

P. H. Eloy · M. C. Nollevaux · J. B. Watelet
J. P. Van Damme · S. T. Collet · B. Bertrand

Endonasal endoscopic resection of an ethmoidal solitary fibrous tumor

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Abstract Solitary fibrous tumor (SFT) is an uncommon spindle cell tumor that typically arises at the level of the pleura in adults. However, SFT has also been reported in various extrapleural sites including orbit, meninges, liver, lung, salivary glands, retroperitoneum, mediastinum. In the head and neck region, SFT has been documented in the external auditory canal, larynx, thyroid, sublingual gland, tongue, parapharyngeal space and the infratemporal fossa. The nose and the paranasal sinuses are a rare site for SFT with only 14 publications in the world literature. We present an additional case of a SFT arising at the level of the right ethmoid sinus successfully removed in one piece endoscopically and review the corresponding literature.

Keywords Solitary fibrous tumor · Ethmoid sinus · Endoscopic surgery · Spindle cell tumor

Introduction

Solitary fibrous tumor (SFT), formerly known as localized fibrous mesothelioma or benign fibrous mesothelioma [33] is an uncommon spindle cell tumor of a mesenchymal cell origin that typically arises at the level of the pleura during adulthood. However, SFT has also been documented in a large variety of extrapleural sites.

P. H. Eloy (✉) · J. P. Van Damme · S. T. Collet · B. Bertrand
Department of Otorhinolaryngology, Head and Neck surgery,
University Hospital of Mont-Godinne, Université Catholique
de Louvain, 5530 Yvoir, Belgium
E-mail: philippe.elay@orlo.ucl.ac.be
Fax: +32-81-423703

M. C. Nollevaux
Department of Histopathology, University Hospital
of Mont-Godinne, Université Catholique de Louvain,
5530 Yvoir, Belgium

J. B. Watelet
Department of Otorhinolaryngology, Head and Neck Surgery,
University Hospital of Gent, De Pintelaan, 185, Gent, Belgium

The nose and the paranasal sinuses are a rare location for SFT with only 14 publications in the world literature [1, 2, 6, 11, 14, 15, 17–20, 23, 30, 34, 35].

We present an additional case of a SFT arising from the right ethmoid sinus successfully and completely removed endoscopically in one piece and review the corresponding literature.

Case report

A 26-year-old woman was referred to the ENT department for surgery of an expanding process of the right nasal cavity. The patient was complaining of a progressive unilateral right-sided nasal obstruction that became bilateral and permanent with time. There was no history of bleeding, headache or ear symptoms. The anterior rhinoscopy revealed a solid and glistening tumor obstructing the nasal vestibule (Fig. 1). A computed tomography (CT) scan of the paranasal sinuses demonstrated an expanding process occupying the entire right nasal fossa from the nasal vestibule anteriorly to the nasopharynx posteriorly. The maxillary sinus was opaque with a lateral displacement and “erosion” of the medial wall (Fig. 2a). The tumor extended upwards to the right ethmoid sinus without breaching into the anterior cranial fossa. The nasal septum was deviated to the left by compression of the tumor. The left anterior ethmoid cells and the frontal sinus were opacified. A magnetic resonance imaging (MRI) showed a space-occupying lesion of the right nasal cavity with a hypointense signal on T₁-weighted images. On T₂-weighted images, the signal was hypointense and heterogeneous with areas of hyperintense signal (Fig. 2b). The mass showed a prominent and inhomogeneous enhancement after Gadolinium injection (Fig. 2c). There was retention of secretion in the right sphenoid and maxillary sinuses. A selective angiography demonstrated that the nutrient arterial supply came from the anterior ethmoidal artery. A biopsy of the tumor was performed under local anaesthesia. The first pathological hypothesis was a

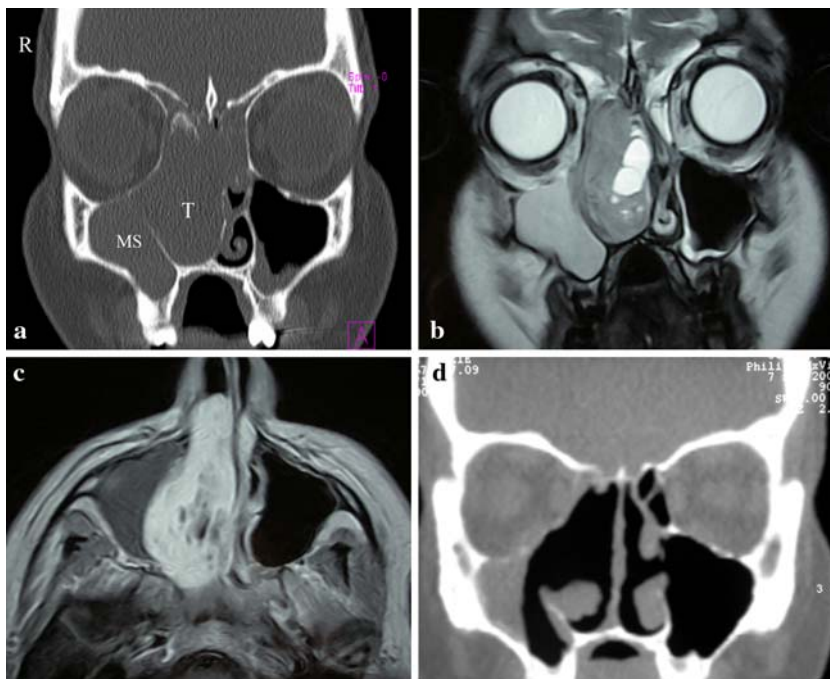


Fig. 1 Nasal endoscopy: large lobulated mass obstructing the right nasal vestibule

“hemangiopericytoma-like” tumor. The tumor was removed using an endoscopic endonasal approach that permitted an *en monobloc* removal of the lesion with concomitant opening of the different air cell compartments of the right ethmoid sinus and a middle turbinectomy. The mass was delivered through the oral cavity. A septoplasty was also performed during the same procedure.

Gross pathologic examination revealed a gray–white polypoid mass measuring 50×35×28 mm and presenting with a smooth and glistening surface. On cut section, the tumor showed a fleshy appearance, exhibiting sometimes more myxoid and microcystic areas.

Fig. 2 a Preoperative sinus CT scan: coronal view—total obstruction of the right nasal cavity by the tumor, lateral displacement and “erosion” of the intersinonasal wall, left sided nasal septum deviation, opacification of the left anterior ethmoid air cells. **b** Magnetic resonance imaging: T₂-weighted sequence—coronal cut. Large tumor filling completely the right nasal fossa. Secretion in the right maxillary sinus. Left-sided nasal septum deviation. **c** Magnetic Resonance imaging: axial cut—T₁-weighted image, tumor enhancement after gadolinium injection, hypointense signal in the right maxillary sinus full of mucoid secretion. **d** Postoperative CT scan: the right operated ethmoid sinus is free of disease. The right middle maxillary antrostomy is wide and patent. The nasal septum is straightened



Microscopically the tumor displayed a variable cellularity with zones of hypocellular fibrous stroma alternating with zones of hypercellularity constituted by a proliferation of bland-looking spindle cells haphazardly distributed within a collagenous stroma (Fig. 3a, b). The spindle cells showed an ovoid or round nucleus, without obvious cytological atypia, and a well-defined cytoplasm. The vascularity was prominent, with numerous thin, often dilated or branching vessels, sometimes surrounded by a densely population of more ovoid tumor cells, drawing a hemangiopericytoma-like pattern (Fig. 3b).

Immunophenotypically the tumor cells stained diffusely for vimentin (mesenchymal nature marker), CD 34 (endothelial cells marker) (Fig. 3c) and Bcl-2 (“anti-apoptotic” protein marker) (Fig. 3d) but not for S100 protein (indicative for Schwannian differentiation), cytokeratin (epithelial cells marker) and [alpha] smooth muscle actin (smooth muscle and myofibroblast marker). Based on the morphologic findings and the immunophenotypic results, the final diagnosis of SFT was made. The postoperative course was uneventful and the patient is still free of symptoms after 6 months. The postoperative CT scan demonstrated the absence of recurrence (Fig. 2d).

Discussion

Solitary fibrous tumor is a slow-growing tumor of mesenchymal origin that commonly arises from the pleura in adults [5, 7, 33]. The clinical behavior of pleural SFTs is unpredictable. Approximately 80–88% of pleural SFTs behaves in a benign fashion and is cured

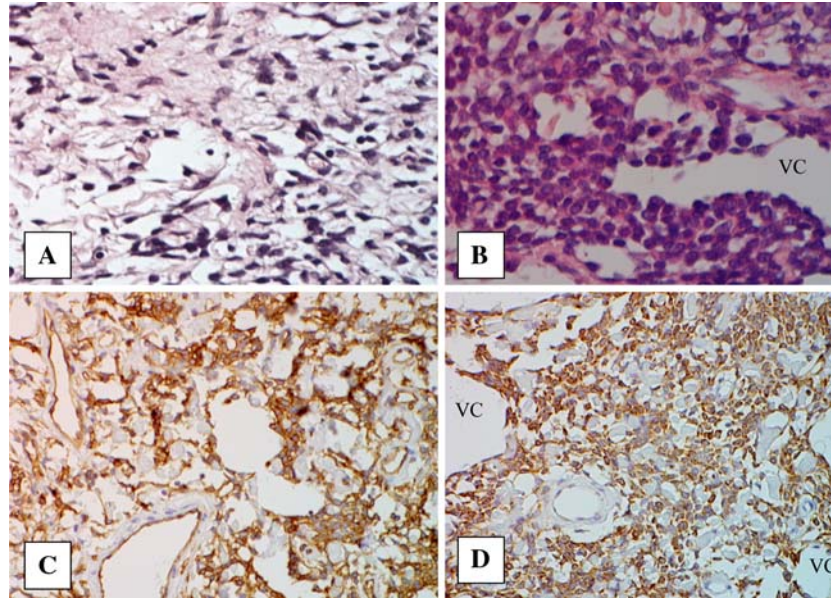


Fig. 3 a Histopathologic examination: light microscopy, hematoxylin–eosin (H and E) staining; high magnification: hypocellular area showing spindle cells with an ill-defined cytoplasm, scattered in a fibromyxoid background. **b** Histopathologic examination: light microscopy, hematoxylin–eosin (H and E) staining; high magnification: hypercellular area with more ovoid cells surrounding

branching vessels, thus creating a “hemangiopericytoma-like” pattern. **c** Histopathologic examination: light microscopy; high magnification: the vast majority of the tumor cells stained diffusely and strongly for CD34. **d** Histopathologic examination: light microscopy; high magnification: numerous tumor cells (mainly their cytoplasm) expressed the Bcl-2

with surgical excision. In contrast, 12–20% of the pleural SFTs are associated with invasion, recurrence and metastasis [7].

In the head and neck region, SFT has been documented in the external auditory canal [12], lachrymal sac [26], epiglottis [28], larynx [3], thyroid [23], sublingual gland [21], parotid gland [27], tongue [29, 32], lower gingival [10], orbits [25], parapharyngeal space [9], nasopharynx [8], hypoglossal nerve [4] and infratemporal fossa [24]. The nose and the paranasal sinuses are a rare site for SFT with only 14 publications in the world literature [1, 3, 6, 11, 14, 15, 17–20, 23, 30, 34, 35]. The disease occurs equally in men and women, ages ranged 40–60. The symptomatology is invariably dominated by nasal obstruction and sometimes by epistaxis. Nasal endoscopy disclosed a large expanding process that can be located either at the posterior aspect of the nasal cavity [30], the sphenoidal recess [1] or extends largely in the surrounding sinuses [6, 14, 17, 18, 34, 35]. Some cases exhibit extension into the anterior cranial fossa [11, 15].

From a clinical point of view the differential diagnoses for a SFT in the sinonasal cavity are numerous: epithelial neoplasms, esthesioneuroblastoma, meningioma, lymphoma, nasopharyngeal angiofibroma, hemangiopericytoma, schwannoma, fibromatosis, malignant fibrous histiocytoma and fibrosarcoma [15].

SFTs show some characteristic radiologic findings such as: a well-circumscribed solid mass hypo to isointense to muscle on T₁-weighted images, heterogeneous hypo or hyperintense on T₂-weighted images and a

prominent and heterogeneous contrast enhancement [13, 15, 16].

These findings correspond well to the histologic findings of a fibrous stroma intermixed with prominent vascular channels [15, 16].

The definitive diagnosis of SFT is based on the histopathological examination. On gross examination, SFTs of the nasal cavity and paranasal sinuses are well-circumscribed, often polypoid masses, with a smooth external surface and a pale appearance on cut section. On permanent sections of formalin-fixed tissue, the microscopic findings in SFT typically reveal a proliferation of ovoid–spindle-shaped cells randomly distributed, along a “patternless–pattern”, within a collagenous, sometimes keloid-like, stroma of variable vascularity. The tumor cells present with a faint eosinophilic cytoplasm and exhibit nuclei with evenly distributed chromatin and small nucleoli [5, 15]. The cells are randomly distributed in a collagenous background of variable vascularity and cellularity [13]. Often a portion of the tumor has prominent branching vessels and a hemangiopericytoma-like pattern [13, 15, 17, 35]. This was present in our case leading to confusion with the diagnosis of hemangiopericytoma.

The histopathologic features of SFTs may also be confused with those of a schwannoma, fibrous histiocytoma, low-grade fibrosarcoma or a nasopharyngeal angiofibroma. A broad panel of immunostains is therefore useful for differentiating these neoplasms. SFTs are strongly positive for CD34 and vimentin and uniformly negative for keratin, desmin and S100 protein. CD34

immunoreactivity can be considered a specific marker for SFTs and is noted for its absence in other epithelial and spinal tumors [31]. Although not specific of SFTs, the adjunction of Bcl-2 in the panel of immunostains may help to distinguish between the different entities, as Bcl-2 is frequently positive in SFTs but not, for example, in hemangiopericytoma.

Surgery is the first-line of treatment. Complete en bloc removal is the aim of the treatment. For tumor localized in ethmoid, an endonasal endoscopic surgery is the treatment option [22] if there is no invasion of the anterior cranial fossa and no extension to the infratemporal fossa. Endoscopy provides a wide and mobile panoramic view, good visualisation and magnification, angled vision and illumination [3, 23]. Compared to the traditional external approaches, this type of treatment also has the advantages of no external incision, less blood loss, low postoperative morbidity and shorter hospital stay (1 or 2 days postoperatively). Yet, this kind of surgery should be reserved to experienced surgeons in endoscopic surgery particularly in case of large tumor because of the trends for bleeding [18].

Conclusion

Sinonasal locations of a SFT are rare. The clinical manifestation is an apparent benign tumor with an inclination to local aggressiveness. Diagnosis should be considered in the differential diagnosis of all expanding process in the nasal and paranasal cavities and spindle-cell lesions. The diagnosis of these tumors is based on the histopathologic findings of two fundamental architectural features present in various degrees: the solid spindle type and the diffuse sclerosing type. Immunohistochemical analysis (CD 34 and vimentin positivity) are essential to make the definitive diagnosis. Local excision is the first line of treatment. Endoscopic treatment of ethmoidal solitary tumor is reliable [1] if the surgeon has experience in endoscopic sinus surgery [2], if the tumor has no major extension to the brain or the infratemporal fossa and [3] taking into account the trend for bleeding of such a tumor. Yet a long follow-up is recommended because of the unpredictable behavior.

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