NASO-ETHMOID SCHWANNOMA WITH INTRACRANIAL EXTENSION

Case report

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ABSTRACT - Intranasal schwannomas are rare lesions, specially when they present with an intracranial extension. The fifth case in the medical literature of a naso-ethmoid schwannoma with extension into the anterior cranial fossa is presented. The magnetic resonance findings and the details of the combined intracranial / transfacial operative approach used are described. The possible origin and the clinical characteristics of this rare lesion are reviewed.

KEY WORDS: schwannoma, intranasal schwannoma, paranasal sinus tumor.

Schwannoma naso-etmoidal com extensão intracraniana: relato de caso

RESUMO - Schwannomas intranasais são lesões raras, principalmente quando apresentam um extensão intracraniana. Estamos apresentando o quinto caso da literatura médica de um schwannoma naso-etmoidal com extensão para o interior da fossa craniana anterior. São descritos os achados da ressonância magnética e os detalhes da via de acesso cirúrgico combinada intracraniana/transfacial. A possível origem e as características clínicas dessa lesão rara são revistas.

PALAVRAS-CHAVE: schwannoma, schwannoma intranasal, tumor de seio paranasal.

Any nerve with a schwann cell sheath may give origin to a Schwannoma and so, this neoplasm may develop in almost any part of the body¹, ². Although up to 45% of all schwannomas occur in the head and neck region³, the involvement of the nasal cavity and paranasal sinuses is rare, with approximately 40 cases reported³-18. From those cases, only four have been associated with intracranial extension into the anterior cranial fossa⁴-8,18.

CASE REPORT

A 40-year-old white woman presented to the Department of Neurosurgery of the Hospital Santa Marcelina in March, 1995 with a 3-year history of frontal headaches, gradual loss of the olfaction, altogether with a bulging deformity in the frontal area. Eight months prior to admission the patient became anosmic and developed a painful swelling of the frontal area, over the mentioned deformity. At examination she had no neurological abnormalities besides a bilateral anosmia. Magnetic resonance imaging revealed a mass lesion that filled the superior part of the naso-ethmoid complex and extended superiorly into the anterior cranial fossa (Fig 1). The tumor was successfully excised through a combined intracranial and transfacial procedure. A bifrontal craniotomy was performed and after extradural elevation of the right frontal lobe a white and smooth tumor was displayed. After the tumor was debulked its intracranial portion was completely removed extradurally to the level of the cribiform plate. The projection of the tumor into the ethmoid sinus and nasal cavity was then totally removed through a lateral rhinotomy. Microscopic examination revealed a tumor that consisted of regions of dense spindle cells, arranged in short bundles and forming interlacing fascicles, and of regions of a loose, myxoid matrix. The histological diagnosis, confirmed by immunohistochemical studies, was of a benign schwannoma (Fig 2). Shortly after the surgery the patient returned to her previous work and more than five years after the surgical treatment she is doing quite well, with a persistent bilateral anosmia. Recent image studies demonstrated the absence of any residual or recurrent tumor (Fig 3).

DISCUSSION

Schwannomas are, in the overwhelming majority of cases, benign slowly growing tumors that characteristically expand and thin the bony confines of

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the cavities and foramina in which they arise. The tumor in the present case, as well as the other four reported in the literature, presumably arose from one of the intranasal nerves and grew until it eroded the floor of the frontal fossa to reach the intracranial compartment. The precise origin of the intranasal schwannomas is obscure, as there are many nerve branches in the region. They may arise from any of the following nerves: (a) general sensory branches of the ophthalmic and maxillary divisions of the trigeminal nerve; (b) autonomic fibers (parasympathetic) from the sphenopalatine ganglion and (c) autonomic fibers (sympathetic) derived from the carotid plexus. As the olfactory nerves are covered by glial cells they cannot give rise to nerve sheath tumors.

All the cases of nasal schwannomas with intracranial extension, including the one we are reporting, presented with a long history of nasal symptoms. The tumors were large at the diagnosis and all of them were completely resected by an intracranial approach or by a combined intracranial / transfacial approach.

The CT or MR images are non specific. The features presented are those of a benign, slow growing tumor of greater signal intensity than polyps or mucoceles. The differential diagnosis must include papilloma, sarcoma, carcinoma, and lymphoma.

Although a schwannoma within the nasal cavity, specially with an intracranial extension, is a rare occur-
rence, it should be part of the differential diagnosis of intranasal lesions elected to be submitted to biopsy and CT or MR imaging should be taken prior to the biopsy procedure.

REFERENCES


Fig 3. Post-operative magnetic resonance images. T1-weighted coronal (A) and sagittal (B) images obtained after IV administration of gadolinium demonstrated the absence of any residual or recurrent tumor. Some degree of residual atrophy of the frontal lobe is shown.