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Caso clínico

Primary nasal meningioma: an unusual diagnosis

Meningioma nasal primário: um diagnóstico incomum

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Resumo

Introdução: Os meningiomas são tumores comuns que, por vezes, podem originar-se em locais extracranianos podendo estes representar uma disseminação secundária da lesão intracraniana ou tratar-se de um meningioma primário sem qualquer relação com as estruturas subdurais, sendo este um achado raro. Uma vez que a forma de apresentação do meningioma primário mimetiza outras patologias da área da cabeça e pescoço é necessário que esta entidade seja considerada no diagnóstico diferencial das mesmas.

Caso clínico: Doente do sexo masculino, 60 anos, recorre ao serviço de urgência de Otorrinolaringologia por queixas com dois meses de evolução, de uma massa intranasal de crescimento progressivo, associada a obstrução nasal e epífora. Ao exame objectivo apresentava uma massa de grandes dimensões que ocupava a totalidade da fossa nasal direita. Tanto a tomografia computorizada como a ressonância magnética identificaram uma massa volumosa sem qualquer componente intracraniano associado. O doente foi submetido a exérese cirúrgica completa da lesão. O diagnóstico foi estabelecido através do estudo histológico e imuno-histoquímico. Actualmente o doente apresenta-se assintomático e sem sinais de recorrência.

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Discussão: Os meningiomas são tumores normalmente com características benignas mas que, por vezes, podem apresentar um comportamento agressivo com destruição local. Imagiologicamente apresentam-se como uma massa isodensa ou hiperdensa que na ressonância magnética é realçada pelo contraste. O estudo imuno-histoquímico é fundamental para o diagnóstico. O tratamento de eleição é a exérese cirúrgica.

Palavras-chave: Meningioma Primário Extracraniano, Tumor, Nasal

Abstract

Introduction: Meningiomas are common tumors that sometimes appear in extracranial sites representing a secondary dissemination of the intracranial lesion or a primary meningioma with no relation with the subdural structures, which is a rare finding. The clinical presentation of a primary meningioma of the head and neck mimics other pathologies of this area and for that reason it should be included in the differential diagnosis when studying these lesions.

Case Report: A 60-year-old male patient presented at the Emergency Otorhinolaryngology Department with a two months history of an intranasal mass associated with nasal obstruction and epiphora. On physical examination it was verified a large mass that occupied the whole right nasal cavity. Both computed tomography and magnetic resonance imaging identified a bulky mass without any intracranial component associated. The patient underwent a complete surgical excision of the lesion. The diagnosis was established through the histological and immunohistochemical study. Currently the patient is asymptomatic without signs of recurrence.

Discussion: Meningiomas are tumors that usually have benign features but sometimes may exhibit an aggressive behavior with local destruction. Imaging normally present an isodense or hyperdense mass that is enhanced by contrast on magnetic resonance imaging. The immunohistochemical study is fundamental for the diagnosis. The treatment of choice is surgical excision.

Key-words: Primary Extracranial Meningioma, Tumor, Nasal

Introduction

Meningioma is a slow-growing tumor arising from meningocytes (arachnoid cap cells) of arachnoid granulation tissue. It is the most common nonglial intracranial neoplasm, constituting 15-20% of all intracranial tumors[1].

Approximately 6-17% of all meningiomas can be found in extracranial or extraspinal sites. The head and neck is the most common site, usually resulting from the extension of a primary intracranial tumor (secondary extracranial meningioma)[2].

Less than 2% of all meningiomas arise at a primary ectopic site (primary extracranial meningioma (PEM))[3]. PEMs refer to meningiomas arising outside the subdural compartment as primary tumors, having no relation to any subdural structures[4]. PEMs in head and neck constitute 80% to 90% of all PEMs[4]. In

this location the tumors are usually associated with the presence of a mass, nasal obstruction, epistaxis, exophthalmos, swelling, local pain or impaired vision[4]. These are the symptoms and signs that bring the patient to the Otorhinolaryngology specialist, however these are inespefic findings that can be presented in multiple other pathologies. Therefore PEMs should be considered as a differential diagnosis in the study of these type of lesions.

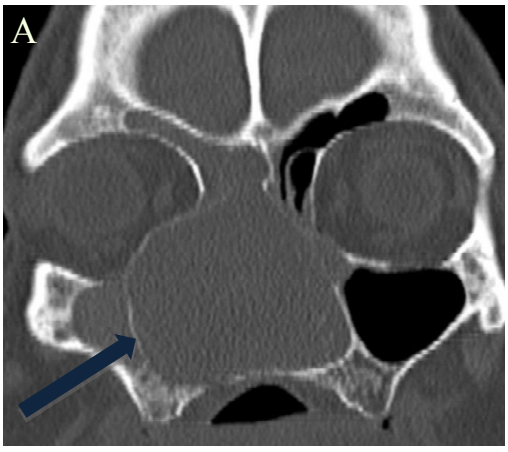
Clinical case

A 60-year-old man presented at the Emergency Otorhinolaryngology Department with complaints of an intranasal enlarging mass associated with nasal obstruction and epiphora with two months of duration. He denied recurrent epistaxis, pain or changes in visual acuity. There was no past history of trauma of the face or nose. He had worked with woods for over 40 years without nasal protection.

On physical examination he presented with swelling of the right lateral side of the nose. A mass of hard consistency occupied all the right nasal cavity, reaching the nasal vestibule and causing contralateral displacement of the nasal septum. Oculomotricity was preserved and neck palpation was innocent.

A Computed Tomography (CT) scan (Figure 1A), requested to evaluate the disease's extension, revealed a massive naso-ethmoidal mass, with homogeneous soft tissue density and with slight enhancement after administration of intravenous iodinated contrast. The mass totally occupied the nasal cavity and the ethmoidal cells, with anterior extension to the nasal vestibule and posterior extension to the nasopharynx, causing marked left deviation of the nasal septum. No intracranial component was identified.

The subsequent Magnetic Resonance Imaging (MRI) (Figure 1B) exhibited a bulky mass with heterogeneous sign in T_2WI, low sign in T_1WI and intense enhancement with contrast. This lesion caused apparent destruction of the nasal septum, right nasal turbinates, medial wall of the maxillary sinus and the infero-medial wall of the right orbit, "touching" the inferior rectus muscle without evidence of direct compression of the right eyeball. The lesion measured 5.0 cm (transverse) by 8.0 cm (anteroposterior) by 4.8 cm (craniocaudal). There was no evidence of anomalous enhancement zones in the intracranial compartment.



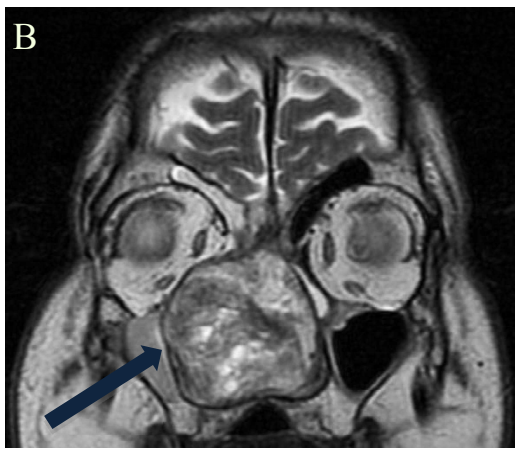
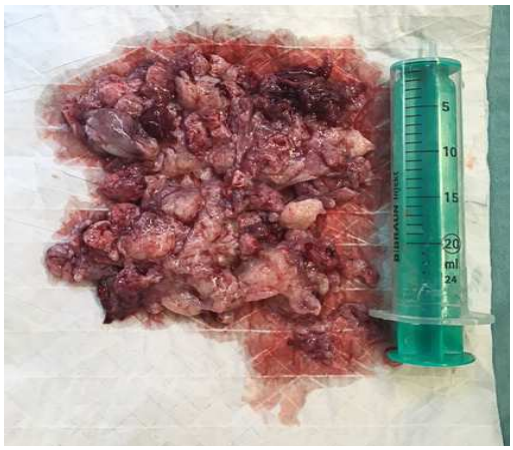


Figure 1A: TC image shows a massive naso-ethmoidal mass. **Figure 1B**: MRI image reveals a bulky mass intensely enhanced with contrast.

The patient was submitted to a complete excision by endoscopic sinus surgery. Intraoperatively it was verified that the lesion apparently originated from the posterior region of the nasal septum. All the mass was removed (Figure 2). Bone integrity of the nasal walls was verified with no evidence of cerebro-spinal fluid leakage.

Histopathology of the specimen showed proliferation of fusiform and oval cells, in some areas disposed in a spindle type, without atypia or mitosis. The immunohistochemical study (Figure 3A and 3B) showed immunoreactivity of the neoplastic cells to vimentin, without expression of EMA, S100, CD31, AE1AE3 and SMA. Despite the absence of EMA expression, diffuse immunoreactivity to vimentin and morphological aspects favored the hypothesis of meningioma. After six months of follow up the patient remains free of recurrence.

Figure 2*:* Image of the mass removed during surgery.



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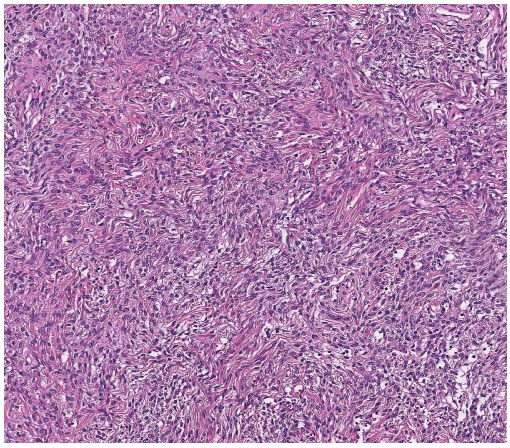

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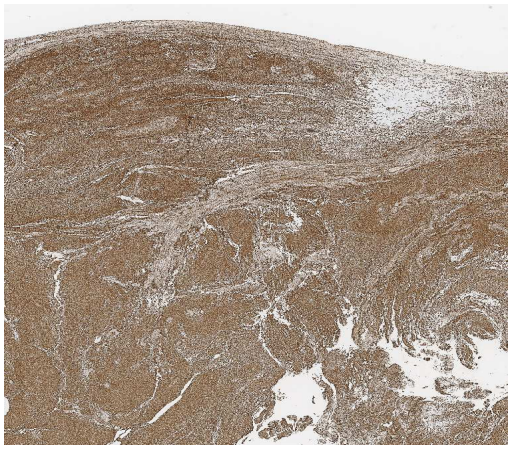

Figure 3A: Haematoxylin and eosin staining of the specimen showing fusiform and oval cells disposed in a spindle type.

Figure 3B: Immunoreactivity of the cells to vimentin staining.

Discussion

Histologically meningioma has benign characteristics, however, it can be aggressive, with local destruction of tissues and even metastasis[2].

The origin of PEMs is still unclear and multiple hypotheses have been proposed, including:

1) Origin from ectopic meningocytes in cranial and peripheral nerve sheaths and in subcutaneous tissue.
2) Arising from misplaced arachnoid cells in an extracranial location during embryologic development.
3) Stemming from arachnoid cap cells trapped in the cranial suture during birth and molding of the head[4].

PEMs in head and neck are more frequent in female and have two peaks in age distribution. The first occurring during the second decade of life and the second during the fifth through seventh decades[4].

On CT scan meningioma typically constitutes an isodense or hyperdense mass. MRI generally reveals an isointense mass on T_1WI and T_2WI is intensely enhanced with contrast[5].

The ultra-structure of meningioma shows two types of cells with rounded or oval and elongated nuclei in spindle type[2].

Immunohistochemical studies are indispensable in the differential diagnosis of primary extracranial meningiomas, excluding other benign tumors of peripheral nerve origin. The positive expression of EMA and vimentin antigen demonstrates the dual epithelial and mesenchymal characteristics of the cells found in these tumors[2].

Surgical resection is the first-choice treatment due to tumor radioresistance and recurrence is very rare following adequate surgical resection[2,5].

Conflicts of interest: No conflicts of interest were declared by the authors.

References

1- Leestma JE. Brain tumors. Am J Pathol. 1980;100(1):239-316.

2- Aiyer RG, Prashanth V, Ambani K, Bhat VS, Soni GB. Primary extracranial meningioma of paranasal sinuses. Indian J Otolaryngol Head Neck Surg. 2013;65(Suppl 2):384-387.

3- Friedmann CD, Costantino PD, Tietelbaum B *et al*. Primary extracranial meningiomas of the head and neck. Laryngoscope. 1990; 100:41–48.

4- Liu Y, Wang H, Shao H, Wang C. Primary extradural meningiomas in head: a report of 19 cases and review of literature. Int J Clin Exp Pathol. 2015; 8(5):5624-5632.

5- Wang Z, Wang Y, Zhao X, Zhang J. Primary sphenoid sinus meningioma resection via transnasal transsphenoid approach: a case report. J Int Med Res. 2015 Apr;43(2):270-5.