CLINICAL PHOTOGRAPHS

Skull base meningioma extended to the left orbital, infratemporal and pterygomaxillary fossae

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Meningiomas are the most common intracranial tumors, representing about 15% of all intracranial primary tumors[1]. The site of origin can be variable, many tumors having the origin in the orbit or the intracranial cavity, with a possible extension into the maxillary or the ethmoid sinus. In 1982, Friedmann et al. classified the meningiomas according to the sinuses that can be involved, the most common sites of primary origin being the nose, the maxillary sinus and the frontal sinus[2]. Almost 6–17% of all meningiomas can be located extracranially or extraspinally[1].

Skull base meningiomas can be diagnosed in people of any age, most frequently in the 4th - 6th decade. The clinical symptoms have a direct correlation to the anatomical site involved, usually patients presenting nasal obstruction, proptosis, epiphora, pain or nose bleeding[1,3,4].

World Health Organization (WHO) classified meningiomas according to morphologic criteria into three grades[5]: grade I – benign meningiomas (80-90%), with low risk of recurrence or aggressive growth[5,6]; grade II – tumors with an increased mitotic activity and three or more of the following features: increased cellularity, small cells with a high nuclear/cytoplasmic ratio, prominent nucleoli, uninterrupted patternless or sheet-like growth, or foci of spontaneous or geographic necrosis[5-7]; grade III – anaplastic meningiomas - tumors with malignant characteristics resembling carcinoma, sarcoma or melanoma and a high recurrence rate[5-7].

The gold standard for the therapeutic approach for all meningiomas is complete surgical resection, including its dural attachment. In case of sphenoid sinus or infratemporal fossa involvement, the resection is most often partially, thus the radiation therapy may be needed. Surgery is not indicated in those cases with medial sphenoid wing or cavernous sinus involvement[8]. As meningiomas are highly vascular tumors, pre-operative embolization is often needed[8].

A 15-year-old girl was admitted in our Clinic with a history of fronto-temporal meningioma extended to the infratemporal, pterygomaxillary and intraorbital fossae, diagnosed 6 months prior, accusing persistent bilateral nasal obstruction (predominantly on the left nasal fossa), bilateral decreased visual acuity, predominantly left anterior rhinorrhea, left exophthalmia and hyposmia. The patient had undergone one neurosurgical intervention by infratemporal fossa approach 5 months earlier, with partial resection of the intracranial part of the tumor. The histopathological examination revealed a grade II meningioma. 3 months after the neurosurgical procedure, it was noticed an important tumoral growth, approximately 3 cm in coronal view on MRI evaluation.

The craniocerebral MRI revealed an expansive process involving the left sphenoidal wing, with ethmoidal and orbital, infratemporal and pterygomaxillary fossae invasion and intracerebral extranevraxial expansion into the antero-internal left temporal pole (Figure 1a). The tumoral mass measured approximately 6 cm in diameter (increased size since the last MRI performed before the neurosurgical intervention). The left optic nerve presented a curved trajectory with bushing of the external right muscle and orbital apex. In the ethmoid sinus, the tumoral mass developed caudal to the palate; the sphenoid sinus is totally embedded, the pituitary mass lesion is pushed by the upper periphery of the mass, reaching the optical chiasm formation with lift, shaft and bushing above the circle of Willis and the left sylvian artery until the middle horizontal portions of the sylvian valley (Figure 1b).

The ophthalmological examination revealed left eye protrusion with bilateral decreased visual acuity, optic atrophy (right eye partially, left eye almost totally), and no restricted movements for either eyes or diplopia.

The endoscopic endonasal appearance was of a red tumoral mass with midline location, embedding the nasal septum in its posterior half, extending to the nasopharynx and downward to the level of the nasal floor passages, with apparent origin in the skull base

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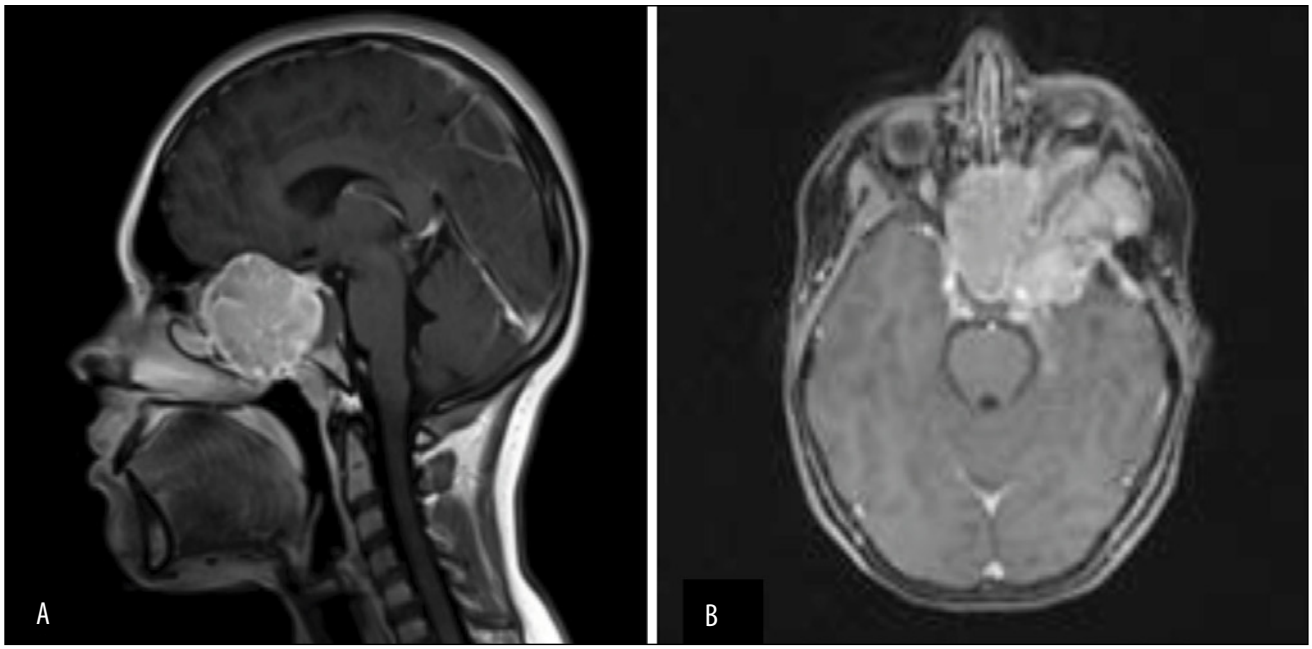

Figure 1 Craniocerebral MRI scan, sagittal (**A**) and axial (**B**) slices - expansive process involving the left sphenoidal wing, with ethmoidal and orbital, infratemporal and pterygomaxillary fossae invasion and intracerebral extranevraxial expansion into the antero-internal left temporal pole

(olfactory fossa) and posterior and superior to both the sphenoid sinuses (Figure 2).

The tumoral extension and the important vascularisation of the tumoral mass (a characteristic feature of meningiomas) make impossible a complete endoscopic resection. It was decided that this case will be managed in a multidisciplinary team of neurosurgeon, ENT surgeon, maxillo-facial surgeon, ophthalmologist, plastic surgeon and anaesthesiologist in order to perform a cranio-facial resection type 2. Given the important tumoral volume and the possible neurologic and haemorrhagic complications that may occur during or after the surgical procedure, it is probable that, even in a multidisciplinary team, the complete tumoral resection is impossible to be achieved. In this situation, the patient will receive postoperative radiotherapy.

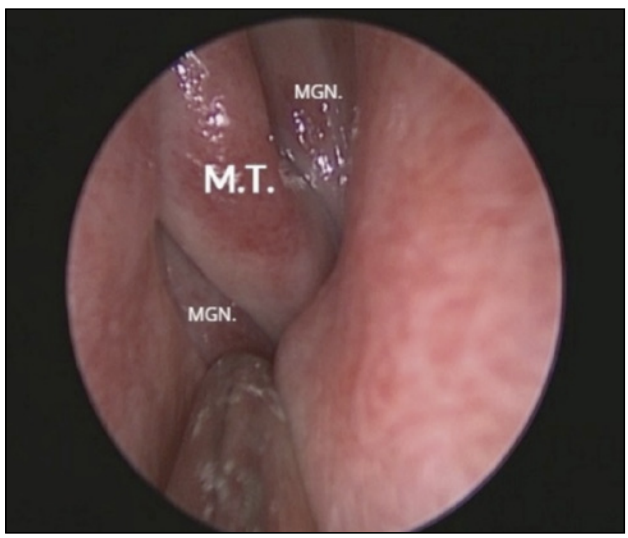

Figure 2 Intraoperatory view - red tumoral mass with midline location, embedding the nasal septum in its posterior half, extending to the nasopharynx, with apparent origin in the skull base (MT-middle turbinate, MGN-meningioma)

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