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SOLITARY FIBROUS TUMOR OF THE SINONASAL TRACT: A RARE CASE INVOLVED THE NASAL SEPTUM

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ABSTRACT

Solitary fibrous tumor (STF) is a mesenchymal neoplasm with fibroblastic differentiation that usually develops in the pleura, but can also affect extrapleural sites. Sinonasal localization is very rare. A 60-year-old man complained right-sided nasal obstruction and bilateral epiphora for 5 years. Based on MRI and CT, the tumor was surgically removed. Final histology examination evidenced diffuse spindle-shaped cells proliferation and less cellular areas with collagenized stroma. The immunohistochemistry showed positivity for STAT6, vimentin, CD34 and CD99. The definite diagnosis was Solitary Fibrous Tumor.

INTRODUCTION

Solitary fibrous tumor (STF) is a spindle cell mesenchymal neoplasm with fibroblastic differentiation that usually develops in the pleura, but can also affect extrapleural sites [1]: in 6% of cases it involves the head and neck region [2]. Sinonasal localization is very rare where it can lead to nonspecific symptoms such as unilateral nasal obstruction. It has a slight prevalence for the male sex compared to the female sex (M:F 5:4). The etiology is unknown although the possible pathogenesis lies in the NAB2-STAT6 gene fusion [3,4].
We describe the case of a tumor with implantation at the level of the right nasal septum, which on preoperative biopsy was classified as an anthrochoanal polyp.
Thanks to CT and MRI study, final histological examination, and positivity for vimentin, CD34, and CD99 on immunohistochemical examination, we were able to reach the correct diagnosis of solitary fibrous tumor.
At 24 months, the patient is free of disease.

CASE DESCRIPTION

A 60-year-old Caucasian man presented at our ENT unit reporting a symptomatology of right-sided nasal obstruction, anosmia, rhinorrhea and bilateral epiphora for 5 years.

His past medical history was positive for cancer: he referred previous surgery for skin melanoma. Physical examination revealed a large, lobulated, hard tumor originating from the nasal septum of the right nasal cavity, extending from the nasal vestibule to the ipsilateral choana (Fig 1).

Computed Tomography (CT) scans showed a oval mass, with lobulated margins, parenchymatous density that displaces the nasal septum to the left, the right nasal wall to the right, the right ethmoid cells superiorly, the unciform process and the right lamina papyracea superoexternally, with which it is in continuity (Fig. 2).
The tumor appeared on Magnetic Resonance (MR) with uneven intensity - mainly medium-low with hypointense and hyperintense components - in T1 and - mainly low with hyperintense components - in T2 and STIR.
A preoperative biopsy under local anesthesia was performed, which identified the mass as an anthro-coanal polyp.

The most important differential diagnosis was made with a metastasis/recurrence of melanoma because of the patient's past history. As reported in the literature, especially melanoma metastases can lose melanocyte lineage-specific markers, presenting unusual morphology and immunohistochemical features similar to SFT, emulating the same. PCR study of mutations for BRAF, NRAS, KIT, GNAQ and GNA11, which confirms the diagnosis of recurrent/metastatic melanoma, is mandatory to recognize the two forms [5].

The patient underwent surgery under general anesthesia for endoscopic exeresis of the tumor. The insertion at the level of the nasal septum was identified after dissection of the mass with a plasma blade, and then a subperoisteal removal of the septal mucosa was performed. During the surgical procedure, it was necessary to causticate the septal branch of the sphenopalatine artery. Finally, uncinectomy and anstostomy media were packed.

Final histology examination evidenced diffuse spindle-shaped cells proliferation and less cellular areas with collagenized stroma, moderate polymorphism, low mitotic index (less than 4 mitoses per 10 HPF).
The immunohistochemistry showed positivity for STAT6, vimentin, CD34 and CD99.
The definite diagnosis was Solitary Fibrous Tumor with free resection margins.
The patient underwent new examination at 3, 6, 12 and 24 months remaining free of disease (Fig. 3).

DISCUSSION

Solitary fibrous tumors are submesothelial, fibroblast-like mesenchymal tumors that typically develop in the parietal or visceral pleura [1]. However, they can be found in extrapleural site: the head-neck district is involved in only 6% of cases [2]. The peak incidence is between the 5th to 6th decade of life with a slight prevalence for the male sex compared to the female sex. In most cases it is asymptomatic or may give signs of itself by causing unilateral nasal obstruction when it totally engages the nasal fossa. MRI shows a hypo- or isointense mass at T1 weighing and hypo- or hyperintense at T2 weighing, with heterogeneous enhancement after contrast [6].
Immunohistochemical examination shows positivity for STAT-6, CD34, CD99, BCL-2 [7].
Criteria for classifying the mass as malignant have been defined by WHO: hypercellularity, increased mitoses (>4 mitoses per 10 high-power fields), cytologic atypia, tumor necrosis, and/or infiltrating margins [8,9].

STF of the sinonasal tract is very rare: only 9 cases have been described in the literature. The case history, considering those with an implantation base at the level of the nasal septum, is further reduced.

In each of them, unilateral nasal obstruction, similarly to our case, was the only sign that led the patient to a specialist examination. The diagnostic procedure consisted of endoscopic examination, imaging tests (CT and/or MRI), and preoperative biopsy. The treatment of choice was endoscopic surgery. If the mass is highly vascularized, exeresis may be preceded by embolization of the mass [10].

In our case, surgery without chemo-radiotherapy was the treatment of choice because the mass did not meet WHO histologic criteria as it was characterized by hypocellularity, low mitotic number (<4 mitoses per 10 high-power fields), absence of cytologic atypia, tumor necrosis, and/or infiltrating margins.
Specifically, endoscopic resection of the mass with excision of its attachment to the mucoperichondrium with cartilage was performed.
The study by Sireci et al., in this regard, retrospectively analyzed the database of two University Hospitals (Genoa and Palermo) on the type of treatment given to sinonasal tumors from 2012 to 2020. A cohort of 32 patients with nasal septal tumors was identified: 28 (87.5%) cases were benign neoplasms and four (12.5%) cases were malignant tumors.
The surgical approach chosen, for benign tumors, was resection of the mucocondrium/periosteum; in contrast, malignant tumors were treated with resection of all layers of the nasal septum [11].
In the case described, we preferred the transnasal endoscopic approach and the "disassembly technique," which consists of oriented anatomic decomposition of the mass to precisely locate the implant base [12].
The plasma blade was used for the dissection of the mass because, as described in the literature, the CO2 laser can produce high thermal energy that can damage the tissue, distorting the pathological examination.

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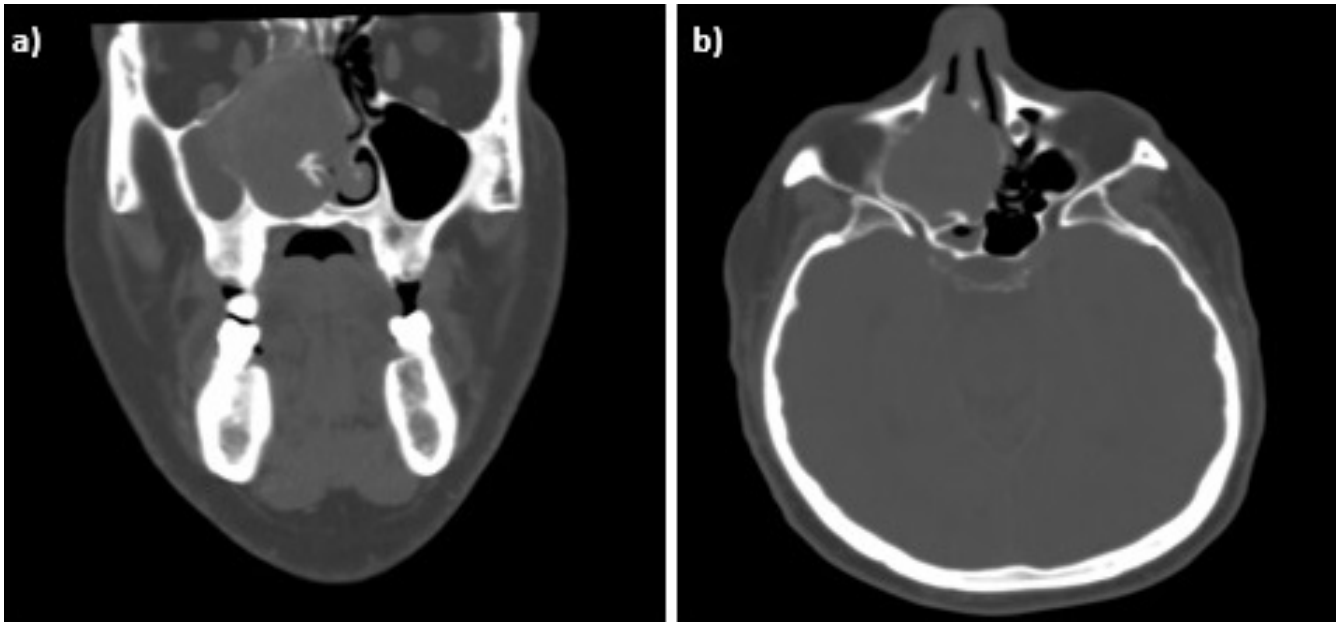

Fig.1 – CT image of the SFT in coronal section (a) and axial section (b)

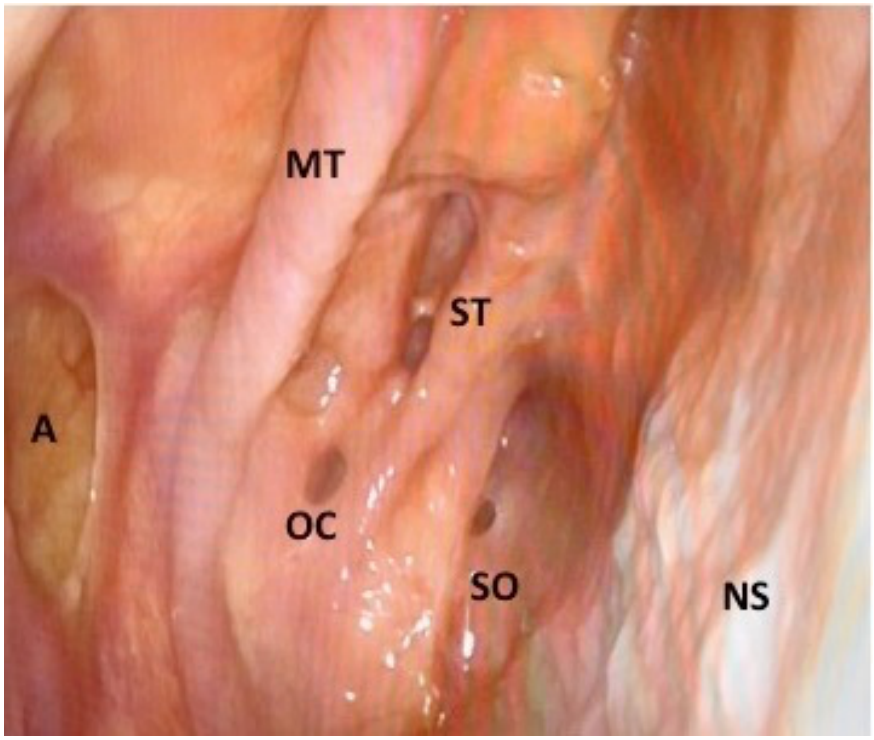

Fig.2 – Control after 2 years. Absence of recurrence in the nasal nostril. A, Antrostomia; MT, Middle Turbinate; OC, Onodi Cell; Superior Turbinate; SO, Sphenoid Ostium; NS, Nasal Septum.