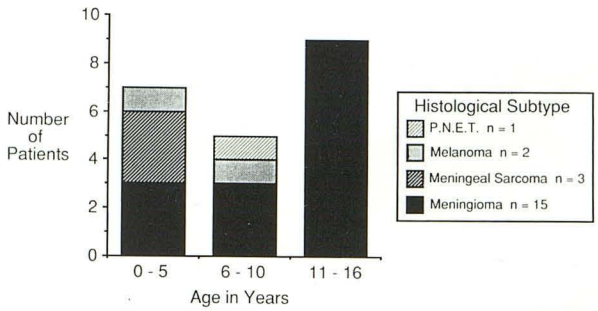

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Fig. 1. Most of the "sclerosing" meningioma consists of almost acellular collagen. At bottom left (*asterisk*) is a more cellular part of the lesion; (Hemotoxylineosin × 495.) (Histology is from case 1. See also Fig. 9.)

Fig. 2. In the more cellular parts of the lesion, a perinuclear halo produces a "fried egg" appearance (*open arrow*). Delicate wisps of cytoplasm mimic astrocyte cell processes (*arrowheads*). Hemotoxylineosin × 235). (Histology is from case 1. See also Fig. 9.)

Fig. 3. In the collagenous and largely acellular part of the lesion, the whorling pattern is evident. (Hemotoxylineosin × 495). (Histology is from case 1. See also Fig. 9.)

Fig. 4. Age distribution of 21 primary meningeal tumors by histologic subtype.

lignancies were alive at 4 and 14 months.

Symptom duration in the group with sclerosing meningioma was generally longer (average 31.5 months) than for the other group with meningiomas (average 15.5 months) and seizures were the predominant symptom in four of the patients with sclerosing meningioma.

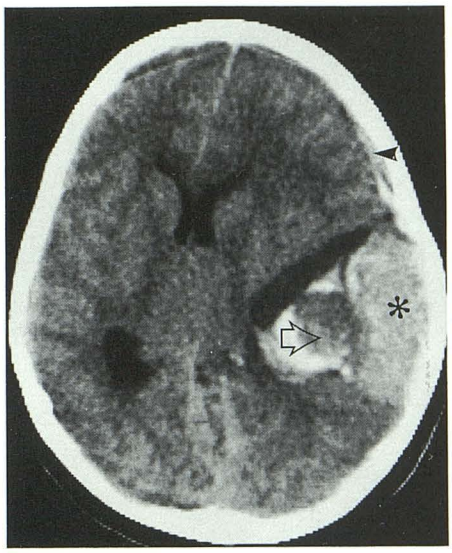
Radiology

Morphologically, the 21 patients presented with 17 intracranial mass lesions (Table 2), two diffuse lesions (Table 3), and two cases of bilateral optic sheath meningioma, for a total of 23 tumors in 21 patients. CT detected all of the mass and diffuse lesions but only one of the four optic sheath tumors, for an overall sensitivity of 83%.

Mass Lesions (n = 17).

1. *Benign (n = 13):*

The CT features of the benign lesions, all meningiomas, are shown in Table 2, and the typical appearance (Fig. 7) was of a hyperdense or isodense mass with variable degrees of calcification and hyperostosis. Following contrast injection, enhancement was homogenous or heterogenous with well-defined margins and variable edema. Two tumors arose in the posterior fossa and three had no dural attachment (two in the



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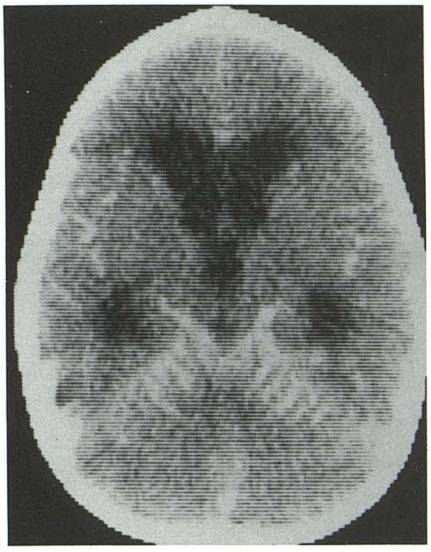

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Fig. 5. Unenhanced axial CT. Malignant melanoma (case 20), showing hyperdense mass (*asterisk*) with proven diffuse components spreading anteriorly (*arrowhead*) and with underlying focus of partially clotted hemorrhage (*open arrow*) responsible for child's acute presentation (nonenhanced scan only done at time of presentation).

Fig. 6. Axial enhanced CT; diffuse malignant melanoma (case 19). Diffuse enhancement is noted over the cerebellar hemispheres. Hydrocephalus is evident.

lateral ventricle and one in the sylvian fissure (Fig. 8)). Neither cyst formation nor hemorrhage was seen in the meningiomas.

The sclerosing meningiomas showed certain features when compared to the transitional or meningotheleal types. Four of the five meningiomas showing calcification were of the "sclerosing" type, in addition, within the meningiomas the only examples of heterogenous enhancement (Fig. 9) and bone lysis (Fig. 10) occurred within this sclerosing group (Table 4).

TABLE 2: CT findings in primary meningeal tumor: presenting as mass lesions

Unenhanced CT		Meningioma n = 11	Meningeal Sarcoma n = 3	Meningeal Melanoma n = 1
Mass:	hyperdense	9	1	1
	isodense	2		
	heterogenous		2	
Calcification		5	1	
Hyperostosis		1		
Bone destruction		1	2	Remodelled bone
Extracranial extension		1	1	
Hemorrhage				1
Enhanced CT		n = 13	n = 3	n = 0
Enhancement:	homogenous	12		
	heterogenous	1	3	
Oedema:	absent	5	1	
	moderate	2	2	
	severe	6		
Margins:	well defined	13	1	
	irregular or fringed		2	
Hydrocephalus		3	2	
Cyst			2	
Total numbers		13	3	1

TABLE 3: Histology and CT findings in primary meningeal tumors presenting as diffuse lesions

Case	Histology	Hydrocephalus	Meningeal Enhancement
19	Melanoma	Panventricular	Superior cerebellar cisterns
21	PNET	Third and lateral ventricles	Right sylvian fissure and basal cisterns

2. *Malignant (n = 4):*

Three meningeal sarcomas and one malignant melanoma presented as mass lesions. CT in those tumors showed a number of "atypical" features (Table 4) that were either seen rarely, or not at all, in our meningiomas and included: hemorrhage—one (Fig. 5); bone lysis—two (Fig. 11); heterogenous enhancement—three (Figs. 11 and 12); and cyst formation—two (Fig. 12); as well as poorly defined margins—two (Fig. 11).

Diffuse Tumors (n = 2). Both diffuse tumors (Table 3) were malignant. Both presented with meningeal enhancement and hydrocephalus (Figs. 6 and 13); one was a malignant melanoma, the other, a PNET.

Optic Sheath Tumors (n = 2). Both patients presented with progressive visual loss, and surgical exploration disclosed small bilateral optic sheath meningiomas within the optic canals of both patients. Only one of those four tumors

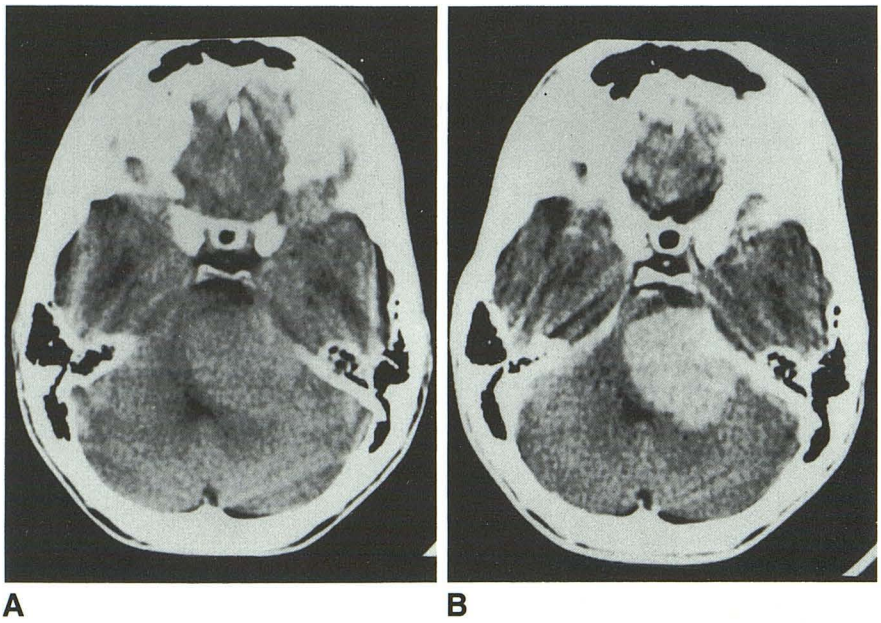

Fig. 7. Unenhanced (*A*) and enhanced (*B*) axial CT; meningiothelial meningioma (case 10). Unenhanced CT shows a hyperdense mass arising from inferior surface tentorium (*A*) that following contrast injection (*B*) shows homogenous enhancement with well-defined slightly lobulated margins.

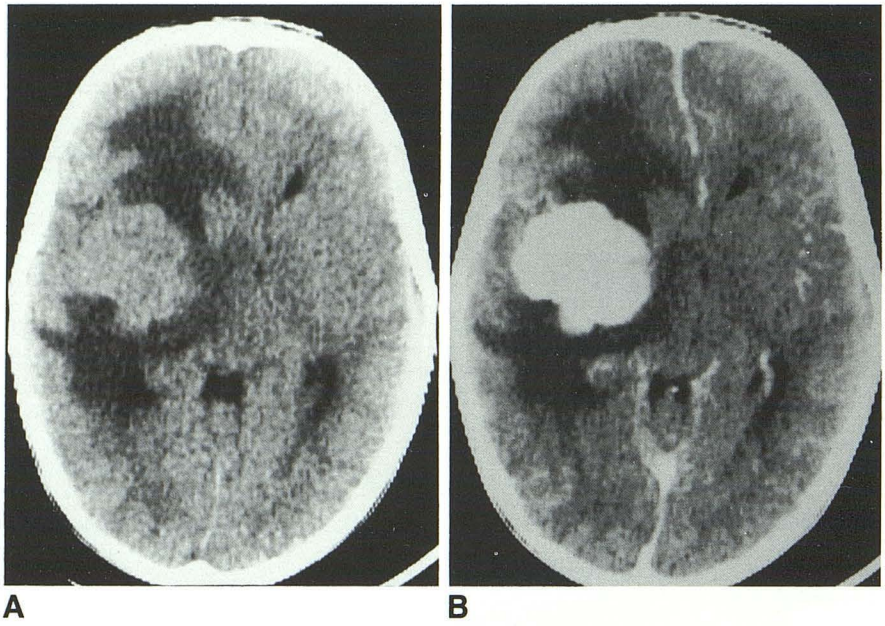

Fig. 8. Unenhanced (*A*) and enhanced (*B*) axial CT. transitional meningioma (case 8) arising in right sylvian fissure. Preoperative diagnosis was of an intraaxial tumor.

were seen preoperatively by virtue of its extension along the orbital portion of the optic nerve (case 14). These tumors do not appear in Tables 2, 3, or 4.

Discussion

Clinical Findings

The low incidence of primary meningeal tumors in our institution (3% of intracranial neoplasms) is in keeping with previously recorded incidences of 1.4%–3.2% of all intracranial tumors in children (1–6). Contrasted with adults in which meningiomas constitute the largest single grouping within primary meningeal tumors and which are predominantly benign histologically, our childhood primary meningeal tumors were more frequently malignant (28.5%) and showed three major groupings: meningioma (71.5%), sarcoma (14.25%), and miscellaneous (14.25%). This ratio of benign:malignant is generally reflected by other authors (1–3, 5, 6) who, however, note a higher incidence of meningeal sarcoma (22%–50%). In addition, sarcomatous degeneration, a feature not seen in our meningiomas, is said to occur in 14% of meningiomas (9). These

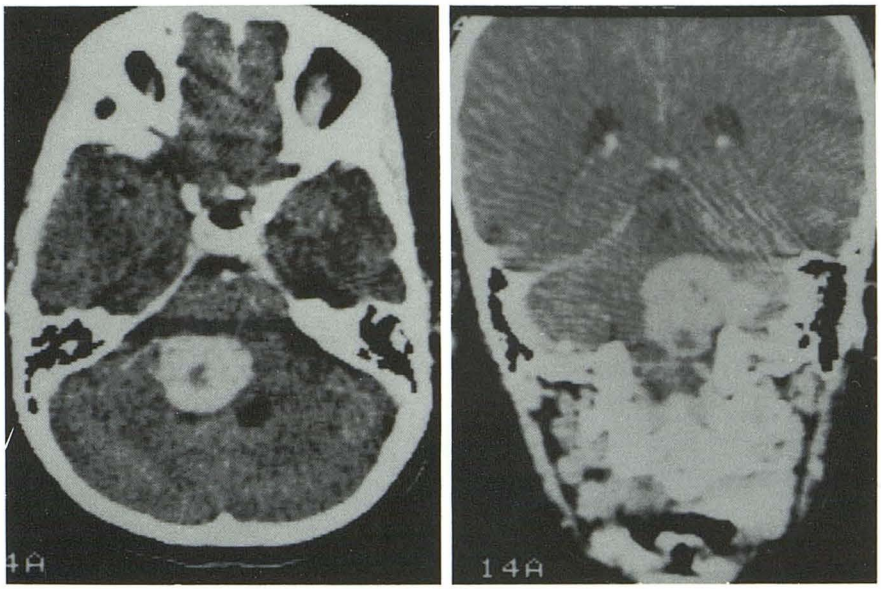

Fig. 9. Enhanced axial (*A*) and coronal (*B*) CT. "Sclerosing meningioma" (case 1) arising from jugular tubercle and adjacent petrous bone and showing heterogenous foci of enhancement. The original histology in this case (astrocytoma) is illustrated in part in Figures 1–3. Figure 2 shows some of those features that led to its original mistaken diagnosis of astrocytoma, and Figure 3 shows some of the features that allowed the diagnosis to be revised to sclerosing meningioma.

two discrepancies may be due in part to our reinterpretation of three meningiomas (cases 2, 4, and 6) with brain invasion as sclerosing meningiomas. Ordinarily, they might have been called meningioma with sarcomatous change. In addition, one tumor originally diagnosed as meningeal sarcoma (case 5) was reinterpreted as a sclerosing meningioma.

The average age of presentation of 11 years 4 months for the meningiomas is almost twice the average of other intracranial tumors noted by Yates et al (10), who gave an average age of 6.3 years. This presentation by meningioma in older children is noted by other authors (5, 6, 11, 12) who also demonstrated an increasing incidence with increasing age (5, 6, 12), a feature reflected in our statistics and illustrated in graphic form in Figure 4. It appears reasonable to assume that the increased numbers of meningiomas noted with increasing age in children simply reflects the lower edge of a curve that begins in children and peaks in adults at the fifth to seventh decade (17, 18). In contrast, our six malignant tumors presented at average age 4 years 5 months, with the three sarcomas all being less than 4 years of age.

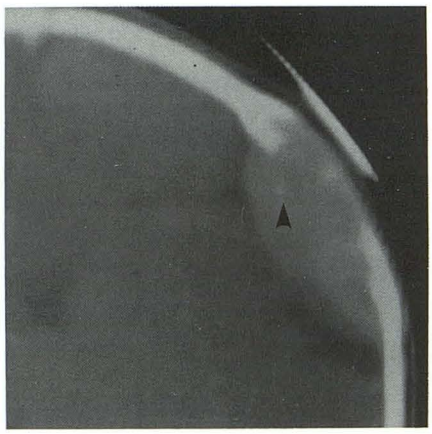

Fig. 10. Enhanced axial CT. "Sclerosing" meningioma (case 5) at "wide" CT window showing extensive bone destruction and large extraaxial mass, as well as a skull contour defect that resulted in presentation as a palpable mass (no evidence of the brain invasion, noted histologically, was seen on this or other sections, at routine window settings). Faint calcification (*arrowhead*) is seen. Original histologic diagnosis was meningeal sarcoma.

The well-known female predominance in meningiomas where M:F rations range between 1.6 and 2.4:1 (15, 18) applies primarily to middle-aged adults (19) and is not reflected in childhood cases that show approximate equivalence between the sexes or a slight male bias (1–2, 7, 9, 11, 12). In addition, the sexes are approximately equally represented where meningiomas are incidentally discovered at autopsy in the very old (19). These facts and the increased clinical manifestations of meningioma in pregnancy, as well as the statistically significant occurrence of meningioma and breast carcinoma in the same patient, suggests that sex hormone receptors may be present in these tumors (19). Kepes (19) has reviewed the data on the presence of estrogen and progesterone receptors in meningioma and suggests that the equivalence between the sexes in childhood meningioma and the female predominance in adults may be a reflection of the fact

TABLE 4: Atypical CT findings correlated with histology

Case	Histology	Hemorrhage	Bone Lysis	Poorly Defined or Fringed Margins	Heterogenous Enhancement	Cyst	Diffuse Tumors
1	Sclerosing meningioma	−	−	−	+	−	−
5	Sclerosing meningioma	−	+[a]	−	−	−	−
16	Sarcoma	−	−	−	+	+	−
17	Sarcoma	−	+[a]	+	+	−	−
18	Sarcoma	−	+	+	+	+	−
19	Melanoma	−	−	−	−	−	+
20	Melanoma	+	−	−	−	−	−
21	PNET	−	−	−	−	−	+

[a] Extracranial extension noted.

Note.—+, present; −, absent.

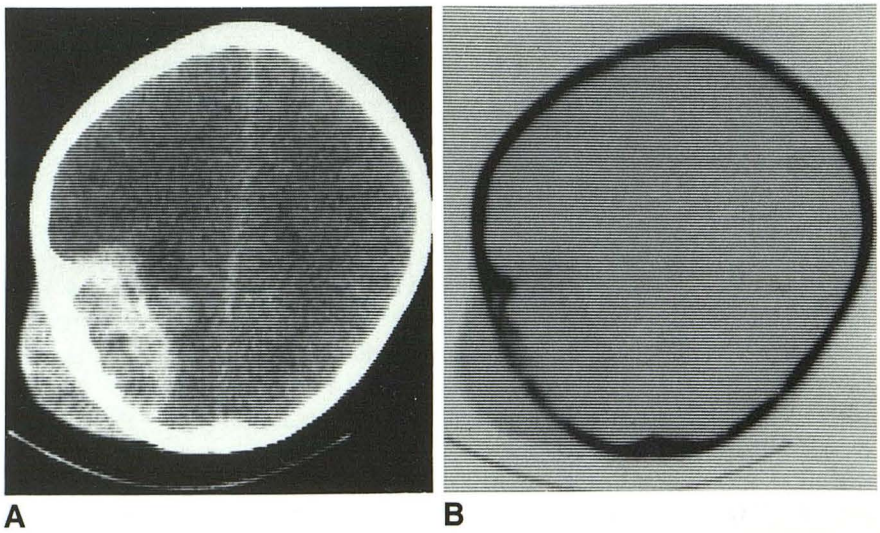

Fig. 11. Enhanced axial CT; meningeal sarcoma (case 17) at regular brain (*A*) and bone (*B*) windows. Heterogenous enhancement with poorly defined margins and bone lysis is seen (presentation was with a palpable mass).

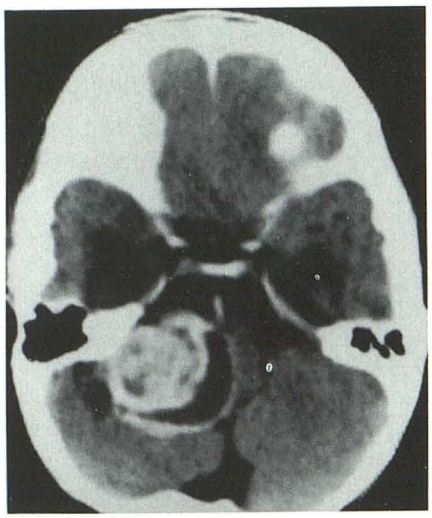

Fig. 12. Enhanced axial CT; meningeal sarcoma (case 16). Heterogenous enhancement of the solid component as well as mural enhancement of the "cystic" component is seen. Temporal horn dilation is seen bilaterally.

that estrogen stimulation has a slow and cumulative effect that does not ordinarily manifest as a female predominance until middle age.

Pediatric meningiomas reportedly occur more frequently in the posterior fossa, 19%–57% (3, 12), however, our incidence of 13.3% (two/15) of meningiomas in this location is closer to the adult figure of 10%–11% (20, 21). Two meningiomas arose in the lateral ventricle (13.3%) and one in the sylvian fissure (6.5%), supporting the concept that the ventricles, 17%–26% (9, 12), and other nondural sites, 12.5% (5), are favored locations.

The increased incidence of meningioma in patients with the central form of neurofibromatosis (NF-2) is well recognized (17) and in children, this association occurs in 19%–24% (9, 11, 12). None of our pathologically proven cases has NF-2, although case 3, 1 year previously, had a low-grade astrocytoma resected from his thoracic cord. If it were known that he had a first-degree

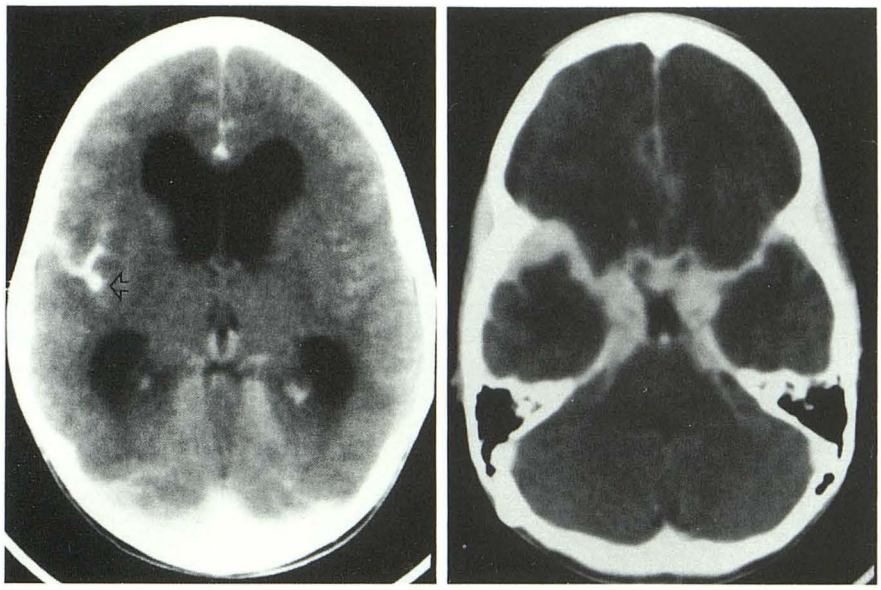

Fig. 13. *A*, Enhanced axial CT; PNET meninges (case 21). Hydrocephalus and meningeal enhancement in the right sylvian fissure (*open arrow*). No intraaxial tumor was seen on this or on any subsequent scan.

B, Same case following shunting of hydrocephalus, showing enhancement of meningeal tumor in basal cisterns.

relative with NF-2, he would fulfill the NIH diagnostic criteria for this disease (22). Unfortunately, the patient is lost to follow-up. Two patients in our care with neurofibromatosis and a radiologic diagnosis of meningioma are not included in this series since histologic proof of meningioma is not available. There are numerous reports of meningiomas following cranial irradiation by 5–25 years (19). Rubinstein et al (23) have reported that 80% of meningiomas seen following cranial irradiation for tinea capitis have a distinctive histologic appearance with high cellularity, extreme nuclear pleomorphism, frequent giant cells and nuclear pseudoinclusions, abundant mitoses, and prominent hyalinized blood vessels. It is not possible to be certain that the meningioma in our case 13 (treated with radiation for medulloblastoma 13 years previously) is in fact radiation-induced since it did not show these changes.

Radiology

CT of adult meningioma typically shows a hyperdense or isodense mass with varying amounts of calcification, hyperostosis, and edema, that following contrast injection shows homogenous enhancement with well-defined round or lobulated borders (18, 20, 24). Tumor detection rate is over 95% and a specific diagnosis is made in up to 90% of the cases (18, 20). Diagnosis is more difficult in certain locations (20), including intraventricular tumors, or where atypical features are present. When these atypical features are focal, a correct diagnosis can still be made in 93% of the cases (25). A misdiagnosis of malignancy based on atypical CT findings is made in 6%–7% of the cases (18, 25).

Eighty-five percent (11/13) of our meningiomas presenting as intracranial mass lesions (Table 2) showed a uniformity of appearance that conformed well to the "typical" adult appearance described above. In contrast, 100% (six/six) of the malignant tumors and only 15% (two/13) of the meningiomas showed "atypical" appearances (Table 4). Both atypical meningiomas were of the "sclerosing" subtype. Case 1 showed only focally heterogenous enhancement with otherwise typical appearances (Fig. 9), an appearance well documented by Russell et al (25) and ascribed to necrosis, scarring, cystic degeneration, or old low-density hemorrhage within meningioma. In contrast, all three sarcomas showed much more extensive heterogenous enhancement (Figs. 11 and 12), a finding that may be valuable in differentiating these from meningiomas in children.

The second atypical meningioma, case 5, showed bone lysis on CT (Fig. 10), a feature well described in adult meningioma (12, 20, 26) and which does not appear to correlate with clinical aggressiveness of the tumor (27). Thus, although this feature was seen in two of our three sarcomas, its frequency in meningioma, 4%–21% (20, 27) makes it of questionable value in differentiating meningioma from malignant meningeal tumors in children.

Microfoci of intratumoral hemorrhage (acute or chronic) are probably commoner than previously realized and appear to be responsible for a

proportion of the atypical CT appearances in meningioma (25), however, CT evidence of gross hemorrhage remains uncommon within meningioma so that an alternative diagnosis should be considered. The only case in which we saw hemorrhage was a malignant melanoma (Fig. 5).

Poorly defined or fringed margins on the cerebral surface of meningeal tumors was interpreted as suggestive of brain invasion and was seen only in our malignant tumors, in two of the three sarcomas. While this fringing pattern may be seen in meningioma, it is significantly more common in malignant or atypical meningiomas (27). Of interest, four sclerosing meningiomas, with documented brain invasion, did not show this sign (Fig. 10).

Large cystic areas were seen in two of the three sarcomas only and no meningiomas. This appearance, although uncommon, has been described in meningioma. Claveria et al (20) noted three major and three minor cysts in 71 tumors. Russell et al (25) noted that these "cysts" may represent adjacent edema, trapped cerebrospinal fluid spaces, focal brain atrophy, or true cystic degeneration. Rarely, a second adjacent pathology may coexist beside the meningioma (20). Beside cystic degeneration, cysts with neoplastic cells in the wall, and with enhancing rims on CT, may be seen in meningioma (28) and cystic change within meningioma may be a cause of mistaken diagnosis of malignancy (20, 25, 29). Bowen et al (28) reviewed the literature on cystic meningioma and noted that these appear to be commoner in pediatric meningioma, 11 of 39 cases occurring in those less than 17 years of age, with six of those under the age of 10 months. In pediatric primary meningeal tumors, Kolluri et al (6) noted cysts in four of 15 meningiomas and two of four meningeal sarcomas and Sano et al (5) noted cysts in 16.7% of his meningeal tumors, all sarcomas. Thus, while it appears that pediatric meningiomas may more commonly be cystic, a significant number of the cystic primary meningeal tumors in this age group are sarcomas (5, 6), and thus cystic change should be regarded with suspicion for malignancy.

Diffuse involvement of the meninges by primary meningeal tumors is uncommon. Of 50 cases of disseminated meningeal and ependymal malignancy described by Ascherl et al (30), no case is attributed to a primary meningeal tumor, and, in our review of 153 primary meningeal tumors cited by eight series (2, 3, 5–7, 9, 11, 12), only two diffuse tumors are noted. One, a meningioma (12), had a large parasellar tumor partially resected, and, at autopsy several weeks later, had widespread tumor in the posterior fossa and spinal meninges. The second case (3), a meningioma with sarcomatous change treated with surgery and radiotherapy, showed widespread intracranial metastases at autopsy 10 years later. Dissemination at or after the time of surgery is a possibility in these two cases. Rubinstein (17) notes that diffuse forms of meningeal sarcoma may be encountered, and that neurocutaneous melanosis as well as primary malignant melanoma, as in our case 19, may be diffuse lesions.

Both our diffuse lesions were malignant, and in view of this and the rarity of benign diffuse meningeal tumors in the literature, we believe that this finding should prompt suspicion of malignancy.

Sclerosing Meningioma

The "sclerosing" meningiomas that represented 47% (seven/15) of our meningiomas do not appear to be a biologically different group of tumors, but form a histologic subtype of meningioma that has been previously described (16). Age at presentation and sex distribution appear similar to the remaining meningiomas. Seizures are a common presentation in this small group and they were symptomatic for longer periods (averaging 31.6 months vs 15.5 months) than the remaining meningiomas. Calcification appears to be more common in this group on CT (57% vs 12.5%). The histologic findings that the bulk of these tumors consists of collagen bundles with a small population of spindly cells may help to explain the heterogenous enhancement seen in case 1 (Fig. 9).

The importance of recognizing this subtype histologically is apparent from studying the original diagnoses in Table 1 where, despite second opinion being sought from other centers, diagnoses including astrocytoma, meningeal sarcoma, and atypical or malignant meningioma were made in five patients, with two receiving radiotherapy. (In this respect, the radiologic findings of "typical" meningioma in five of seven cases may be helpful in suggesting the correct diagnosis.) One reason for the diagnostic difficulty was the belief that childhood meningiomas are likely to be malignant (31). Another was that microscopic examination concentrated on the few viable cells and ignored the whorling architecture of the acellular collagen. In addition, in

tumors showing the histologic pattern of meningiomas, there is uncertainty in the literature as to the criteria for a diagnosis of malignancy, although there is general agreement that the diagnosis implies a tendency to aggressive growth and recurrence despite apparent total removal (31). Features such as focal hypercellularity, high mitotic rate, focal necrosis, atypical mitotic figures, or papillary architecture have been proposed as indications of malignancy (32–35), however, none of the meningiomas in this series showed these features. Brain invasion by tumor has been used to diagnose malignancy (31) and was seen in four of our cases, all in the sclerosing group, yet their prognosis (Table 1) appeared similar to the nonsclerosing (and noninvasive) meningiomas. The one case (case 3) that recurred, and which is presumed to have died, was incompletely resected at the first operation but did not show evidence of brain invasion. This casts doubt on the validity of using brain invasion as an indicator of malignancy in childhood meningioma.

Summary

Primary meningeal tumors are uncommon in children. The largest group, the meningiomas, have very similar CT appearances to those seen in adults. They differ in being more commonly intraventricular or nondural in location and do not show the female predominance seen in adults. It may be possible to differentiate these apparently benign meningiomas from other, usually malignant, meningeal tumors (particularly meningeal sarcomas) by a combination of clinical and CT findings: meningiomas having longer symptom duration, and presenting in an older child with CT findings that are "typical" for adult meningioma, and malignant primary meningeal tumors having a more acute presentation in a younger child and with CT findings of a dural mass that more often includes atypical features such as extensive heterogenous enhancement, hemorrhage, large cystic areas, fringed or poorly defined margins, or diffuse involvement of the meninges.

Sclerosing meningiomas are recognized as a distinct histologic subtype of meningioma, whose importance lies in the possibility of confusion with other meningeal and nonmeningeal tumours. These tumors commonly present with seizures, and with calcification on CT. Histologically, they may show brain invasion without other signs of malignancy, a feature that, therefore, may not be a reliable indicator of malignancy in meningioma and that does not appear to correlate with a worse prognosis in the short term.

The bad reputation previously ascribed to childhood primary meningeal tumors should be confined to that group that are malignant. Meningiomas have a more favorable prognosis, at least in the short term.

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