



Case report

Solitary Fibrous tumor of Nasal cavity: A case report and review of literature

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ABSTRACT

Introduction and importance: Solitary fibrous tumors (SFTs) involving the nasal cavity are extremely rare with few cases reported in the literature.

Case presentation: We present a case of SFT in a 90 year-old male complaining of a slow-growing mass prolapsing through left nostril. Nasal endoscopy and imaging exams revealed a mass occupying the entire left nasal cavity, pushing the nasal septum to the opposite side and extending up to the nasopharynx. Biopsy specimen examination reported sarcoma. The patient underwent complete surgical resection of the mass through left para-latero-nasal approach. Immuno-histochemical analyses confirmed the diagnosis of SFT. The patient has remained free of tumor 2 years after surgery.

Clinical discussion: Clinical and imaging features of SFTs of nasal cavity are not specific. A broad of differential diagnosis is associated with histopathologic features of SFTs. Therefore, immuno-histochemical analyses are crucial to confirm the diagnosis. Complete resection of the mass with clear margins is mandatory to minimize local recurrence.

Conclusion: SFTs of nasal cavity are very rare neoplasms which continue to pose challenges to practitioner. Pathological examination and mainly immunohistochemical studies are important to establish the diagnosis. Complete resection of the tumor is the key for good outcome.

1. Introduction

Solitary fibrous tumors (SFTs) are rare neoplasms which typically occur in the pleura [1]. However, because of their mesenchymal origin, they have been reported to derive from many different anatomic locations such as the urogenital system, lungs, liver, orbit, thyroid, larynx and salivary glands [2,3]. SFTs involving the nasal cavity are extremely rare. To our knowledge, only about 40 cases have been reported in the English literature [4,5]. Clinical, imaging features of SFTs are not specific and they have a broad of differential diagnosis. Herein, we share our experience on the presentation of SFT in a 90 year-old male, mistaken for sarcoma, and we characterize pathologic features and management strategy of this rare entity. This case report has been reported in line with the SCARE criteria [6].

2. Presentation of case

A 90 year-old male was referred for otolaryngologic evaluation with a mass in the left nasal cavity which was present for over 4 months with gradual progression. There was no medical history, particularly, no previous sinus disease or sinus surgery. On clinical examination, there was a voluminous mass prolapsing through the left nostril (Fig. 1). The latter was fully obstructed with a deflection of the nasal septum to the right side. There were no palpable cervical lymph nodes, facial deformities, ocular signs or neurological disorders.

Nasal endoscopy through the right side, revealed a smooth-textured and grayish mass prolapsing through the left choana into the nasopharyngeal space. The mass pushed the nasal septum toward the right side.

A computed tomography (CT) scan showed a large, homogeneous mass occupying the entire left nasal cavity. The mass extended up to the nasopharynx and displaced the medial wall of the left maxillary sinus (Fig. 2A). A destruction of the nasal septum and the left lamina

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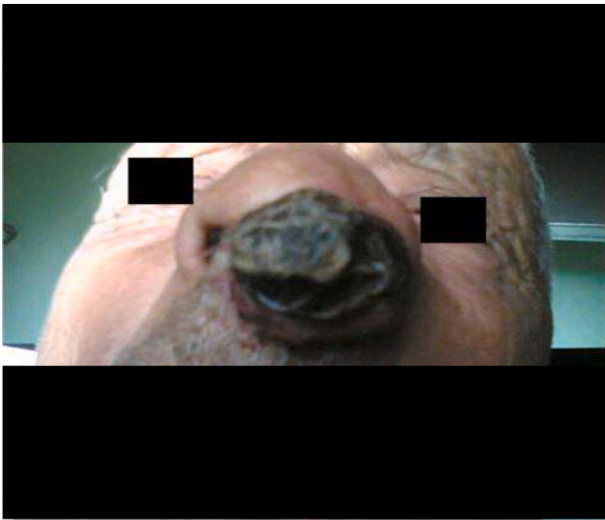


Fig. 1. Huge mass prolapsing through left nostril.

papyracea has been noticed with and a complete opacification of the left maxillary sinus (Fig. 2B).

Examination of the biopsy specimen reported malignant tumor compatible with sarcoma. A chest and abdominal CT scan and bone scan were performed and revealed no abnormalities.

The patient underwent surgical resection through left para-latero-nasal approach. Intraoperative findings showed a soft to firm vascular mass originating from the middle turbinate head, filling the left nasal cavity, extending posteriorly up to the choana and pushing the nasal septum to the opposite side. There was a fluid retention of the left maxillary sinus with no tumor involvement. A total en bloc resection of the mass was performed and hemostasis was achieved. The specimen was sent for histopathological examination.

Microscopic examination revealed spindle-shaped cells dispersed within a collagen-rich stroma with numerous thick-walled vessels and ectatic vascular areas. There were no features of mitoses or polymorphism. Immuno-histochemical staining was positive for CD-34 and bcl-2, and negative for S100 protein, desmin and actin. Thus, the diagnosis of SFT was established (Fig. 3).

The patient has been followed up regularly by endoscopic surveillance and CT scan controls with no signs of recurrence 2 years after surgery.

3. Discussion

SFTs were first considered as mesothelial neoplasms. On the basis of ultra-structural and immuno-histochemical studies, they have been shown to arise from mesenchymal fibroblast-like cells. Mesenchymal origin of SFTs was further supported by the description of extrapleural locations [2]. SFTs of the head and neck are uncommon. They have been documented in external auditory canal, lacrimal sac, epiglottis, larynx, nasopharynx, thyroid, sublingual gland, parotid gland, and tongue [7]. Those of nasal cavity and paranasal sinuses are extremely rare [4,8].

According to the literature, there are no specific symptoms. The most common clinical one was progressive nasal obstruction. The other reported symptoms were epistaxis, rhinorrhoea, anosmia, headache, facial pain, exophthalmos and visual disorders due to compression of the orbit [9,10]. Our patient presented a slow-growing mass prolapsing from the left nostril. The endoscopic appearance of SFTs was that of smooth, oval-shaped or circular encapsulated, red and fibrous unilateral nasal masses [7].

On unenhanced CT, SFT appears as homogeneous iso-attenuated mass occupying nasal cavity, with occasional internal calcifications. Marked enhancement after the administration of contrast material is generally noted due to their high vascularity. Depending on the size, the nasal septum may be deviated with bone structures remodeling, local absorption and even reactive sclerosis [9]. Rarely can the tumor extend to the orbit and cranial cavity, through the cribriform plate and ethmoid roof [2]. In our case, an erosion of the left lamina papyracea the nasal septum has been noticed. On MRI, SFTs are usually homogeneously isointense to gray matter on T1-weighted images and generally appear heterogeneously isointense or hypointense on T2-weighted images. Predominant low signal on T2-weighted images is a characteristic feature of those tumors, but not specific [9]. Nevertheless, low signal on T2 with the enhancement after gadolinium injection is highly predictive of SFT [11,12]. Unfortunately, MRI was not performed in our case because of a lack of accessibility.

Based on the variety of clinical presentations and the absence of specific imaging features, the clinical differential diagnosis of nasal cavity SFTs should be, essentially, made with fibrosarcoma, hemangiopericytoma and nasopharyngeal carcinoma [5].

Biopsy and pathological examination are necessary to establish the diagnosis of SFT. Microscopically, the tumor is composed of spindle cells dispersed within a collagenous stroma. The presence of areas of hyalinization adjacent to collagen deposits is characteristic of SFTs. The tumors are highly vascularized which may result in some confusion between SFT and hemangiopericytoma [2,4]. Some tumors can appear similar to SFTs, such as nasopharyngeal angiofibroma, angioleiomyoma,



Fig. 2. A. Axial CT scan showing homogenous large mass completely filling the left nasal cavity and extending up to the nasopharynx (star). B. Coronal CT scan showing a mass occupying the left nasal cavity. The mass pushed the medial wall of left maxillary sinus and caused erosion of the left lamina papyracea (white arrow) and the nasal septum (red arrow). There was a complete opacification of the left maxillary sinus. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

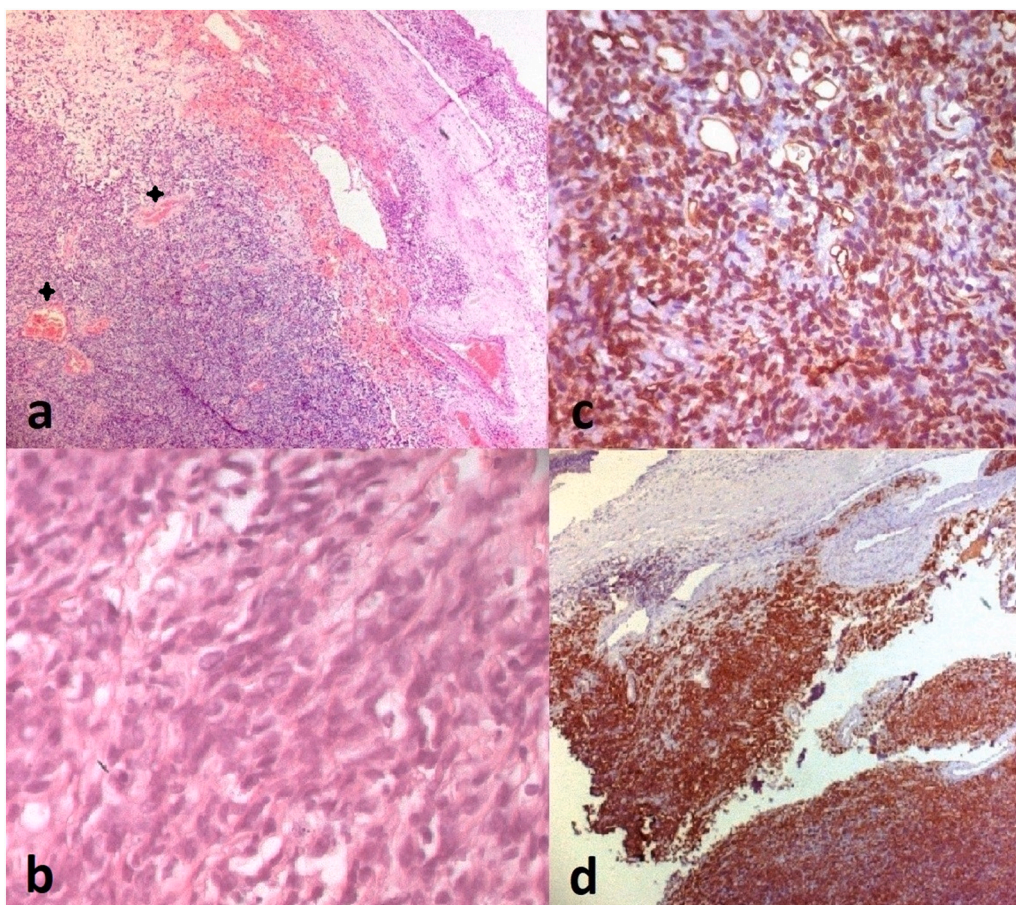


Fig. 3. Microscopic examination showing fusiform cells within a dense collagen stroma with areas of hyalinization (a: HE $\times 40$, b: HE $\times 400$). The tumor is highly vascularized (star). IHC showed positive staining for CD34 (c) and bcl-2 (d).

nerve sheath tumors like schwannoma and neurofibroma [5]. In our reported patient, SFT was mistaken for sarcoma because the latter is composed of spindle cells.

Due to the broad differential diagnosis associated with their histopathologic features, immuno-histochemical studies are mandatory to confirm the diagnosis of SFTs. CD34 and bcl-2 are the first-line markers for the diagnosis of these tumors; a tumor negative for CD34 and bcl-2 is unlikely to be SFT [5,9]. Unfortunately, CD34 is not entirely specific of SFTs and express in a variety of spindle cell neoplasms such as dermatofibrosarcoma protuberans and neural tumors [2]. In addition, SFTs are strongly positive for vimentin and uniformly negative for actin, keratin, desmin, and S100 protein. These findings may help to exclude some lesions, like epithelial tumor, hemangiopericytoma, fibrosarcoma, and neurogenic tumor [9]. In our case, immuno-histochemical analyses were strongly positive for CD 34 and negative for S100 protein, desmin and actin. These findings supported the diagnosis of SFT. Some microscopical features are considered to be typical for malignant SFTs, such as nuclear atypia, increased cellularity, necrosis and more than 4 mitoses per 10 HPF [2,5]. In our case, no malignant features have been observed. It has been reported that morphologic features cannot, necessarily, predict malignant behavior of SFTs. In fact, recurrences and metastases have been well documented in SFTs lacking atypical histologic features [5].

Complete surgical removal remains the cornerstone of the treatment of SFTs. The surgical approach depends on tumor size, extension and aggressiveness. In fact, tumors larger than 10 cm, the presence of histologic malignant component and positive margins are associated with a high risk of local recurrence [5]. Therefore, surgical approach should allow complete resection of the tumor with clear margins. The different

used approaches are medial maxillectomy, lateral rhinotomy, external ethmoidectomy, sphenoidectomy and surgery via the transfacial approach [13,14]. Our patient was operated on through a left paralaro-nasal approach.

In some case reports, endoscopic resection has been reported as suitable approach for the management of SFTs. Indeed, it permits a good visualization and magnification. However, sizable and/or bleeding tumors may obscure visualization leading to incomplete resection [7]. Therefore, endoscopic resection of SFTs should be considered when the size allows an adequate visualization and the surgeon has experience to tackle potentially bleeding endoscopically. In our case, endoscopic treatment was not considered because of the tumor size and the lack of experience of the surgeon. Adjuvant therapy, such as radiotherapy or chemotherapy, is not recommended because of the low risk of recurrence which is usually non-malignant [7].

In general, SFTs are associated with a good prognosis. Complete resection of SFTs is the most important factor in determining long term outcome [7]. Close and long term follow-up is mandatory because of unpredictable behavior. The surveillance is based on clinical examination, endoscopy and Imaging, particularly MRI.

Management of recurrence remains unclear. Surgical excision, if possible, with adjuvant radiotherapy, can be an alternative. However, there is a lack of data regarding long term efficacy of radiotherapy [15].

4. Conclusion

SFTs of nasal cavity are very rare neoplasms which continue to pose challenges to practitioners. Indeed, clinical and imaging features are not specific. A broad of differential diagnosis is associated with

histopathologic features. Therefore, immuno-histochemical studies are crucial to establish the diagnosis as in our case. Complete resection of the tumor with clear margins is the main factor to improve the outcome after surgery with long term surveillance.

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Ethical approval

This is a case report study and ethical approval not required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Mayssa Khribi, Kermani Wassim: conception and design of the study.

Abir Meherzi, Badreddine Sriha: acquisition of data.

Mayssa Khribi: article drafting.

Wassim Kermani, Mouna Bellakhdhar, Mohamed Abdelkefi: final approval of the version to be submitted.

Research registration

Not applicable.

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Declaration of competing interest

None declared.

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