

Secondary Extracranial Meningiomas Extending into the Paranasal Sinuses

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Rezumat

Meningioame secundare extracraniene extinse spre sinusurile paranazale

Meningioamele sunt tumori bine-cunoscute ale sistemului nervos central. Meningioamele extracraniene extinse secundar în sinusurile paranazale, sunt tumori rare care reprezintă aproximativ 2% din totalul meningioamelor. Meningioamele extracraniene ale sinusurilor paranazale pot reprezenta o provocare de diagnostic și tratament. Vă prezentăm o analiză retrospectivă care cuprinde trei cazuri și discutăm aspectele clinice, constatările imagistice, evaluarea diagnostică, precum și opțiunile terapeutice. Localizările tumorale au inclus sinusul frontal, sinusurile etmoidale, sinusul sfenoid și chiar sinusul maxilar. Rezecția chirurgicală completă a fost realizată la un singur pacient, în timp ce rezecția deliberat subtotală a fost efectuată în celelalte cazuri, în scopul de a evita deteriorarea neurologică severă, dar cu un control tumoral suficient.

Cuvinte cheie: meningioame extracraniene, sinusuri paranazale, diagnostic imagistic, rezecția chirurgicală

Abstract

Meningiomas are well-recognized tumors of the central nervous system. Extracranial meningiomas, secondarily extended into the paranasal sinuses, are rare tumors, comprising approximately 2% of all meningiomas. Extracranial meningiomas of the paranasal sinuses may present a diagnostic and therapeutic challenge. We present a retrospective review comprising three cases and discuss the clinical presentation, imaging findings, diagnostic evaluation, and treatment options. The localizations included the frontal sinus, the ethmoid sinus, the sphenoid sinus and even the maxillary sinus. Complete surgical resection was achieved in one patient; meanwhile deliberate subtotal tumor resection was performed in the other cases in order to avoid severe neurological damage with sufficient tumor control.

Key words: extracranial meningiomas, paranasal sinuses, imaging diagnosis, surgical resection

Introduction

Meningiomas are slow-growing and benign tumors with an annual incidence of 6 per 100,000 inhabitants (1). They are the commonest benign intracranial tumor, accounting for 13–26% of all primary intracranial tumors. Approximately 2% of intracranial meningiomas present extracranial dissemination at sites such as the orbit, middle ear, nasal cavity, nasopharynx and paranasal sinuses (1-3). The great part of intracranial meningiomas originates in females, while the largest part of primary extracranial meningiomas entails males (1-4). Diagnosis is established by physical examination, imaging

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studies, endoscopic biopsy of the tumor through the nasal cavity, and occasionally immunohistochemical studies are helpful for accurate diagnosis.

The only curative treatment is surgical extirpation. The development of different skull base approaches, such as fronto-temporal craniotomies with supplementary osteotomies, has encouraged more radical resection. Therefore, visualization and preservation of important functional structures and their vascular supply was improved.

Close cooperation of an interdisciplinary team of neurosurgeons, maxillofacial and otolaryngology surgeons is therefore extremely important.

The aim of this study was to describe the clinical picture, diagnosis and surgical approach to extracranial meningiomas in an interdisciplinary setting.

Case series

During the time period 2003-2009 we encountered three cases of extracranial meningiomas in our department. The first case was a 57-year-old male who presented with complaints of headache and proptosis of the right eye (Fig. 1). The symptoms developed over the course of a year. There were no visual changes and no neurological deficits. Magnetic resonance imaging (MRI) showed an enhancing middle cranial fossa mass extending into the ipsilateral sphenoid sinus, cavernous sinus, ethmoid, invading the orbit, penetrating the infratemporal fossa, pterygomaxillary fossa and even the maxillary sinus (see Fig. 2). The final diagnosis was meningioma of the sphenoid wing extending into the cavernous sinus and paranasal sinuses. A combined infratemporal and lateral rhinotomy approach to the skull base was performed in order to remove the tumor. After incision of the dura, tumor removal was continued along the sphenoid wing, the orbital apex and the anterior clinoid process. While infiltrated dura was excised



Figure 1. Patient with a sphenoid wing meningioma invading the orbit and paranasal sinuses

radically, no aggressive effort was made to remove tumor infiltrating the cavernous sinus or orbital soft tissues deep to the periorbit, to avoid severe functional disturbances of the eye or brain. Also, tumor infiltrating the major vascular supply was left to avoid cerebral infarction. However, the periorbit was resected. Pathology demonstrated cells arranged in whorls, nests, and occasional sheets (see Fig. 3), corresponding to meningioma. Postoperatively, the patient recovered without complications and was directed to radiotherapy because of incomplete tumor resection. At the last follow-up, 2 years post-operatively, no recurrence has been observed.

Case 2 is represented by a meningioma invading the frontal and ethmoid sinus (see Fig. 4). In this case a craniofacial approach, the accepted surgical removal for tumors

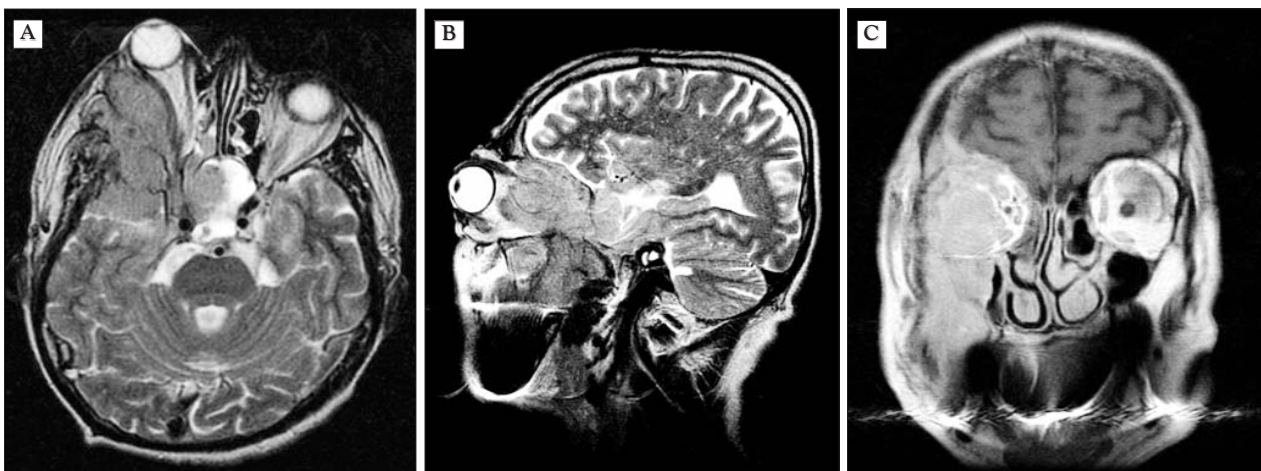


Figure 2. (A) – Axial T1-weighted MRI scan shows an intracranial mass at the anterior pole of the temporal lobe invading the sphenoid sinus, ethmoid and orbit. (B) – Sagittal T2-weighted MRI scan illustrates a mass extending into the sphenoid sinus, orbit and maxillary sinus. (C) – Coronal T1-weighted MRI demonstrates a tumor invading the temporal fossa, orbit, maxillary sinus

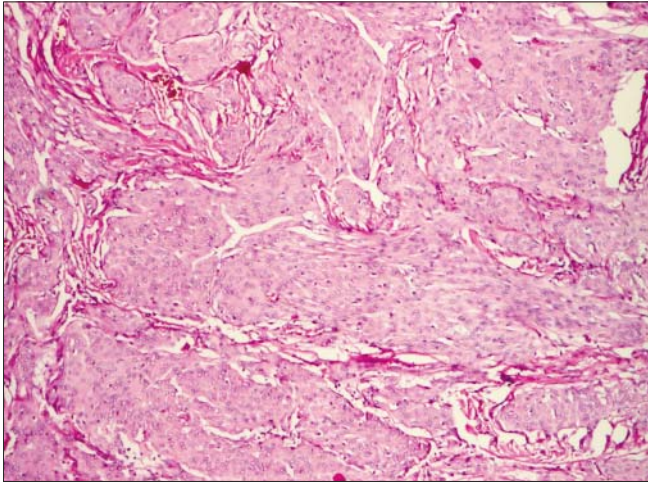


Figure 3. Interlacing bundles of spindle cells arranged in whorls, nests, and occasional sheets. Hematoxylin and eosin X 200

which have breached the anterior cranial fossa, permitted the radical excision of the whole tumor. Three years later, the patient is free of disease.

The third case is a recurrence within the orbit, frontal and ethmoid sinus (see Fig. 5). This patient had been operated 6 years before for a meningioma located on the dura covering the frontal lobe. A craniofacial approach was performed, but an incomplete tumor resection from the orbit was done. At 1 year postoperatively, the patient is free of disease.

Discussion

Meningiomas are the most common nonglial intracranial tumors, arising from arachnoid cap cells and accounting for 15% of all primary intracranial tumors (1-4). Meningiomas are slow growing tumors. Grossly, meningiomas are often firm, pink to grey, well circumscribed globular or lobulated tumors. Nevertheless, these tumors can be aggressive with local destruction and extracranial extension (5-7). Approximately 6-17% of all meningiomas can be found extracranially, primary in the head and neck region: in the nasal cavity and sinuses, the middle ear, the orbit, infratemporal fossa (1-7). According to the origin of meningiomas, these tumors have been classified into primary and secondary extracranial meningiomas (2,4,7). Primary extracranial meningiomas are not associated with an intracranial mass, and are believed to develop from heterotopic rests of meningoepithelial cells. Secondary extracranial meningiomas are extensions from intracranial disease. The tumors may enter the orbit, nasal cavity through preformed bony pathways, surgical defects, or foramina of the skull base (4-7). Meningiomas have been reported to compress the surrounding tissue without infiltrating it.

These skull base lesions often require both CT and MRI for adequate evaluation before treatment. Meningiomas are usually enhance intracranial extradural lesions; however, these tumors may have several different appearances. When bone is involved, the most common findings are of sclerosis or hyperostosis. On T1-weighted MRI meningiomas are isointense to the brain, less commonly they are hypointense (5-7). They become isointense (50%) or hyperintense (40%) on T2-weighted

Figure 4. (A) – Coronal MRI scan demonstrating a tumor invading the frontal and ethmoid sinus. (B) – Sagittal CT scan demonstrating a tumor invading the frontal and ethmoid sinus

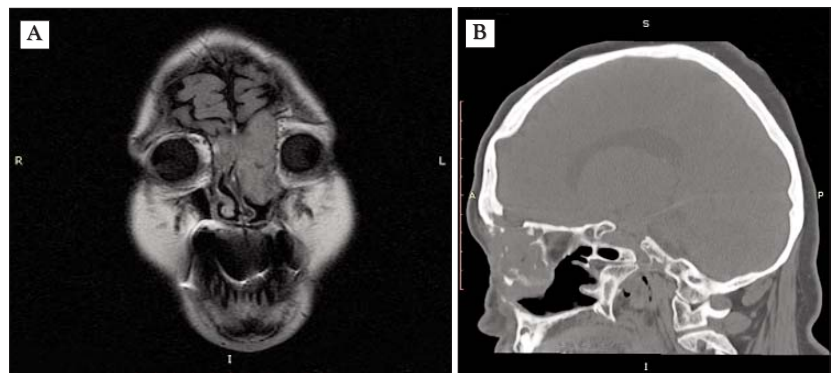
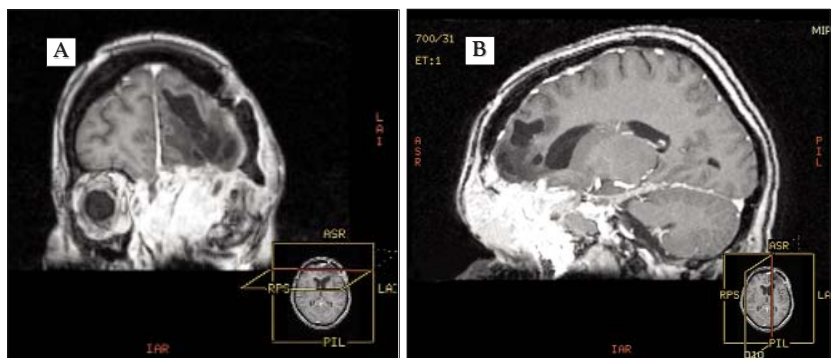


Figure 5. (A) - Coronal MRI illustrating a recurrent meningioma invading the orbit and ethmoid sinus. (B) – Sagittal MRI of the same patient



MR images (5). Meningiomas frequently show intense and homogenous enhancement with gadolinium. Enhancement of thickened adjacent dura, so called dura tail is commonly seen (4-7). On the other hand, angiography is indicated when the tumor site involves mobilization of major vascular structures at the skull base. Preoperatively this information may be used in an attempt to embolize the major feeding vessels (7).

The diagnosis of an extracranial meningioma is made by histological analysis. For practical purposes, 4 patterns have been described: syncytial (or meningothelial), fibroblastic, transitional, and angioblastic (1-7). The syncytial and transitional forms are the most common prototypes while the preponderance of primary extracranial meningiomas of the paranasal sinuses are of meningothelial type (4-7). There have been no reports of angioblastic type among extracranial meningiomas of the head and neck region. Recently, the following variants are described according to the World Health Organization (WHO) classification: Grade I – fibroblastic, transitional, psammomatous, angiomatous. These variants represent more than 90% of meningiomas. Atypical meningiomas (WHO II) show hypercellularity, mitosis and necrosis (5-7). WHO grade II tumors have a higher rate of recurrence (29–40%) than grade I tumors (7–20%), particularly after subtotal resection. Malignant meningiomas (WHO III) display brain invasion, rapid recurrence, and rarely metastasize. In addition, immunohistochemical staining is often necessary to confirm the diagnosis (8). Meningiomas show immunoreactivity with vimentin and epithelial membrane antigen (EMA), and are focally positive for *s*-100, keratin, and CEA, which helps to distinguish them from other tumors such as poorly differentiated carcinomas, gliomas, melanomas, sarcomas, and hemangioblastomas (4-8).

In the evaluation of patients with extracranial meningiomas, one must acknowledge the relationship between meningiomas and neurofibromatosis (3-5). Neurofibromatosis is a genetic disease and responsible gene mutations have been identified. It is classified into two subtypes: neurofibromatosis 1 (NF-1), or Von Recklinghausen's disease, which is the more common and is characterized by café-au-lait spots and peripheral neurofibromas. Patients with NF-I will occasionally have meningiomas. However, NF-II or "central" neurofibromatosis is more often associated with extracranial meningiomas, acoustic neuromas (usually bilateral), and other central nervous system tumors. Friedman et al. (3) recommend that any patient with the diagnosis of a primary extracranial meningioma should be screened for NF-II with MRI and audiometric testing.

The treatment of extracranial meningiomas is primarily surgical (3-7). Surgical extirpation is usually curative. However, due to the close relationship with the cavernous sinus and orbit, several cranial nerves and major blood vessels can be involved. Additionally, these tumours tend to involve the adjacent bones. Thus, the surgical treatment of those lesions requires a complex surgical approach to expose the intracranial and intraorbital structures. An interdisciplinary team approach is essential for merging each specific surgical skill and method, such as microsurgical technique on the one hand, and reconstructive techniques on the other.

Other authors have pointed out the limitation of tumor

resection in cases of cavernous sinus infiltration, including its cranial nerves and the adjacent arteries (9). The incidence of recurrence is reported to differ according to the pathological subtypes. Particularly, resection of the angioblastic type of meningioma is more difficult and recurrence is much more common. Meanwhile, the risk of recurrence is reported to be directly related to the sufficiency of the first resection. However, it is emphasized that some causes of tumor "recurrence" are the failure of early diagnosis, inadequate resection of the involved bony structures, and/or the dural en plaque tumour extension (9). In some cases, deliberate subtotal tumor resection is useful to avoid severe neurological damage with sufficient tumor control and a valuable progression-free survival (6,7).

Simpson described the recurrence rates of intracranial meningiomas after surgical excision, and proposed a grading system based on the degree of surgical excision (10). A grade 1 excision involved removal of the tumor bulk, of the surrounding dural attachment, and any involved bone; grade 2 removal is defined as removal of the tumor and diathermy of its dural attachment; and grade 3 excision was a macroscopic tumor resection with small foci left in situ. The recurrence rate at 5 years was 9% for grade 1 excision, 19% for grade 2, and 29% for grade 3 (10). On the other hand, there is also a role for palliative treatment with radiation. Radiation therapy is reserved for poor surgical candidates and for difficult recurrent lesions along the skull base (9).

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