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CASE REPORT

Malignant Sphenoid Wing Meningioma in Childhood

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ABSTRACT

Paediatric cranial meningiomas are uncommon, constituting 4% of all intracranial tumours in early childhood to adolescent. They frequently show cellular atypia and anaplasia. Paediatric meningiomas arising from the sphenoid wing, although possessing characteristic imaging features, can involve other skull base structures making surgical excision difficult. Prognosis is therefore not as favourable as for lesions at other cerebral convexities. The computed tomographic and histopathological findings, together with the clinical course of a sphenoid wing meningioma that turned malignant, in a 5-year-old boy are presented.

Key Words: *Child; Meningioma*

中文摘要

蝶骨翼惡性腦膜瘤小童病例報告

蘇銳新、吳恆堅

小兒顱內腦膜瘤很罕見，通常佔幼兒至青少年各種顱內腫瘤的4%。此症一般出現腫瘤的細胞變異性及退行分化。起源於蝶骨翼的小兒腦膜瘤雖然具有獨特的影像特徵，但因為可以牽涉其他顱底結構，很難利用手術進行切除。因此與其他大腦穹隆的腫瘤比較，蝶骨翼腦膜瘤預後不那麼好。本文報告一名患有蝶骨翼腦膜惡性瘤的5歲男童，包括其CT和組織病理學結果以及臨床表現。

INTRODUCTION

Cranial meningiomas are uncommon in children. They account for 1 to 3% of all meningiomas[1] and 4% of all categories of intracranial tumour in patients aged younger than 17 years.[2] A distinctive aspect of meningioma in this age-group is its predilection to show cellular atypia[3] and anaplasia in up to 5% of patients.[4] An important subgroup originates from the sphenoid wing. Although these tumours may have typical imaging findings, their broad extent at the skull base causes difficulties in complete surgical resection.[3,5] The recurrence rate is high in incomplete resections.[2] We describe the imaging features, histopathological findings, and clinical course of a sphenoid wing meningioma in a young boy and briefly review similar cases in the literature.

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CASE REPORT

A 5-year-old Vietnamese boy presented to our institution in September 1981 with a three-month history of progressive left-sided visual impairment. Physical examination showed proptosis, third cranial nerve paresis and papilloedema of the left eye. Cranial computed tomography (CT) revealed an enhancing mass arising from the left sphenoid wing that extended into the retro-orbital space and the left parasellar / cavernous sinus region (Figure 1a). There was hyperostosis of the left sphenoid bone (Figure 1b). The imaging features were consistent with a meningioma. An external carotid angiogram showed the tumour feeders that were too fine for preoperative embolisation. The lesion's intracranial portion including its cavernous sinus extension was excised following ligation of left internal carotid artery. Histology revealed tightly packed and abundant calcified psammoma bodies, classical of a benign meningioma. The tumour's intra-orbital component was treated by radiotherapy. Screening for neurofibromatosis yielded negative results. On examination nine months later, the patient showed residual disease in the left orbit and cavernous sinus.

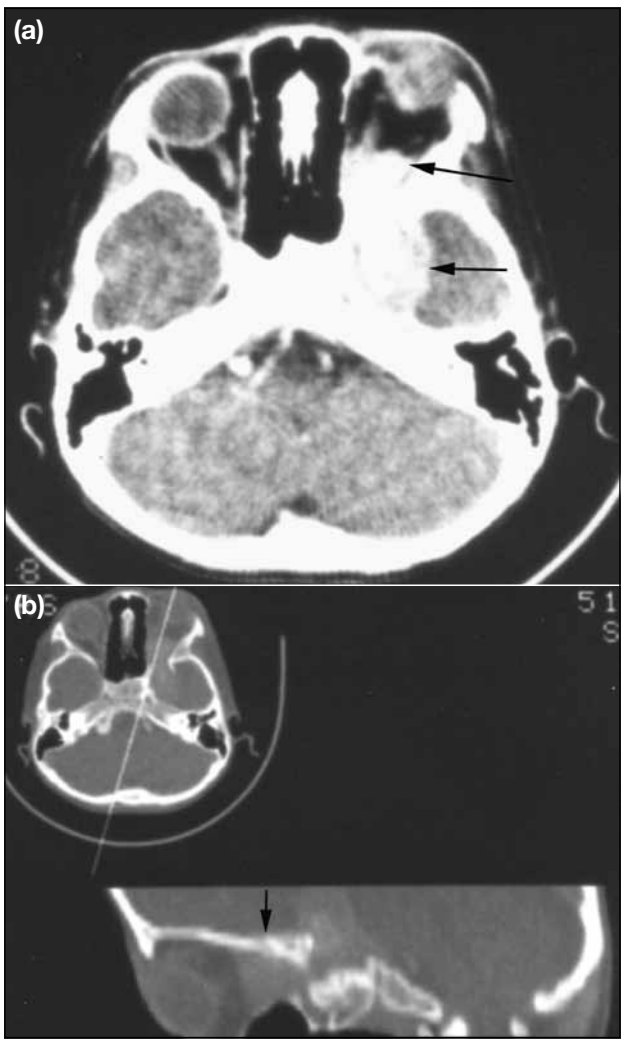

Figure 1. (a) Axial computed tomography through the skull base shows a well-defined uniformly enhancing mass along the left sphenoid wing and spreading to the retro-orbital space and left cavernous sinus region (arrows). Note left proptosis. (b) Sagittal reconstruction along the left orbital axis using a bone algorithm re-demonstrates the extent of posterior orbital involvement. Increased bone density is present in the medial sphenoid wing (arrow).

In December 1985 he was readmitted because of raised intracranial pressure. CT showed a 4-cm diameter lobulated, enhancing mass originating from the anterior and mid skull base extending to the level of the low parts of the lateral ventricles (Figure 2). There was white matter oedema surrounding the tumour, indicating its aggression. A large subfrontal tumour attached to the planum sphenoidale and tuberculum sellae was removed. Mitosis, necrosis, and irregularly arranged tumour cells with indistinct margins were noted histologically, which were characteristics of undifferentiated malignant meningioma.

In May 1986, the patient complained of visual deterioration in his right eye, and CT demonstrated an ill-defined mixed density mass between the frontal lobes (Figure 3). A large left subfrontal tumour, extending contralaterally to involve the right frontal lobe and partially encase the intracranial portion of right optic nerve, was debulked. Histology showed cords and clusters of malignant cells infiltrating cerebral tissues. Presence of large nuclei and prominent nucleoli were consistent with anaplastic meningioma (Figure 4). The boy died five months later.

DISCUSSION

On plain CT, paediatric cranial meningiomas have a heterogeneous appearance and cause a hyperostotic reaction of adjacent bones in about 50% of cases,[6] and intratumoural calcifications are present in 20 to 50%.[3,6] There is heterogeneous enhancement on post-contrast CT, as only the solid components show contrast uptake.[1,2,6] Presence of intralesional haemorrhage, irregular margins, and bone erosions are suggestive but not diagnostic of malignancy.[2,6]

Cyst formation in paediatric cranial meningiomas has been described in up to 50% of cases[3,7] and is an indicator of probable malignancy.[2,6] Tumour cysts are smaller than the solid components, ranging between

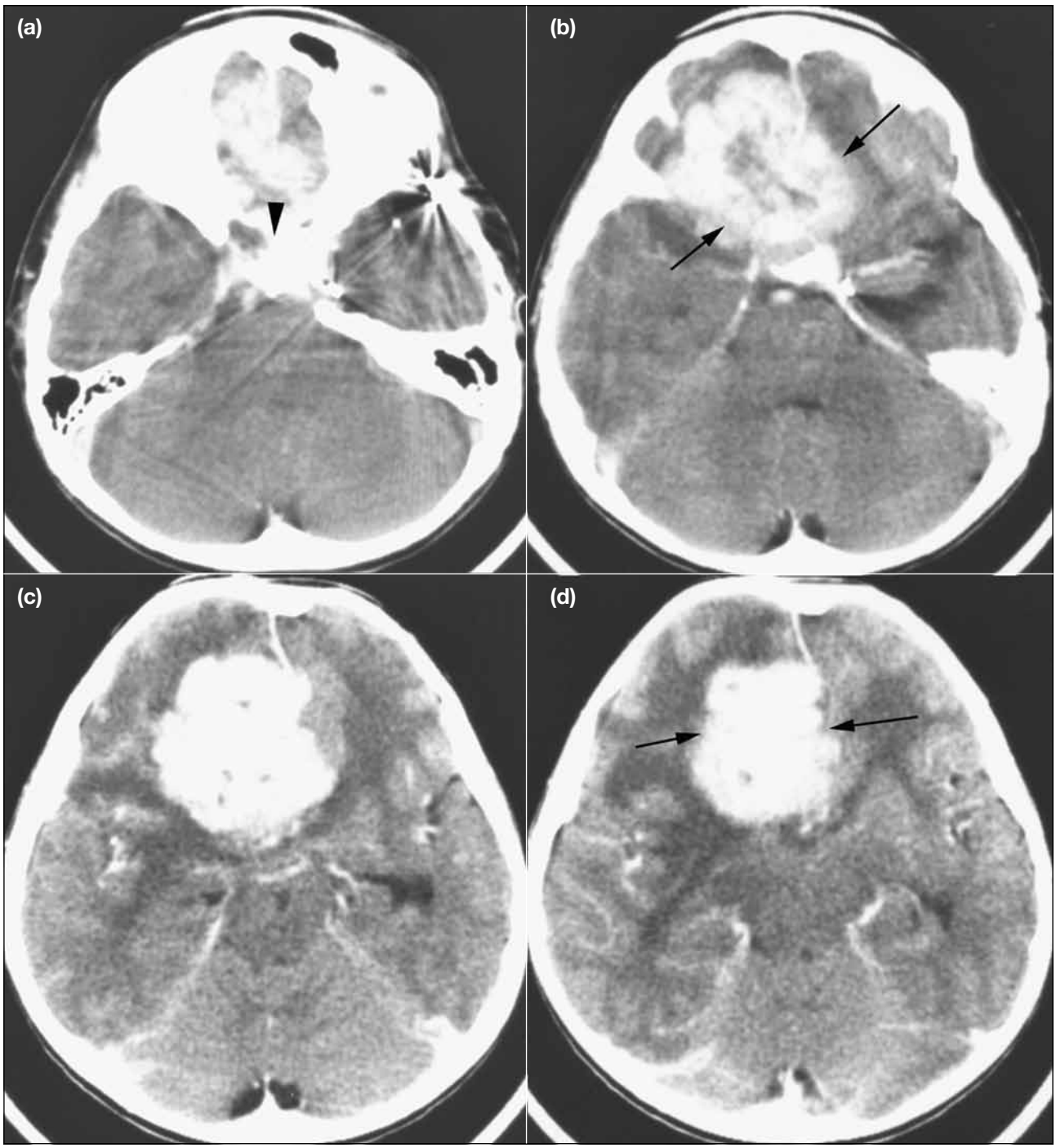

Figure 2. A series of axial computed tomographic scans from the level of the sphenoid wing to the level of the low lateral ventricles shows the tumour's origin at the anterior and mid skull base (arrows in b). The large lobulated enhancing mass with surrounding white matter oedema is most distinctive in (c). The tumour has effaced the lower part of right anterior horn (arrows in d). There is abnormal enhancement of the tuberculum sellae (arrowhead in a).

10 and 30% of the total size.[8] On magnetic resonance imaging (MRI), cyst contents have low signals in T1-weighted images and high intensity in T2-weighted sequences.[8] While meningiomas usually enhance intensely and uniformly, a heterogeneous pattern (probably reflecting intratumoural calcifications and cysts) has also been described.[1,7] CT and MRI are complementary in the preoperative workup of sphenoid-orbital meningiomas, though contrast-enhanced MRI is superior for detecting dural involvement.[9]

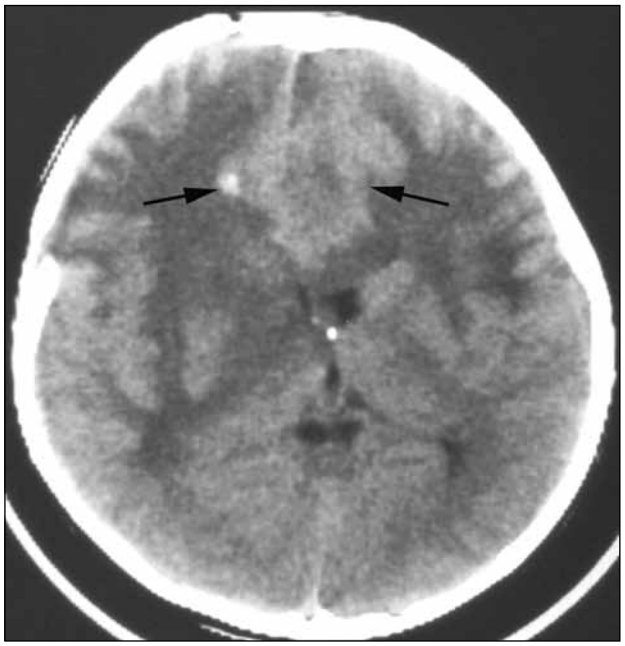

Figure 3. Plain axial computed tomography performed six months after the series shown in Figure 2. This second recurrent tumour between the frontal lobes has indistinct and fringed margins suggestive of brain invasion (arrows). Symmetrical white matter oedema can be either vasogenic oedema or post-radiation reaction.

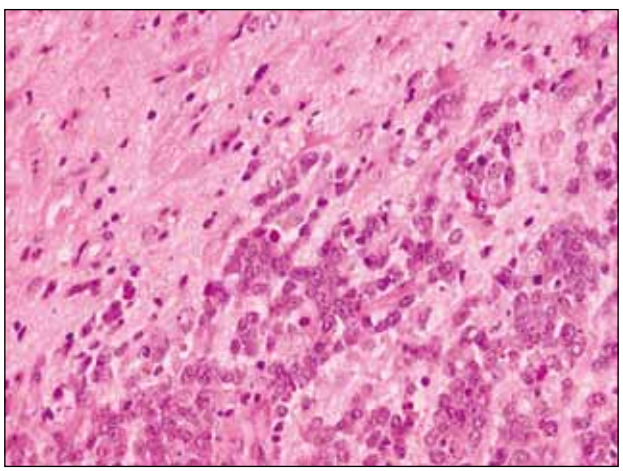

Figure 4. Cords and clusters of malignant meningioma cells infiltrate cerebral tissues. The meningioma cells are pleomorphic with enlarged nuclei and prominent nucleoli. Several mitoses are seen.

Unlike in adults, sphenoid wing meningiomas in childhood are relatively uncommon. Two recent series have quoted a frequency of 1.2 to 9%.[7,10] The present case had CT features strongly suspicious of such a lesion; the dense enhancements, sharp margins, site of origin and sclerotic reaction of adjacent bone were the characteristics. Two reports have described almost identical radiological abnormalities.[3,8] Similar to other skull base meningiomas,[1] those along the sphenoid wing may extend to the middle cranial fossa, pterion, cerebello-pontine angle and the petroclival regions.[3] They are predominantly large and solid.[3,11] In our patient, discovery of another skull base meningioma three years following radiotherapy to the left orbit raised the remote possibility of it being a radiation-induced lesion. More probably the tumour had arisen from residual disease at the tuberculum sellae, as demonstrated by CT. Besides, the average period from cranial radiation treatment in childhood to clinical onset of recurrence is nine years.[12]

At this stage, the CT abnormalities were suspicious of malignancy. The only noticeable sign was extensive frontal lobe white matter oedema. However, peritumoural oedema is an unreliable marker, since in one series four meningiomas depicting this feature were histologically benign.[8] Conversely, a high-grade meningioma may not be surrounded by oedema.[1] The well-defined margins and uniform enhancement characteristics of the recurrent tumour seemingly typified a benign process. Yet the histopathological picture was overtly anaplastic, reinforcing the notion that CT criteria can be indeterminate in distinguishing malignant meningioma from those that are benign. It was in the context of the tumour's second malignant recurrence that the bifrontal white matter oedema and indistinct margins were regarded as ominous, the latter correlated positively with microscopic brain invasion.

With infiltration of the adjacent bone, complete surgical excision of sphenoid ridge meningiomas without causing significant neurological compromise is difficult.[12] There is risk of peri-operative mortality in children with small blood volumes.[3] Preoperative embolisation is mainly reserved for tumours in relatively inaccessible locations such as the parasellar regions.[2,3] Success depends partly on the size of the feeders, which were too fine to embolise in our patient. Therefore, tumours of the sphenoid wing have a less favourable prognosis in terms of recurrence and survival compared to those affecting the cerebral convexities.[3,4,13] This caveat also applies to adults.[14,15]

In radiology practice, an insight into the clinical course of paediatric sphenoid wing meningiomas is helpful. A survey from 1989 up to the present yielded eight cases from six separate publications (Table[2,3,8,10,11,16]).

Table. Childhood sphenoid wing meningiomas from 1989 to 2010.[2,3,8,10,11,16]

Series	Sex / age	Location	Clinical presentation	Imaging findings	Pathological type	Outcome
Ferrante et al,[16] 1989 (15 cases)	M / 13 years	Left sphenoid wing	Proptosis	Angiography: characteristic blush	Meningothelial	Alive and well: 3 years after total resection
Glasier et al,[11] 1993 (5 cases)	F / 3 months	Left sphenoid wing to floor anterior cranial fossa	Not stated	Solid. MRI: hyperintense T1 T2, PD; intense enhancement	Meningotheliomatous	Not stated
Darling et al,[8] 1994 (8 cases)	M / 9 years	Right sphenoid wing / middle cranial fossa	Headaches	Large, solid 10 x 7 x 8 cm	Meningotheliomatous	Not stated
	F / 18 years	Left sphenoid wing / pterion	Headaches	Solid 3 x 3 x 4 cm	Meningotheliomatous	Not stated
Tufan et al,[2] 2005 (11 cases)	F / 17 years	Right sphenoid wing / pterion	Right visual deficit	Solid. CT: strong enhancement	Meningothelial, WHO grade I	Alive and well: 12 years after total resection
Rushing et al,[10] 2005 (87 cases)	Not specified	Sphenoid wing only	Not stated	Not stated	Atypical, WHO grade II	Alive and well: 10 years post-surgery
Arivazhagan et al,[3] 2008 (33 cases)	Not specified	Sphenoid wing only	Not stated	Not stated	Benign	Alive and well: total resection
	Not specified	Sphenoid wing / petroclival	Visual disturbance	Solid. MRI: moderate-to-marked enhancement	Benign	Alive; partial resection

Abbreviations: MRI = magnetic resonance imaging; PD = proton density; CT = computed tomography; WHO = World Health Organization.

Apart from one case classified as atypical,[10] the rest belonged to the benign meningothelial subtype. The patient with atypical tumour was alive and well 10 years post-surgery. Three of the remaining seven cases were alive post-surgery.[8,11] Of the two patients in a recent series,[3] one was disease-free after complete resection, while the other with petroclival involvement still had residual disease despite undergoing multiple removals. In two other reports relating to a 13-year-old boy and a 17-year-old girl,[2,16] both had total resection and were alive and disease-free after 3 and 12 years respectively, indicating that a good outcome was achievable with complete resection.[2,7]

The unpredictable biological behaviour of cranial meningiomas makes offering a prognosis difficult.[10,12] Thus, Caroli et al[12] found an increased frequency of atypical histopathology on initial presentation. The recurrence rate was high in children who had subtotal removal and almost all progressed to a malignant histology.[12] In our patient, rapid recurrence of a second malignancy (leading to death within five months) gives credence to this thesis. This unpredictability was further exemplified in a six-year-old boy with a supratentorial atypical meningioma that metastasised to subcutaneous tissues and lymph nodes of the upper neck.[17] Contrarily, some children with malignant-appearing meningiomas may have a good outcome.[10] Histology and immunochemistry remain the gold standard in arriving at a final diagnosis.

In summary, our case illustrates the difficulties encountered in the management of a sphenoid wing meningioma. CT identified its characteristic features and showed the tumour's broad extent at the skull base. Total surgical resection was incomplete, leading to recurrence and an adverse outcome.

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