

# Extracranial meningioma in paranasal sinuses. A case presentation and literature review

## Meningioma extracraneal en senos paranasales. Presentación de caso y revisión de la literatura

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## Summary
Meningiomas are the most common primary non-glial intracranial neoplasms of the central nervous system, predominantly located extra_axially, derived from meningothelial cells. Extracranial, non-dural, or ectopic meningiomas are uncommon entities, with few cases reported in the literature, whose final diagnosis is achieved histopathologically. This report presents the clinical case of a female patient with recurrent headache and chronic rhinorrhea, whose imaging findings initially suggested a nasal polyp as the primary differential diagnosis. However, due to the lesion's inherent characteristics and its unusual location, an ectopic meningioma was suspected and later confirmed after complete surgical resection and histopathological analysis. As a diagnostic challenge, imaging findings reveal distinctive characteristics that are fundamental for their identification, diagnosis and management.

## Resumen
El meningioma es la neoplasia intracraneal primaria no glial más común del sistema nervioso central, predominantemente de localización extraaxial, derivado de las células meningoteliales. Los meningiomas extracraneales no durales o ectópicos son entidades infrecuentes, con poco registro en la literatura, cuyo diagnóstico se logra mediante análisis histopatológico. Se describe el caso clínico de una paciente con cefalea recurrente y rinorrea crónica, con hallazgos imagenológicos que inicialmente sugerían un pólipo nasal. Sin embargo, debido a las características inherentes de la lesión, a pesar de su ubicación inusual, se sospechó un meningioma ectópico, confirmado tras resección quirúrgica completa y análisis histopatológico. Dados su rareza y presentación atípica, este caso ilustra cómo los hallazgos imagenológicos pueden orientar hacia un diagnóstico poco habitual, convirtiéndose en una herramienta clave para el radiólogo en escenarios clínicos complejos.

## Clinical case
A 63-year-old female patient consulted for long-standing symptoms characterized by recurrent headache and rhinorrhea. Physical and neurological examination revealed no abnormalities. An initial brain magnetic resonance imaging (MRI) scan (Figure 1) was performed, with the only finding described as an ethmoid polyp, for which conservative management was indicated.

Two years later, with persistent symptoms accompanied by pain in the left eye, a new brain MRI was performed (Figure 2), which revealed a solid, extra-axial, left paramedial lesion at the base of the anterior cranial fossa. The lesion showed intra- and extracranial growth with intermediate signal intensity similar to that of gray matter in T1-, T2-, and FLAIR-weighted sequences, with homogeneous enhancement and no diffusion restriction. The mass showed a broad base of implantation with a dural tail and ectopic growth into the sphenoid sinus and ethmoid cells, with bone erosion and displacement of the straight gyrus without perilesional edema. A surgical procedure was performed three months later, which confirmed a meningioma by histopathological study.

## Discussion
Meningiomas are common benign meningoepithelial tumors of the central nervous system, accounting for an average of 20% of brain tumors (1). They have an annual incidence of approximately six cases per 100,000 and a prevalence of 50 to 70 per 100,000 (2). They have a female predilection with a ratio of 2:1 compared to men and are more common after the fifth decade of life (1-3).

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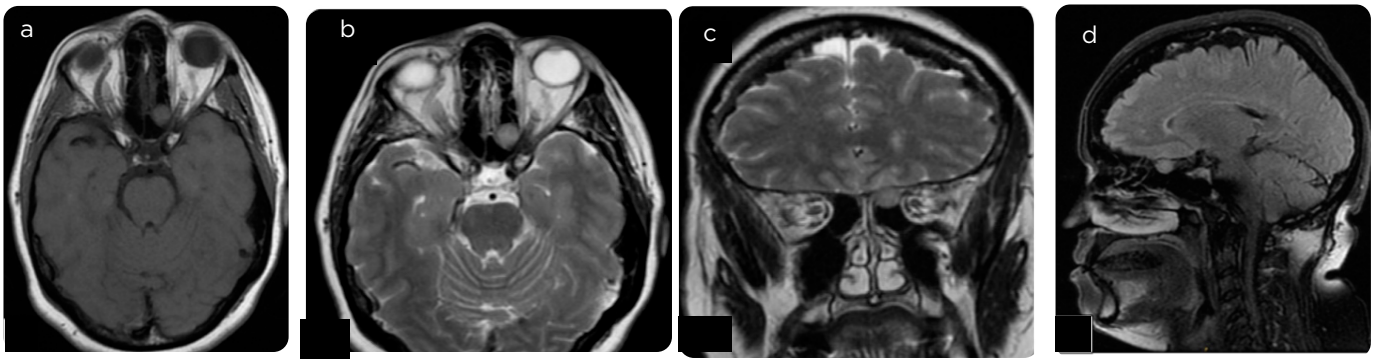



Figure 1. Brain MRI. a) Axial sequences with T1 and b) T2 information. c) Coronal T2 and d) sagittal sequences with spin echo T2 and FLAIR information: well-circumscribed polypoid lesion in the ethmoid cells with intermediate signal in the different sequences.

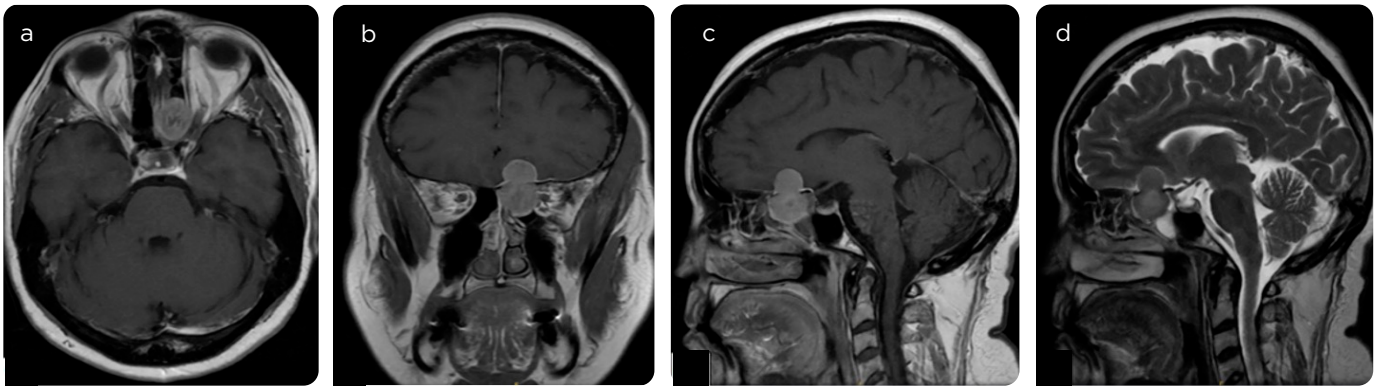



Figure 2. Follow-up brain MRI two years later. Sequences with T1 information with contrast medium: a) axial, b) coronal, c) sagittal, and d) sagittal in sequence with T2 FSE information. A solid, extra-axial, left paramedial lesion is identified in the anterior cranial fossa, anterior to the sella turcica, with extracranial extension. Predominantly intermediate signal in sequences with T2 information and moderate, homogeneous enhancement with contrast medium, with a slightly heterogeneous appearance at its lower margin. In these sequences, the dural tail can be delineated, with no edema of the adjacent cerebral parenchyma.

The World Health Organization (WHO) classification of central nervous system (CNS) tumors categorizes meningiomas into several subtypes and classifies them histologically into three grades (I, II, and III). They are predominantly benign (98%), with only 2% classified as malignant (3).

Meningiomas exhibit typical diagnostic characteristics. On non-contrast computed tomography (CT), they appear as a lobulated, high-density, homogeneous mass with well-defined margins and a broad dural attachment base. On magnetic resonance imaging (MRI), they vary from low to intermediate signal intensity on T1-weighted sequences and from medium to high signal intensity on T2-weighted sequences, generally without diffusion restriction. They usually enhance homogeneously after administration of iodinated contrast medium on CT or gadolinium contrast medium on T1-weighted MRI sequences. In addition, they may show other classic diagnostic features such as hyperostosis of the underlying bone, cerebrospinal fluid (CSF) separation, and a dural tail (2, 4). However, they may show less common features, such as heterogeneous or ring enhancement on contrast-enhanced studies, as well as cysts, hemorrhage, or intratumoral fat content (4).

This tumor is usually found in the parasagittal and lateral aspects of the cerebral convexity, sphenoid wings, olfactory sulcus, and middle cranial fossa (1, 3, 4). Less commonly, they are located in the cerebellopontine angle, optic nerve sheath, choroid plexuses, and sella turcica (1, 3). In unusual cases, they may arise outside the dura mater with extracranial, cranial, or combined extensions, or rarely as entirely intraosseous lesions (1).

Extracranial meningiomas are extremely rare entities, accounting for less than 2% of all meningiomas, and are classified into two categories: primary and secondary extracranial meningiomas (5, 6).

Primary extracranial meningiomas are believed to arise from displaced embryonic arachnoid cells or multipotent mesenchymal cells, while secondary meningiomas arise as a direct extension of an intracranial mass (5).

Primary or ectopic extracranial meningiomas are typically found in the head and neck region, particularly in the sinonasal tract (paranasal sinuses, temporal bone, nasopharynx, and nasal cavity) (5). Up to one-third of meningiomas in this location destroy the skull base with secondary intracranial extension (4). Other anecdotal locations include the mediastinum, jaw, lungs, and skin (1, 6).

Ectopic meningiomas of the sinonasal tract, due to the coexistence of multiple tissues in this location, have a wide spectrum of differential diagnoses that include both benign and malignant neoplasms. Among the most common are epithelial tumors such as squamous cell carcinoma. In the group of neurogenic tumors, olfactory neuroblastoma and schwannoma stand out. Entities of vascular origin include nasoangiofibroma,

almost exclusive to the pediatric population, and finally, sinus polyps should also be considered (5, 7).

Secondary extracranial meningiomas arise as a direct extension of an intracranial mass (5). This extracranial extension accounts for up to 20% of cases and occurs through preformed bone pathways and foramina, mainly at the base of the skull. It predominantly involves the orbits, the external table, the retromaxillary, nasopharyngeal, retromandibular, and cervical regions, and less frequently, the external auditory canal (4-6).

Due to their slow growth, extracranial meningiomas are usually asymptomatic. However, when they reach a certain size, symptoms are related to the mass effect. Since the sinonasal tract is a common site for this pathology, patients often present with nasal obstruction, proptosis, epiphora, pain, and occasionally epistaxis, as well as neurological manifestations (7, 8).

The use of neuroimaging, such as CT and MRI, despite not revealing the inherent characteristics of a meningioma, is essential in the morphological evaluation of these lesions; the role of histopathology in diagnostic confirmation is noteworthy. Neuroimaging plays a crucial role in determining the size, extent, and involvement of adjacent structures, as well as in preoperative planning, since the main therapeutic approach is complete surgical resection, which has a low recurrence rate (5, 7).

## Conclusion

Extracranial meningiomas, especially those located in the paranasal sinuses, are a rare entity that can mimic other more common pathologies and, therefore, make initial diagnosis difficult. Although most are benign lesions, they can exhibit atypical characteristics and locally aggressive behavior. For this reason, complete surgical resection and histopathological analysis are essential to establish a definitive diagnosis and reduce the risk of recurrence.

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