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Case Report

Periorbital and Nasal Solitary Fibrous Tumor

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Abstract

The nasal solitary fibrous tumor is an infrequent benign unilateral tumor, were originally first described as a rare mesenchymal neoplasm that commonly involves the pleura, originating from spindle cells, described in 1931 by Klemperer and Rabin. It is further sub-classified as a benign type of mesothelial tumor; it has been reported in other regions such as: the orbit, nasopharynx, thyroid gland, parapharyngeal space, tongue, major salivary glands, lung, mediastinum, extremities, the liver also rarely affect the nasal cavity and paranasal sinus.

The diagnosis is primarily histological, it consists in a positive immunohistochemical finding of CD34.

Author report a case of 36 years-old man presented with a 3-month history of persistent progressive left nasal bleeding, left nose obstruction and left eye pain is reported. Physical assessment and nasal endoscopy revealed a pain small rubbery mass palpable in the superonasal quadrant of the left eye and the tumor arose from the left ethmoid sinus and extended to the left nasal cavity and the periorbital area. Further imaging by CT and MRI disclosed a large left-sided nasal and periorbital cavity homogeneous mass, occupying left maxillary sinus, left ethmoid, extending to the left frontal sinus and left orbit. The tumor successfully was removed by endoscopy sinus surgery without complication. The tumor had spindle-shaped cells and was positive for CD34, CD99, Bcl-2 and desmin, histopathological analysis of the specimen was consistent with nasal solitary fibrous tumor.

This paper describes the presentation, diagnosis, and treatment of a male patient with a Periorbital and Nasal Solitary Fibrous Tumor.

Keywords: Solitary Fibrous Tumor; Paranasal Sinus; Periorbital Área; Endoscopic Sinus Surgery; Relapse; Immunohistochemistry

Abbreviations

STF: Solitary Fibrous Tumor; ESS: Endoscopic Sinus Surgery; CT: Computed Tomography; MRI: Magnetic Resonance Imaging

Introduction

Solitary Fibrous Tumors (SFT) is an uncommon neoplasm, also known as benign (90%) fibrous mesothelioma or submesothelial fibroma [1], is one of the different types of existing mesothelial tumors. The SFT was first described as a primary spindle cell tumor of the pleura generally associated with serosal surfaces in 1931 by Klemperer y Rabin [2,3].

Due to its mesenchymal origin, it has been reported a wide variety of extrapleural sites of the tumor including the pericardium, mediastinum [4], lungs, kidneys, the liver [5] and are found in head and neck: major salivary glands [5-8], nasopharynx, oropharynx, parapharyngeal space [5,10], eyelids, orbit, tongue [5,12], thyroid gland [5,13], larynx [5,11,14] and can also affect the nasal cavity and paranasal sinus [3,9,11].

To the year 2018 up to 92 cases of sinonasal area were reported in English- language literature, most of them provided by single case [15]. SFT can occur worldwide, in all races and ethnic groups, both women and men can get it, but usually diagnosed between 40 - 50 years. The symptoms: sinusitis, nasal obstruction, hyposmia, rhinorrhea, epistaxis, facial pain, exophthalmos, headaches [17]. The diagnosis of SFT is based on the physical examination, nasal

endoscopy, computed tomography scan (CT), magnetic resonance imaging (MRI) and histologic analysis with immunohistochemistry.

SFT are treated by complete surgical resection [16-18], this is recognized as the best definitive treatment for SFTs. The surgical treatment could be lateral rhinotomy, medial maxillectomy, external ethmoidectomy, endoscopic sinus surgery (ESS), degloving [16] or combined transfacial approach.

The tumors show immunoreactivity for cluster of differentiation (CD)34 [20]. Other markers include CD99, vimentin and B-cell lymphoma (BCl-2). Vimentin is non-specific because it is expressed by most mesenchymal and many epithelial neoplasms. SFT's do not confirm immunoreactivity for S-100 protein, epithelial membrane antigen, keratin [18]. This marker help differentiate SFT of Nasal Cavity and Paranasal Sinus from other benign or cancer lesions.

Malignancy is rare in the sinonasal tract however characterized SFT as malignant if the tumor display necrosis and infiltrative margins, hypercellularity, cytological atypia, increased mitoses (more than 4 mitoses per 10 high power field [21]).

The most reliable prognosis factor of SFT of nasal cavity and paranasal sinus dependent on the tumor completely removed with free margins.

Case Report

Author report a case of 36 years-old man presented with a 3-month history of persistent progressive left nasal bleeding, left nose obstruction and left eye pain.

Physical examination revealed a pain small rubbery mass palpable in the superonasal quadrant of the left eye, in the nasal endoscopy a rough, solid, reddish mass that bleeds easily arose from the left ethmoid sinus and extended to the left nasal cavity, the orbit and the maxillary sinus, septal deviation area II-IV to the right. There was no palpable latero-cervical lymphadenopathy of the neck.

Further imaging by CT and MRI disclosed a large left-sided nasal and periorbital cavity homogeneous expansile mass, occupy left maxillary sinus, left ethmoid, extending to the left frontal sinus, and left periorbital region.

Axial CT scan demonstrated a huge expansile enhancing mass, occupying the left nasal cavity with extension to the frontal sinus, maxillary sinus and pushing laterally on the medial wall of the maxilla, without invading the anterior cranial fossa (Figure 1).

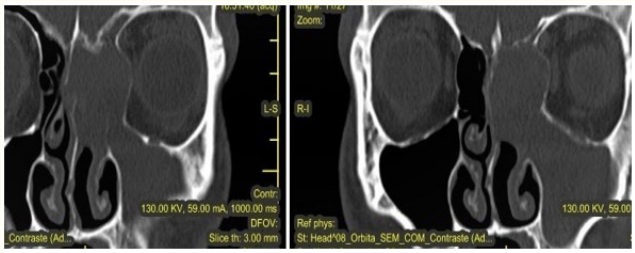

Figure 1

The coronal CT scan (Figure 2) showing a tumor in the nasal cavity, maxillary sinus, ethmoid, extending to the orbit on the left side.

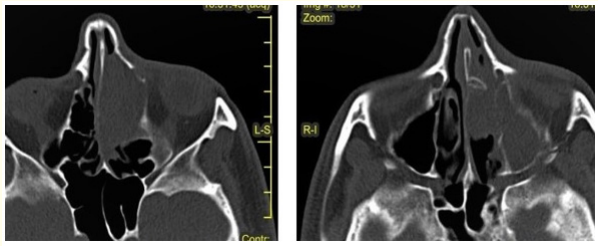

Figure 2

Axial and Sagittal MRI showed expansion of the mass in the left nasal fossa into the left periorbital space, with a hypointense mass on T1 -weighted images and heterogenous isointense mass on T2 -weighted left-side nasal mass (Figure 3).

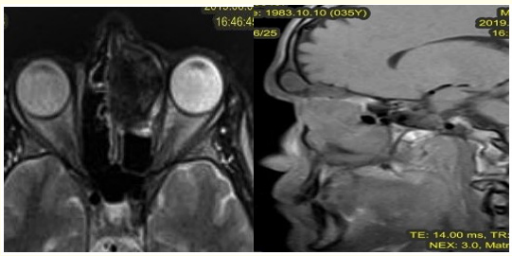

Figure 3

The patient underwent an ESS under general anesthesia, proceed with the excision of the mass that bleed easily and profuse,

without complications. Since the tumor was large and hard to remove in-block, it was resected in fragments.

Histopathologically, the lesion consisted of a non-encapsulated nasal mucosa mass with spindle-shaped cells within a collagen-rich stroma, no mitosis or necrosis, covered by ciliated pseudostratified epithelium. The immunohistochemistry (definitive diagnosis) showed positivity for CD34, CD99, Bcl-2, actin and desmin. Negative for protein S-100 [22,23].

The histological and immunohistochemistry analysis compatible with SFT. In six months follow up no relapse was observed.

Discussion

SFTs are benign rare neoplasms that occur as pleural or serosal tumors [5,6]. Their origin is discussed, SFT grow from mesenchymal rather than the mesothelial tissue. This explains a variety of extrapleural sites of occurrence, SFTs can present itself in any part of the body and have been reported often in the head and neck region up to 5-27% of cases [16], SFT may involve meninges, oral cavity, soft tissue of head and neck, parapharyngeal space [5,10], tongue [5,12], nasopharynx, oropharynx, eyelids, orbit, thyroid gland [5,12], larynx [5,14], major salivary glands [5,8] and can also affect the nasal cavity and paranasal sinus.[3,9,11]. Five percent to 10% of extrapleural SFTs are recurrent [6,18].

The first SFT case arising in nasal cavity in the English literature was reported in 1996 in a 59-year-old woman [23]. To the best of our knowledge there are total 92 reported cases of SFT of the nasal cavity and paranasal sinus so far [24]. So, the present case will be 93rd case of solitary fibrous tumor of nose and paranasal sinuses. Nasal SFT usually presents with unilateral nasal mass, nasal obstruction, rhinorrhea, headache and epistaxis [17]. In this case, the nasal SFT symptoms the patient had been noted are progressive unilateral nasal obstruction, epistaxis and left eye pain for 3 months.

The rhinoscopy and endoscopic findings were a rough, solid, reddish mass that bleeds easily [17,25,26].

Ct shows a lobulated soft tissue mass, well-circumscribed and smooth that sometimes contain scattered calcifications. Larger lesions may have rounded low-attenuation areas due to necrotic or cystic change whereas smaller tumors tend to enhance homogeneously.

On MRI, benign SFT have relatively homogeneous low-to-intermediate signal intensity relative to skeletal muscle on both T1-weighted imaging and T2-weighted imagining because of fibrous tissue, as well as intensive enhancement.

Predominant low signal on T2-weighted images is unusual for other nasal lesions; this feature is an important diagnostic clue for SFT.

Malignant tumors at CT or MRI should be considered in a nasal SFT when a variable contrast enhancement and a central focus of heterogeneity is identified [27,28].

The main differential diagnosis of sinonasal SFT are often confused with lymphoma, fibrous histiocytoma hemangioma, inverted papilloma, schwannoma, fibromatosis, low-grade fibrosarcoma, juvenile angiofibroma, angiomatous polyps, and hemangiopericytoma [28-31].

The definitive diagnosis of SFT is based on histopathological examination and the inmunohistoquemistry. Macroscopically, it is seen as a pedicled exophytic mass and histologically, it is composed of ovoid or spindle-shaped cells randomly distributed along a patterners-pattern within a collagenous stroma of variable vascularity [28,29]. One important finding that aids in the diagnosis of an SFT is the presence of areas of hyalinization usually adjacent to collagen deposits whereas the vascularity encountered is variable [32].

Inmunohistologically SFT is negative for desmin, S-100, actin and epithelial membrane antigen but positive for vimentin, CD34, also can find some other antibodies as BCL-2, CD10, CCD99. The presence of CD34 and Bcl-2 in the immunohistochemical analysis are the most sensitive markers to confirm the diagnosis of SFT [21,22,33].

Rare cases of malignancy of an SFT are sometimes seen, characterized by hypercellularity, pleomorphism, necrosis and increased mitosis [5,32].

SFTs have been treated successfully by ESS in the nasal cavity and paranasal sinuses [26,34] in the cases that SFT was limited in the nasal cavity and paranasal sinuses. In the present case, we could successfully remove a large tumor in the ethmoid sinus, nasal cavity, maxillary sinus and in the periorbital area by ESS in many pieces. ESS is considered the first-line approach for achieving the complete removal of benign tumors such as an SFT.

Conclusion

Solitary fibrous tumor of nasal cavity, paranasal sinuses and the periorbital area is usually a benign, uncommon entity, local invasion or recurrence of the tumor has been demonstrated. its typical imaging characteristics, including bony remodeling and thinning, isointense or hypointense signal on T2-weighted images, marked enhancement on contrast-enhanced T1-weighted images, and a washout TIC pattern may help to suggest this diagnosis preoperatively.

Endoscopic sinus surgery is considered the first line approach for the complete resection of tumors as SFT of the nasal cavity and paranasal sinus and their management differ from other tumors.

Surgeons should be aware of the abundant bleeding during surgery and biopsy as well.

SFT must be differentiated from other nasal and paranasal sinus tumors based on histopathological characteristics and immunohistochemistry analysis (CD34, vimentin, BCL2, CD99). This marker are the most accurate means of diagnosis.

Conflict of Interest

There are not conflicts of interest.

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