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The use of an endoscopic endonasal approach for a secondary intraorbital meningioma: illustrative case

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BACKGROUND Meningiomas are the most frequent primary tumors in the central nervous system (CNS), but intraorbital location is uncommon and accounts for only 0.2% to 4% of all CNS meningiomas. Lesions in this compartment could be classified as primary, secondary, or ectopic. The close relationship with the optic nerve sheath is a landmark to identify the tumor as primary, whereas secondary tumors commonly come from an extension of an intracranial meningioma, and ectopic meningioma is a concept not yet completely established.

OBSERVATIONS The authors present a rare case of a secondary intraorbital meningioma operated through an endoscopic endonasal approach. Secondary meningiomas at the medial orbit are very uncommon, given their more common superior and lateral location as an extension of sphenoid meningiomas. The endoscopic endonasal route provides direct access to the medial orbit. The authors present an illustrative case of a meningioma located at the medial orbit and resected through an endoscopic endonasal approach that provided excellent visualization and anatomical exposure. Additionally, the authors review the concept and possible similarities between secondary and ectopic intraorbital meningiomas.

LESSONS An endoscopic endonasal approach should be considered as a feasible treatment option for intraorbital meningiomas, especially if they are in the medial orbital wall.

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KEYWORDS intraorbital meningioma; brain tumor; endoscopic endonasal approach

Meningiomas are the most frequent benign primary tumors in the central nervous system (CNS), accounting for approximately 36.6% of all cases.[1] They originate from the arachnoid cap cells and may be located anywhere in the CNS, including the extradural compartment.[2–4] An intraorbital site is uncommon and accounts for only 0.2% to 4% of all CNS meningiomas.[5,6]

Intraorbital meningiomas are classified as primary, secondary, or ectopic. Primary lesions account for 30% of intraorbital meningiomas and originate on the surface of the optic nerve sheath (ONS). Secondary lesions account for 70% of cases and usually extend intracranial lesions into the intraorbital compartment with intraosseous invasion.[7] Ectopic orbital meningiomas (EOMs) have no apparent relationship to the ONS, optic canal, or intracranial meninges, and its existence is controversial. Several case reports support the existence of EOMs,[4,7–17] whereas some authors believe that EOMs may originate from dura mater extensions, as previously demonstrated by the presence of dural tail to the lamina cribrosa region in a putative ectopic lesion.[18] However, the pathway used to reach the orbit ectopically remains unclear.

We report a case of meningioma with extension to three different compartments (medial orbit, ethmoid, and cribriform plate), and we review the concept of secondary and ectopic meningiomas and the feasibility of endoscopic endonasal access to superior, medial orbital lesions.

Illustrative Case

A 39-year-old female with no significant past medical history presented with progressive ocular asymmetry for 12 months with no

ABBREVIATIONS CNS = central nervous system; CT = computed tomography; EOM = ectopic orbital meningioma; MRI = magnetic resonance imaging; ONS = optic nerve sheath.

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diplopia. Static facial inspection revealed a discrete proptosis on the left side with a tendency to lateral ocular deviation. She was intact on neurological examination, with normal intrinsic and extrinsic ocular motricity and visual acuity (Fig. 1).

Magnetic resonance imaging (MRI) showed a lesion isointense on T1- and T2-weighted images and homogeneous contrast enhancement. The lesion was centered at the left medial orbit, measuring 2.6 × 2.3 × 2.0 cm (anterior × lateral × superior), with extension into the anterior ethmoid cells and the intracranial space through the cribriform plate. The lesion had a significant intraorbital/extraconal component displacing the medial rectus muscle laterally and the oblique and superior rectus muscle superiorly, with no involvement of the ONS and no signs of optic nerve compression (Fig. 1).

Given the patient's history and imaging features compatible with meningioma, surgical resection was offered, and two different approaches were discussed with the patient: transcranial access through a frontal craniotomy with superomedial orbitotomy and an endoscopic endonasal approach. The risks of olfaction loss were discussed, but, considering the less invasive procedure, the patient opted for the endonasal endoscopic surgery.

After general anesthesia was established, the patient was placed in the supine position with the back of the bed elevated above the heart level, and the right thigh was prepped for potential fascia lata and fat grafting. The head was secured in a Mayfield head holder and tilted 20° to the right.

Given the location of the lesion on the left cribriform plate and orbit, the exposure of the anterior skull base was predominantly on the left with wide sphenoethmoidectomy. On the contralateral side, only an anterior ethmoidectomy was performed. Opening of the frontal sinuses was achieved with a Draf III procedure due to tumor extension anterior to the anterior ethmoid artery (Fig. 2). A right-side nasoseptal flap was harvested and stored in the right rhinopharynx. The superior portion of the nasal septum attached to the cribriform plate was removed, and the anterior ethmoidal arteries were coagulated and divided prior to tumor dissection.

After broad exposure, it was possible to remove the portion of the lesion located inside the most anterior ethmoidal cells and to visualize the region of the anterior skull base adjacent to the cribriform plate. We then performed drilling of the lamina papyracea, which allowed access to the intraorbital portion of the tumor and exposure of the anterior skull base dura mater (Fig. 3). Despite the extraconal location, the violation of the periorbita by the tumor allowed orbital fat to herniate into the surgical field and made tumor dissection more challenging (Fig. 4). The use of surgical cottonoids allowed fat displacement and sharp tumor dissection, resulting in gross total resection of the orbital tumor component. The ONS was not visualized during the procedure, and it is possible to observe the area around the superior oblique muscle displaced at the end of the dissection (Fig. 4).

The dura mater was opened at the cribriform plate region with exposure of the tumor in the intracranial compartment. The lesion had a soft consistency with a clear dissection plane with the normal tissue. The whole lesion, including the involved dura mater, was resected. A multilayered strategy was used for skull base closure. An inlay fascia lata graft was placed covering the dural defect, followed by the nasoseptal flap that covered the anterior skull base dura and the exposed orbital content. The patient had an uneventful

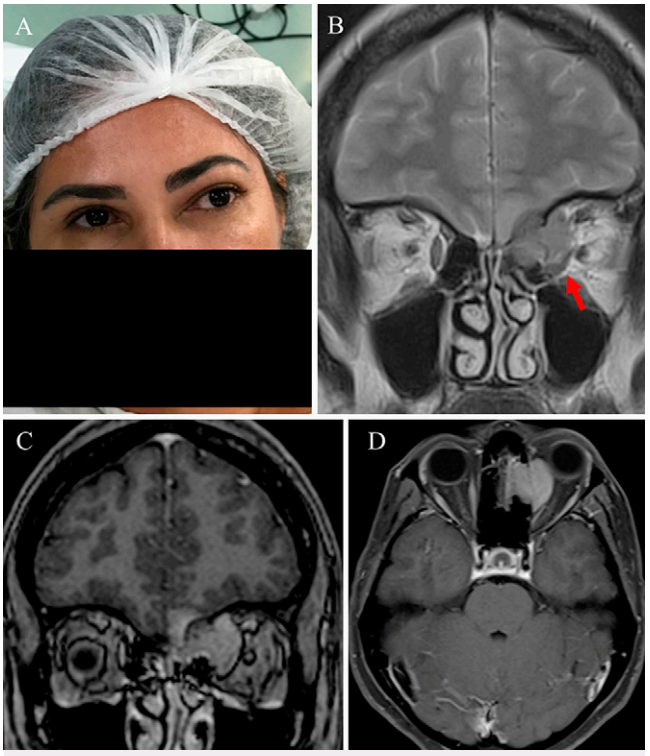

FIG. 1. Photograph (**A**) showing the patient's discrete proptosis on the left side with a tendency to lateral ocular deviation. T2-weighted MRI (**B**) showing an isointense intraorbital lesion with displacement of rectus medialis muscle (*red arrow*). Coronal (**C**) and axial (**D**) contrast-enhanced T1-weighted MRI shows homogeneous enhancement with an evident tricompartmental (anterior skull base, ethmoid, and orbit) lesion suggesting a diagnosis of meningioma.

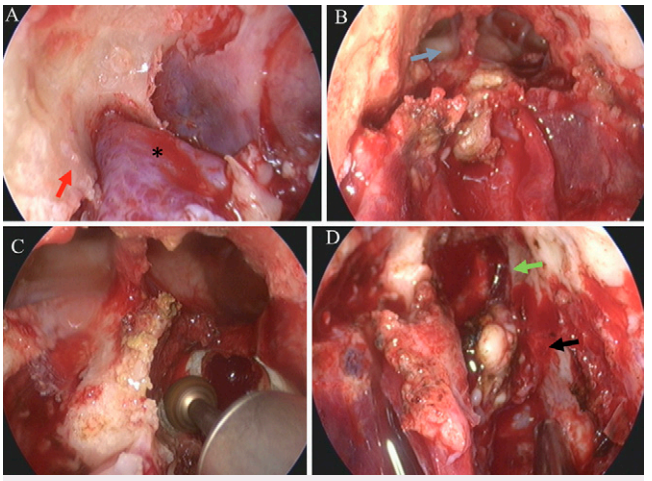

FIG. 2. Images showing the Draf III approach. Note the right frontal process of the maxilla (*red arrow*, **A**) and the septal mucosa (*asterisk*) before the frontal sinus opening. One can see the frontal sinus already opened (*blue arrow*, **B**) and after enlargement and wide frontal sinus exposure (**C**). After left anterior ethmoidal opening (*green arrow*, **D**), it is possible to identify part of the tumor that extends into the nasal compartment (*black arrow*).

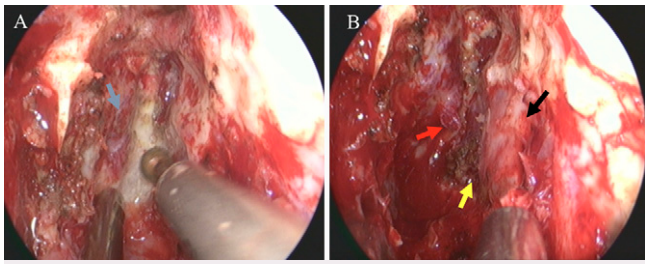

FIG. 3. After removal of the anterior ethmoidal cells and resection of the nasal tumor in the ethmoidal region, we drilled the anterior skull base to expose the dura mater of the anterior skull base (*blue arrow*, **A**). We opened the medial orbital wall to expose the intact periorbita (*black arrow*, **B**), so it is possible to visualize the spot where the tumor invades the orbit (*yellow arrow*) and a small rupture in the dura mater where the tumor invades the intracranial compartment (*red arrow*).

postoperative course with 1 day in the intensive care unit, and she was discharged home on postoperative day 4.

There were no complications in the postoperative period, and complete improvement of the left proptosis was noted at the 3-week follow-up (Fig. 5). The patient complained of anosmia, and the rest of her neurological examination remained intact with normal visual acuity and extrinsic ocular motricity. Six-month postoperative MRI confirmed complete tumor removal and contrast enhancement at the anterior skull base and medial wall of the orbit, compatible with good vascularization of the nasoseptal flap (Fig. 5).

Discussion

Observations

Intraorbital meningiomas are classified as primary, secondary, or ectopic. Primary lesions account for 30% of intraorbital meningiomas,

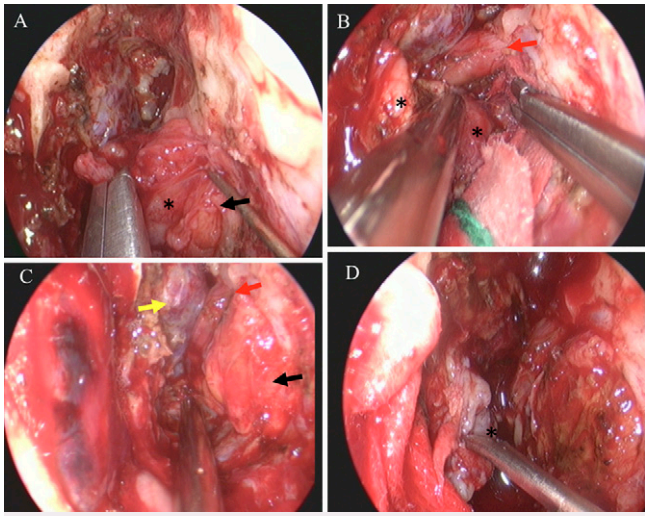

FIG. 4. After opening the periorbit, we started dissection between the tumor (*asterisk*, **A**) and the intraorbital fat tissue (*black arrow*). It is possible to see at the end of tumoral dissection its close relationship with the superior oblique muscle (*red arrow*, **B**). After complete removal of intraorbital tumor, we could see the orbital fat tissue, the superior oblique muscle, and the frontal basal dura mater (*yellow arrow*, **C**). After dural opening, we could access the intracranial compartment tumor (*asterisk*, **D**) with complete removal.

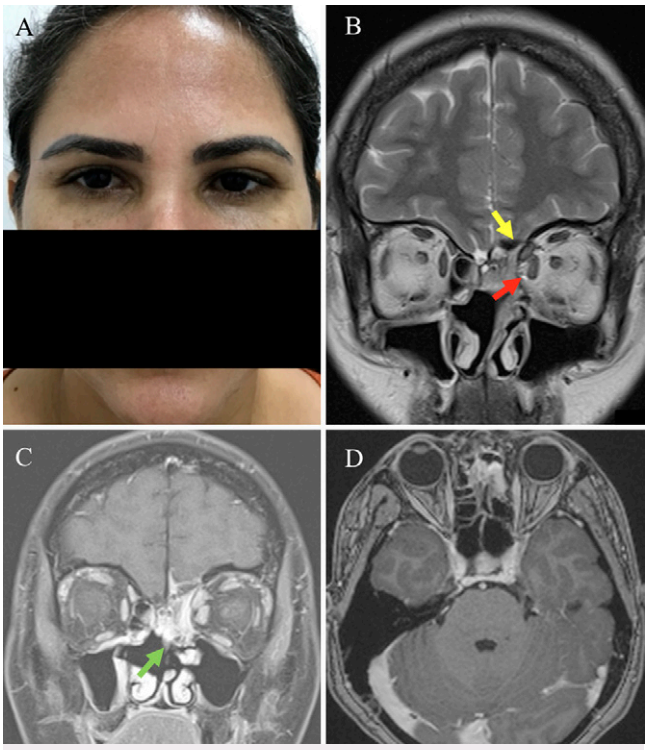

FIG. 5. Postoperative photograph (**A**) showing proptosis recovery. T2-weighted MRI scan (**B**) shows the normal position of the rectus medialis muscle (*red arrow*) and superior oblique muscle (*yellow arrow*). Coronal (**C**) and axial (**D**) contrast-enhanced T1-weighted MRI scans show the nasoseptal flap with homogeneous enhancement in the medial wall of the left orbit covering the anterior skull base (*green arrow*).

and their origin has a clear relationship with the ONS. Secondary lesions account for 70% of cases and usually extend intracranial lesions into the intraorbital compartment with intraosseous invasion.[7]

Given the absence of optimal management algorithms, observation of patients with vision stability is a viable alternative.[19] Conservative treatment is then reserved for patients without significant visual dysfunction or progression of visual loss, whereas radiotherapy and surgery, combined or not, are offered to patients with rapidly progressive visual loss.[20]

Secondary meningiomas account for the majority of orbital meningiomas. These meningiomas extend into the orbit via the perioraminal chiasmatic sulcus between the arachnoid of the ONS and the dura mater via the intracanalicular region, involve the optic nerve, and originate primarily from the planum sphenoidale region or the tuberculum sella.[21,22] In addition, they can emerge from the cavernous sinus, falciform ligament, clinoid region, sphenoid wing, frontoparietal area, or olfactory sulcus.[23] The growth pattern of meningiomas is typical, and their growth follows the pathway with the least resistance.[24]

EOMs have no apparent relationship to the ONS, optic canal, or intracranial meninges, and its existence is still controversial. In 2018, Huang et al.[8] showed that EOMs are extremely rare, with only 20 cases described in the literature. Although the authors further reinforced that the epidemiology of EOMs might be underestimated due to doubts regarding correct diagnosis, the authors also noticed that most of the lesions were in the superomedial portion of the orbit (55%).[8]

Huang et al.[8] observed that proptosis was present at diagnosis in up to 70% of the patients, and no patient had an initial complaint of visual loss. This fact reinforces the nonrelationship of ectopic and secondary meningiomas with ONS but cannot explain the origin of these tumors.

One of the oldest studies about this subject showed that the only orbital region with the presence of arachnoid cells would be the ONS, and, consequently, the EOM should originate from the ONS and then migrate to a region away from this structure.[25] Other theories include meningoceles that regressed throughout development[26] and possible meningeal tissue retained outside the CNS with a possible embryogenesis-related alteration.[4,11] Often there is asymmetry of the ipsilateral sinus, even when the sinus is not affected.[4,8]

In two reported cases, the expression of a protein is frequently found in meningeal cells of the lamina propria of the olfactory bulb (calponin),[26] raising the hypothesis that the tumor cells would reach the orbit through the frontoethmoidal sutures.[8] Furthermore, in 11 of the 20 reported patients, the lesions were adjacent to the frontoethmoidal suture, strengthening the theory of migration of cells capable of transforming into meningiomas. However, this information could change a large portion of ectopic meningiomas into secondary orbital meningiomas, because the origin of the tumor would be intracranial.

Diagnostic imaging should include computed tomography (CT) and contrast-enhanced MRI of the skull base and orbits. CT and MRI may reveal images with the classic meningioma characteristics, and there may be adjacent ipsilateral ethmoidal asymmetry.[8] In addition, reactive hyperostosis or pneumosinus dilatans may help in the differential diagnosis[7] and determination of the tumor site of origin.

Besides the importance of anatomical repair to endoscopic endonasal access, the neurovascular bundle (anterior ethmoid artery and nerve) may help in better understanding the origin of a meningioma classified as ectopic located in the medial portion of the orbit as actually being an orbital meningioma secondary to the growth of an intracranial meningioma through this neurovascular bundle into the orbit.

Thompson and Gyure[2] demonstrated, in their article about extracranial meningiomas of the nasal sinus tract, that arachnoid cells are present in the sheath of nerves and vessels exiting through the skull. Therefore, there is the possibility that the origin of meningiomas from undifferentiated multipotent mesenchymal cells, such as fibroblasts and Schwann cells, could explain the origin of ectopic meningiomas related to the anterior ethmoid neurovascular bundle. In addition, arachnoid cells may be present in the anterior ethmoidal nerve sheath, which, in association with its proximity to the orbital cavity, could suggest the migration of potential tumorigenic cells from the small nerve sheath into the orbit. This hypothesis may explain the origin of ectopic meningiomas of the medial portion of the orbit. However, the greater possibility is that these cases are secondary meningiomas originating from the intracranial compartment with migration and growth through the small nerve sheath.

Given the damaging potential of access to the intraorbital region, frequently requiring incisions in the face,[27] recent advances in endonasal endoscopic surgery are increasingly gaining acceptance as an operative route, and anatomical knowledge of them is essential for the planning and execution of complex endoscopic orbital surgeries. However, an exclusive endonasal endoscopic approach for the removal of intraorbital meningioma is an underreported topic.

Recently, Jeon et al.[28] showed the separate or combined use of superciliary transorbital access and endoscopic endonasal access for orbital tumors, allowing 360-degree access to the orbit without the need for orbitotomy and with low morbidity. However, in their sample, only two cases were meningiomas, one of which was a sphenoorbital meningioma and another of the olfactory groove, both considered secondary orbital meningiomas. In our patient, the evaluation of the preoperative MRI, given the location of the lesion, allowed us to realize that there could exist a correlation with the anterior ethmoid neurovascular bundle, which made the entire endonasal endoscopic route an excellent way to access the ethmoid, orbital, and intracranial portions of the lesion.

Benefits related to the endoscopic approach include no retraction of brain tissue, early devascularization of the tumor, and the possibility of obtaining complete resection of the lesion. Disadvantages include disruption of normal nasal sinus anatomy, damage to the olfactory apparatus, and risk of cerebrospinal fluid rhinorrhea.[29] Orgain et al.,[30] in a retrospective multi-institutional study, observed that all patients undergoing unilateral endoscopic endonasal surgery with total resection of meningioma of the olfactory groove reported some degree of olfactory preservation (75% characterizing the loss as mild or moderate). Youssef et al.[29] demonstrated the preservation of olfaction from transnasal endoscopic surgery in a patient with olfactory drop meningioma. This was possible because of a few elements: the unilateral origin of the tumor from the lamina cribriform, olfactory function on the side contralateral to the previously intact lesion, and preservation of the contralateral olfactory epithelium during the surgical procedure.

Lessons

We observed that for the diagnosis of a primary intraorbital meningioma, the relationship of the tumor with the ONS is essential. The differential diagnosis of a secondary or ectopic intraorbital meningioma is still controversial, but the presence of an intracranial tumor with intraorbital extension through the bone is highly suggestive of a secondary tumor, manly if the ONS is dislocated and intact. The origin and the real existence of EOMs remains unclear. The possible relationship between the migrations of arachnoid cells by neurovascular bundles through the orbit should be considered as an explanation for the concept of ectopic meningioma. Subsequent studies with larger samples may contribute to define it. However, it is highly probable for our case and others reported in the literature that the origin of the lesion is from the cribriform plate with a secondary growth toward the orbit region through the neurovascular bundle as a secondary meningioma, making the concept of ectopic meningioma less probable. The endoscopic endonasal approach for meningiomas of the medial orbit remains challenging, but we believe that, with an adequate preoperative anatomical study and surgical skills, this route could be less invasive with a good aesthetic result.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Guedes, Melo, Oliveira. Acquisition of data: Guedes, Júnior, Melo, Neto, Oliveira. Analysis and interpretation of data: Guedes, Júnior, Beer-Furlan, Oliveira. Drafting the article: Guedes, Júnior, Oliveira. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Guedes. Statistical analysis: Guedes, Oliveira. Administrative/technical/material support: Guedes, Júnior, Melo, Neto, Oliveira. Study supervision: Guedes, Beer-Furlan, Oliveira.

Supplemental Information

Previous Presentations

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