CASE REPORT
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Ethmoid Schwannoma: about the management of a rare tumor of sinonasal cavities manifested by an orbital complication

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ABSTRACT

We report a rare sinonasal tumor initially manifesting with orbital cellulitis with impaired oculomotricity and take stock of the management methods. Here, we present the case of a 72-year-old patient presenting to the ophthalmology emergency department with acute orbital cellulitis. Computed tomography revealed an ethmoidal mass associated with frontoethmoidal muco-pyocele. It was fully removed endoscopically, and a pathological diagnosis of ethmoid schwannoma was made. Magnetic resonance imaging after ten months showed no sign of recurrence. The patient presented with an acute orbital cellulitis with acute loss of visual acuity and impaired oculomotricity within 24 hours, associated with fever and local pain. Tomodensitometry showed a large heterogeneous tumor arising from the anterior ethmoid sinus and extending to the frontoethmoidal recess and the orbital cavity with lamination of lamina papyracea. The tumor was removed by endonasal radical excision. The final pathological analysis revealed benign schwannoma. There was no sign of recurrence or late complication during the 10-month follow-up. Schwannomas of the nasal cavities are rare tumors but must be mentioned among the differential diagnoses of endonasal and endosinusal masses, sometimes observed with orbital complications. On the basis of the location and extension, endoscopic resection is a valid strategy.

Keywords: Emergency, endoscopic surgery, ethmoid sinus, orbital cellulitis, schwannoma

Introduction

Schwannomas are benign tumors of nervous origin with a slow growth profile. A significate part (25%–45%) of them arise in the ENT area[1] but less than 4% of these tumors are sinonasal (1-3). We found very few case reports of a schwannoma arising from frontoethmoidal area (4, 5). This pathology peaks among individuals aged 30–40 years but can occur at any age (1-3, 6). The incidence of solitary schwannoma is similar in men and women and has no predilection for race (7).

Schwannomas originate from the Schwann cells in myelin sheaths. They mostly occur in the septum and maxillary and ethmoid sinuses in terms of sinonasal location (3, 8, 9). The presentation of a sinonasal schwannoma is similar to that of other benign nasal tumors. Schwann's sheaths primarily cover the nerve fibers of peripheral and sympathetic nervous systems. If the mass is located within the nasal cavity, a simple resection is often possible. In larger masses, a larger endoscopic resection may be necessary; however, an external approach is relatively rare (1, 2). The examination of a nasal schwannoma should include nasal endoscopy, sinus computed tomography (CT) scan, and magnetic resonance imaging (MRI) to assess its extent and guide the surgical approach for resection (1). In this report, we describe the case of an ethmoid schwannoma and review the current literature.

Case presentation

A 72-year-old man presented to the ophthalmology department of a tertiary referral hospital with rapidly progressive right orbital cellulitis over 24 hours, including ptosis and inferior chemosis. This was associated with watery eye, intense periorbital and frontal pain, limited oculomotricity, and diplopia in the peripheral gaze owing to the limitation in almost all fields of the gaze. Intraocular hypertension was measured at 36

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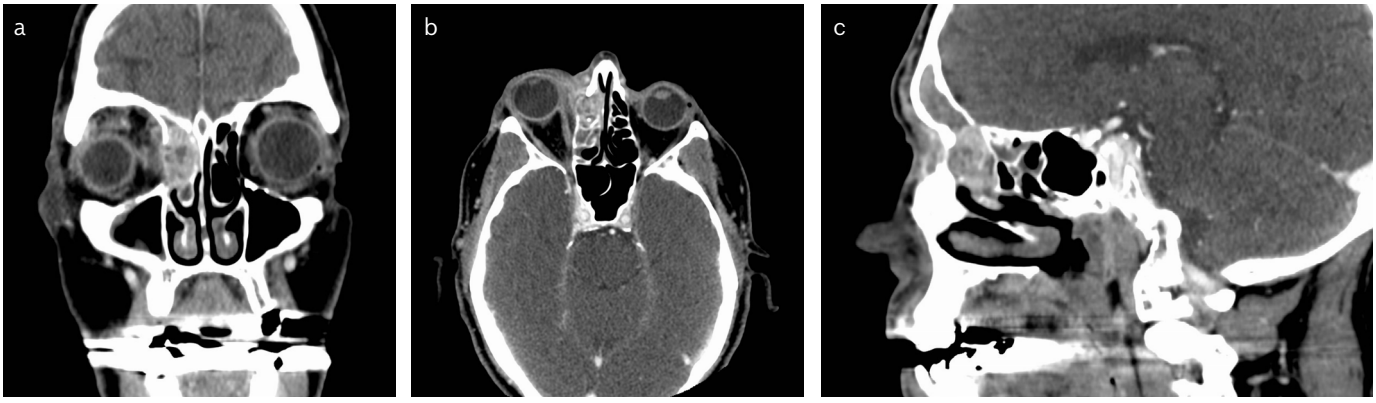

Figure 1. a-c. Emergency computed tomography scan showing the heterogeneous 16 × 17 × 20 mm right ethmoid-orbital mass, with exophthalmos and erosion of the lamina papyracea. Complete filling of the right frontal sinus and part of the ethmoid cells with an inflammatory material was noted (sections: (a) coronal, (b) axial, and (c) sagittal).

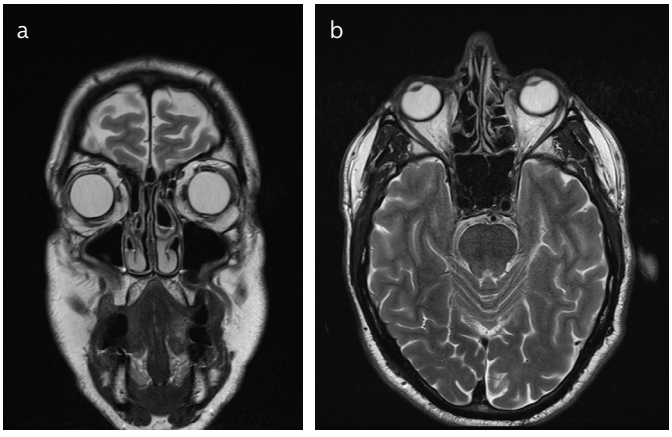

Figure 2. a, b. Four-month postoperative magnetic resonance imaging showing no residual tumor formation. Ethmoid cells and frontal sinuses were drained (sections: (a) coronal and (b) axial).

mmHg secondary to the mass effect necessitating immediate per-os acetazolamide. The patient was also suffering from fever of 38.5°C on admission and major biological inflammatory syndrome. He noticed no nasal or ophthalmological symptoms preceding this acute episode, except those of an allergic rhinitis he had been suffering from for years. He was being treated with levetiracetam for a right temporal complex partial focal epilepsy, without recurrence, for two years.

A same-day emergency computed tomodensitometry found a 16 × 17 × 20 mm right ethmoid-orbital mass, with exophthalmos and lamina papyracea erosion (Figures 1a-c). There was also complete filling of the right frontal sinus and part of the ethmoid cells with inflammatory material. The patient was, therefore, referred to our otolaryngology department for surgical management.

Main Points:

- Schwannomas arising from frontoethmoidal sinuses are rare.
- It should be considered in the differential diagnosis for sinonasal masses with orbital complications.
- Depending on the location and extent of the tumor, endonasal endoscopic excision could be a suitable management strategy.

Through an endoscopic approach, several polypoid masses were discovered on the right side, some of which had a fleshy appearance filling the middle meatus and extending down to the lower edge of the middle turbinate. Several biopsy specimens were sent, both fresh and formalin-fixed. Progressive excision of the mass filling the middle meatus allowed approach to the unciform process and enabled performing an antrostomy leading to healthy maxillary mucosa. The mass was then resected step by step as it had completely lysed the anterior ethmoid cells forming a vast open cavity up to the nasofrontal recess. Once the frontal cavity was opened, abundant pus spurted out, and bacteriological samples were taken. As the lamina papyracea was pushed laterally by the mass and appeared partially lysed, it was removed to examine the periorbital area, which was intact. The posterior ethmoidal cells were healthy. Unfortunately, we were not able to confirm the exact origin of the tumor endoscopically. The immediate postoperative stay was uneventful, and the patient was discharged home after three days of favorable biological and ophthalmological monitoring. Intraocular pressure and visual acuity were normalized after decompression. Postoperative care using daily saline douching, amoxicillin-clavulanate antibiotic, and methylprednisolone was prescribed. Endoscopic postoperative care was provided at regular intervals. An MRI at four months post-surgery found no residual tumor formation (Figure 2a, b).

Bacteriological samples found rare pansensitive Hemophilus influenzae. Frozen sections showed no signs of lymphoproliferative pathology or malignant neoplasm. Beneath the uninterrupted respiratory epithelium, pathological analysis of formalin-fixed sections showed a tumoral lesion consisting of spindle cells organized in a fascicular pattern. The cellular density was variable amongst the tumor imparting a biphasic appearance with a predominance of Antoni-A areas associated with occasional Antoni-B areas suggestive of the diagnosis of a schwannoma. No peripheral nerve was observed. There was no significant atypia or mitotic activity observed and no evidence of necrosis. Immunohistological analyses were performed on sections of fixed and paraffin-coated material using the Benchmark Ultra (Roche) automatons and anti-CD34 and S100 antibodies. Diffuse spindle cell immunopositivity for S100 and absence of CD34 and expression in the neoplastic cells were observed (Figures 3a, b). This profile was, thus, consistent with an ethmoidal schwannoma.

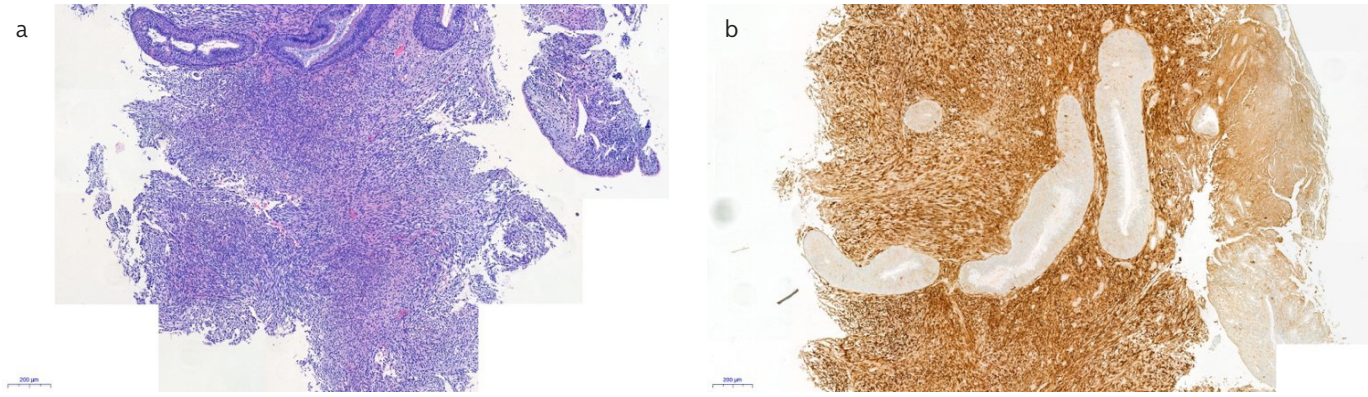

Figure 3. a, b. Hematoxylin and eosin, 5x. Spindle cell tumor with fascicular pattern and no significant atypia. Note the predominance of Antoni A pattern (a). mmunohistochemistry, 5x, showing diffuse positivity for protein S100 (b).

We are currently at more than eight months of follow-up without recurrence and without visual or functional nasal sequelae.

The patient gave oral informed consent for this case report.

Discussion

In this report, an ethmoid schwannoma with acute ophthalmologic presentation was completely resected endoscopically and confirmed pathologically. Schwannomas can develop almost anywhere in the body with the exception of the cranial optic and olfactory nerves because they lack sheaths containing Schwann cells (2, 3, 7). A little over 100 cases have been described and histologically confirmed in the literature since 1943 (3, 7).

The most frequently found benign tumors developing from nerve sheaths are schwannoma and neurofibroma (7). These tumors are accompanied by various signs and symptoms related to the anatomical site involved, the nerve of origin, or the compression of adjacent nerves. Despite their common origin, these lesions differ clinically and histologically (2). Schwannomas are predominantly isolated, encapsulated, and usually well-circumscribed lesions that are oval, round, or fusiform in shape. They contain hyaline blood vessels (7). These benign tumors do not tend to invade the axons, but rather repress them. This means that the nerve can theoretically be preserved surgically. A classical schwannoma typically has a histological pattern of Antoni A and Antoni B areas (3). Although there is a risk of malignant degeneration, this is rare (8%) and is found in longstanding lesions (2).

In the head and neck region, vestibular schwannomas, also called acoustic neuroma, are among the most common tumors (3, 7). Endonasal damage is quite rare and accounts for about 4% of them (2). In the nasal region, the ethmoidal sinus is most often involved, followed by the maxillary sinus (3, 9). Sinonasal schwannomas originate in the branches of the cranial nerves, such as the ophthalmic and maxillary branches of the trigeminal nerve or the autonomic nervous system, but their exact origin is often unclear in practice (3).

The most common symptoms are unspecific, such as nasal obstruction, epistaxis, hyposmia, and pain. Our patient did not have any of these before the acute event. Exophthalmos, facial swelling, and epiphora are less frequently observed but were present in our patient who experienced periorbital and palpebral edema. Orbital cellulitis with visual acuity reduction, alteration of color vision, visual field impairment, or with Gunn's sign need a rapid and aggressive management to prevent risks of severe complications such as cavernous sinus thrombosis, cranial neuropathies, meningitis, or death (4). Schwannomas localized in the sphenoid sinus can also result in paralysis of adjacent cranial nerves.[10,11] Nasal schwannomas generally have a benign clinical course, with possible loco-regional complications including intracranial extensions that have also been reported (5, 11, 12). Schwannomas have the potential for slow growth; therefore, the CT scan often finds a soft tissue mass with preservation of adjacent bone margins. This helps to differentiate schwannomas from the more aggressive malignant tumors that tend to destroy the adjacent bone. A mucocele is the most frequent complication but does not absorb the contrast on a CT scan (4, 8). MRI can, therefore, be useful to obtain information about the nature of the lesion and to detect damage to the base of the skull. An endonasal biopsy is also useful to confirm the diagnosis (1-3). Sometimes an angiogram may also be necessary to rule out an angiofibroma before a biopsy or surgical management (2).

Once a diagnosis has been established, treatment most often consists of a complete surgical resection with an approach that allows optimal exposure adapted to the location and extent of the tumor (2, 3, 7). In some patients, this can be carried out via an endonasal endoscopic surgery (1, 11). In others, surgery may involve various combinations of lateral rhinotomy, Caldwell-Luc procedure, or external fronto-ethmoidectomy (5, 12, 13). Skull base surgery may be necessary for schwannomas of the sphenoidal sinus. Finally, some extensive intracranial tumors require a craniotomy (5, 12). An attitude of watchful waiting can be considered in case of difficult accessibility and the absence of loco-regional complications, owing to its benign and slowly evolving nature. It also depends on the size of the lesion, and a pathological sample confirming the diagnosis must be the rule (2, 3).

The main differential diagnoses are those of unilateral tumor pathologies of the nasal cavities, such as carcinoma, inverted papilloma, sarcoma, neurofibroma, and meningioma, for tumors located near the skull base (3, 7).

Small biopsy fragments and fragmented samples, as in our case, can make the diagnosis more complex. In these patients,

an immunohistochemical analysis is essential to confirm the diagnosis (7). Schwannomas classically present biphasic histological profiles of Antoni A and B. This profile allows a correct diagnosis to be made and has no prognostic significance. In the nasal cavity and paranasal sinuses, the Antoni A profile is dominant (64.3%).

Second, the protein S-100, classically used to identify Schwann cells, is invariably positive in all schwannomas (7). In our patient, the discovery of a diffuse positivity for the S-100 protein in the tumor cells, therefore, strongly supported the diagnosis of a schwannoma.

We reported the management of a solitary schwannoma of the ethmoid sinus with acute orbital complication. The lesion showed a histological feature typical of a conventional schwannoma, including diffuse S-100 immunostaining. Schwannoma arising from the sinonasal tract are rare and should be considered in the differential diagnosis for sinonasal masses with orbital complication. Depending on the location and extent of the lesion, endoscopic endonasal resection is a valid management strategy.

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