

## **RELATO DE CASO: MENINGIOMA MENINGOTELIAL NASAL**

Case report: nasal meningothelial meningioma

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**Abstract:** Meningiomas are the most common primary brain tumors nowadays. They arise from the dura mater being composed of neoplastic meningothelial cells. Their incidence increases with age, being more common in women and occurring more commonly in people with previous history of cranial irradiation, being their extracranial variant a rare event and of pathogenesis still uncertain. The present study analyzes the case of a 46-year-old female patient who was admitted to Santa Casa de Montes Claros Hospital on June 29th, 2018 after resection of a nasal lesion and anatomopathological study with a result compatible with grade I meningioma. The study also approaches the characteristics and classifications of meningiomas, as well as their clinical and therapeutic repercussions. It also pervades the diagnostic criteria and an analysis of the signs and symptoms referring to the nasal extracranial presentation, which applies to the patient reported in the present case.

Keywords: Meningioma; Brain tumors; Extracranial.

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### INTRODUCTION

Meningioma is a common type of intracranial neoplasm that arises from the meningothelial arachnoid cells around nerves or heterotopic transposed cells at the time of closing the middle line in fetal life in the meninges.<sup>1</sup> Deletions of chromosome 22 are frequent in this neoplasm, being the target observed the NF2 gene, which is often muted.<sup>2</sup> Regarding the histology, it is varied observing three described grades : Grade I - low proliferative potential; Grade II - low malignancy grade and Grade III malignant tumors.<sup>3</sup> Epidemiologically, it presents the ratio 2 women :1 man, and the prevalence increases with age, reaching its peak at 70 years.

Nasal meningothelial meningioma is a rare variant, with a frequency of 3% of all types of meningiomas. <sup>2</sup> This type of tumor is characterized by cells of inaccurate limits, homogenous cytoplasm, large and oval nucleus. The cells are arranged in lobules separated by fibrous stroma. <sup>3</sup>

The objective of this study is to report a case of a patient with nasal meningothelial meningioma.

#### **CASE REPORT**

G. S., Woman, 46 years old, was admitted to the department of oncology clinic of Hospital Santa Casa de Montes Claros on June 29th of 2018, after confirmation of meningioma in the nasal fossa. She reports that approximately 2 years ago she began to present persistent nasal obstruction. After suspicion of chronic sinusopathy she was forwarded to the Otorhinolaryngology, where she was subjected to nasofibrolaryngoscopy with resection of nasal lesion in January 2018. After resection, the piece was sent for anatomopathological study, revealing Histology compatible with grade I meningioma in the nasal fossa, being forwarded oncologic to evaluation. Subsequently, immunohistochemical study of the piece was requested, which showed results consistent with meningothelial meningioma grade I.

In previous history, the patient reports resection of two tumors in the central nervous system (both benign -SIC), in which there was total visual loss on the left and partial on the right part as a sequel to the therapeutic procedure. She denies comorbidities, smoking, alcoholism, use of chronic use of medications and allergy history. In addition, she informs family history of

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colon cancer in maternal uncle. Inhabitant of urban area and housewife occupation. She has no other complaints.

Under the anatomopathological study, macroscopy showed whitish nodule, delimited, firm-elastic to cuts, measuring 4.5 x 2.0 x 1.0 cm, with some stone-hard areas; histological sections revealed polypoid lesion lined by respiratory epithelium formed by proliferation of cells forming lobes delimited by slender collagen tissue, being the cells uniform with oval nucleus and delicate chromatin. The lesion reached the limits of fragments. In immunohistochemical analysis it was demonstrated that it was a hypercellular neoplasm, composed by cells slightly elongated, elongated nuclei and inconspicuous nucleoli. There was no invasion of brain parenchyma, atypia, increase in the number of mitosis or necrosis. It was also found expression of epithelial membrane antigen, and progesterone receptor, being the findings indicative of meningothelial meningioma grade I.<sup>4</sup>

Magnetic resonance imaging was requested of the skull and face and copy of histopathological examination of the resected tumor in 2002 for clinical exclusion of intracranial primary site, in addition to follow-up with an otorhinolaryngologist. Until the time of writing this article the examinations had not been carried out.

#### DISCUSSION

Meningioma, a tumor that is frequent in the central nervous system, being the tumors in the extracranial sites, such as the present case, a rare event. Some risk factors are involved in the development of this type of tumor, among them, the previous irradiation and female sex. The pathogenesis of the extracranial meningiomas is uncertain, hypotheses however some are embryogenesis, considered. During when arachnoid cells appear on the sheaths of the nerves or vessels that come out through the skull, the displaced bodies of Pacchionian, are highlighted, compressed or trapped in an extracranial region. Traumatic events also explain the case due to the displacement of arachnoid islets, which are originated from undifferentiated or multipotent mesenchymal cells as well as fibroblasts, Schwann cells or a combination of both, possibly explaining the diverse pathological spectrum of the meningiomas. <sup>4,5</sup>

World Health Organization (WHO) classifies meningiomas in 3 categories: being the grade I (benign) that has a low rate of recurrence being the most common histological types the syncytial or meningothelial, transitional, or mixed and fibroblastic or fibrous, usually in this grade; grade II (atypical) which is defined by the presence of at least one of the criteria: (1) 4 to 19 mitoses per 10 fields of large increase, (2) three or more of the following criteria: uncharacteristic architecture, hypercellularity, small cells. macronuclei and necrosis; and grade III are the histological subtypes rhabdoid papillary, as well as anaplastic, and characterized by mitoses above 20 per 10 fields of large increase and standard frankly undifferentiated, approaching the aspect of sarcomas, carcinomas or melanomas. Our patient had tumor Grade I, therefore having a positive outcome. 4,5

A variety of nonspecific signs and symptoms are reported, such as congestion, epistaxis, hydrorrhea, hyposmia, nasal obstruction, headache, maxillary pain, periorbitary edema, EXOFALMIA, proptosis, ptosis, with reduced visual acuity and hearing loss. Under nasal endoscopy, the lesion is presented as a lobulated, well circumscribed mass, which may or may not bleed, pink or gray color in the reported case, the patient had nasal obstruction. <sup>5,6</sup>

The diagnosis is made through the examination of pathological material, confirmed by histopathology and immunohistochemistry, and may exhibit the following findings: immunoreactivity for vimentin, EMA pancytokeratin, in addition of and positive reactions for CK7, protein S100, CAM 5.2, progesterone receptors and Ki-67 index (>1%), being the last two ones positive on examination of the patient in question, in addition to EMA. 1,5

Regarding the differential diagnosis, benign and malignant neoplasms are found, as well as neurogenic tumors which can be cited olfactory neuroblastoma and melanoma; tumors of mesenchymal tissue, such as aggressive psammomatoid ossifying fibroma and also vascular tumors, such as paraganglioma.<sup>1</sup>

Being a complete resection feasible, surgery is the treatment of choice. The adjuvant radiotherapy has been suggested for the treatment of patients with meningioma of the central

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nervous system. However, its role to the extracranial meningiomas has not been elucidated yet. <sup>1,5,6</sup>

## FINAL CONSIDERATIONS

Being the extracranial meningiomas, mainly in the head and neck region, a rare event, the report of this case becomes pertinent to the increase of knowledge about this oncologic manifestation. The diagnosis is based on symptoms, physical complementary examination and examinations computed such as tomography and nuclear magnetic resonance, for assessing the extent and surgical planning, and confirmed by the immunohistochemical study to perform the differential diagnosis of primary extracranial meningioma or extension of other benign tumors of peripheral origin. The treatment of choice is surgical excision, once that meningiomas are resistant to radiotherapy and have good prognosis

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and rare recurrences after surgery.

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