An Aggressive Solitary Fibrous Tumor with Evidence of Malignancy: A Rare Case Report

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Abstract Solitary fibrous tumor (SFT) is rare mesenchymal neoplasm that has been originally and most often documented in the pleura. Recently, the ubiquitous nature of the SFT has been recognized with reports of involvement of numerous sites all over the body, i.e., upper respiratory tract, breast, somatic tissue, mediastinum, head, and neck, etc. The diagnosis of SFT still remains an enigma in our field. Furthermore, malignant SFT is extremely rare and only two cases have been reported in the oral cavity till date. Here, we present a rare case report of an aggressive solitary fibrous tumor which presented as a palatal mass and extended throughout the middle cranial fossa and exhibited features of malignancy.

Keywords Solitary fibrous tumor · Malignant solitary fibrous tumor · Palate · Mesenchymal neoplasm

Solitary fibrous tumors (SFT's) are rare mesenchymal neoplasms first documented and frequently encountered in the pleura. It was once referred to as localized fibrous mesothelioma because it was thought to be of submesothelial origin [1]. Since then, numerous hypotheses have been laid down regarding their origin and are now believed to arise from dendritic mesenchymal cells [2]. Recently, the ubiquitous nature of this tumor has been recognized with reports of SFT arising in various extrapleural sites like respiratory tract, lung, breast, somatic tissue, mediastinum, head, and neck, etc. However, involvement of oral cavity is

a rarity [3] with buccal mucosa being the common site of occurrence [4].

Solitary fibrous tumors come under the spectrum of fibroblastic proliferations which are extremely varied with a wide range of histomorphologic patterns posing a diagnostic challenge [5]. In addition, reports of a SFT exhibiting malignant features occurring in the oral cavity are extremely rare [6, 7]. We describe a case of an aggressive solitary fibrous tumor which attained a massive size and showed evidence of malignancy.

Case Report

A 57-year-old male patient presented to our craniofacial unit with a chief complaint of mass on the palate associated with difficulty in swallowing for 3 months. This was accompanied with pain and loss of vision in the right eye since a year. History revealed persistent headache on the right side with no response to analgesics and also hearing loss associated with the right ear. There was no other relevant medical history.

Extraoral examination revealed minimal proptosis of the right eye, with the inability to adduct and no obvious facial asymmetry. (Fig. 1). Intraorally, a diffuse swelling was observed extending from maxillary first molar to soft palate. Uvula and median palatal raphae were deviated to the left. The overlying mucosa was intact and normal in color. On palpation, the swelling was nontender and soft to firm in consistency.

Panoramic radiography revealed destruction of the ramus and thinning of the palate. CT and MRI scans were performed which showed a well defined soft tissue growth in the right skull base region measuring around $9.3 \times 7.1 \times 9.2$ cm. It was seen destroying the middle cranial

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Fig. 1 Extraoral photograph depicting minimal proptosis of the right eye with no obvious asymmetry

fossa, sphenoidal sinus, pituitary fossa, and extending intracranially. It was extending superiorly into right orbit infiltrating the orbital apex, laterally into the infratemporal fossa, inferiorly to the parapharyngeal and retropharyngeal space eroding the parotid gland and ramus of mandible. Anteriorly, it was eroding the maxillary sinus, pterygoid plates, ethmoidal sinus and nasal cavity (Fig. 2a–c). Small necrotic areas were also evident in the lesion. This picture was suggestive of a malignancy and probable diagnosis of lymphoma and malignant nerve sheath tumor was considered.

The patient underwent a thorough preoperative review and clearance was given for surgery with due risk to life considering the proximity of the lesion to vital structures. The lesion was then excised after tracheostomy using modified Weber Ferguson approach.

Macroscopic examination of the excised specimen revealed a lobulated dense rubbery mass measuring 11×5 cm, creamish white in color which was firm in consistency (Fig. 2d). Microscopically, the lesion was highly cellular with areas of myxoid degeneration. The cells were round, ovoid to spindle with vesicular to hyperchromatic nuclei, and sparse cytoplasm and were seen arranged in sheets around numerous dilated blood vessels exhibiting a hemangiopericytoma like pattern (Fig. 3a, b). There were hypocellular areas consisting of densely hyalinised keloid-like collagen interspersed between the tumor cells (Fig. 3c and Fig. 4a, b) The tumor

Fig. 2 CT and MRI scans showing a soft tissue growth in the right skull base region with its various extensions (a, b, c). Gross specimen revealed a lobulated dense rubbery mass (d)

Fig. 3 (a) Photomicrograph of the hypercellular lesion depicting sheets of bland round to ovoid cells with minimal cytoplasm. (H&E, 40×); (b) Extensive vascularity arranged in a hemangioperictoma like pattern. (H&E, 100×) Inset : Perivascular hyalinization. (H&E, 250×); (c) Hypo-cellular areas exhibiting characteristic keloid-like collagen. (H&E, 100×) Inset: Van Gieson stain enhancing the dense collagen. (Van gieson, 100×); (d) Numerous mast cells distributed throughout the lesion (H&E, 100×) Inset: Toluidine blue stained section demonstrating the same. (Toluidine blue, 250×)

Fig. 4 Photomicrograph shows hypocellular areas exhibiting hyalinization intermixed with densely cellular areas of tumor cells arranged in no particular pattern (a, b, H&E, 100×)

cells in several areas exhibited pleomorphism, hyperchromatism of nuclei, mitotic figures, and areas of necrosis (Fig. 5a–d). Additionally, perivascular hyalinization (Fig. 3b) mononucleate stellate giant cells and presence of numerous mast cells (Fig. 3d) were also observed. Immunohistochemically, the tumor cells were positive for CD34, CD99 (Fig. 6a, b) while EMA, SMA, CK, desmin, and S-100 were negative.

The histomorphologic features, immunohistochemical profile, and clinical aggressive behavior led to the diagnosis of malignant SFT. The prognosis in this patient was very poor and patient expired within a month of surgery.

Discussion

The first detailed histologic description of SFT in pleura is credited to Wagner in 1870 but it was first described as a distinct entity by Klemper and Rabin (1931) and they called it as localized fibrous mesothelioma [8]. They proposed the submesothelial cell as the cell of origin for this tumor. Since then, there has been considerable debate regarding its origin with various hypothesis of it arising from mesothelium, fibroblast, primitive cells, pluripotential mesenchymal cells, etc. Recently due to various reports of its ubiquitous occurrence in almost every site of the body,

Fig. 5 Photomicrograph showing features of malignancy, i.e., hypercellularity (a), atypia (b), mitosis (c, Arrows) and areas of necrosis (d). (H&E, 250 $\times$)

Fig. 6 Tumor cells strongly positive for CD34 (a) and CD99 (b)

it is now believed to arise from dendritic mesenchymal cells [2, 9]. Over the years, it has been referred to by various names like benign mesothelioma, solitary fibrous mesothelioma, myofibroblastoma, etc. [9] reflecting the controversies regarding its histogenesis [8, 10].

SFT have been reported in various extrapleural sites like upper respiratory tract, breast, somatic soft tissue, paranasal sinus, orbit, spinal cord, head, and neck [2, 8, 11]. Involvement of oral cavity by SFT is distinctly uncommon and there are only 56 case reports till date with most common site being the buccal mucosa [3, 12–14].

The mean age of occurrence is around 52.3 years and affects mainly middle aged and elderly patients [14]. They are usually slow growing, asymptomatic and largest size reported in literature is around 4.8 cm in the oral cavity [9, 14] while sizes of up to 26 cm have been reported in other sites [15]. The present case was similar in age and sex predilection to that of a classical SFT but exhibited aggressive clinical behavior attaining a huge size and causing considerable debilitation to the patient.

Microscopically, SFT exhibit wide histomorphologic spectrum with areas of haemangiopericytoma like

appearance, storiform, and herringbone like appearance posing a diagnostic challenge [16]. But, characteristically, they are thought to have a patternless pattern with alternating hypo and hypercellular areas. They are composed of round to ovoid/bland spindle cells with vesicular nuclei and sparse cytoplasm with thick hyalinised collagen (keloid-like) interspersed between them. They exhibit prominent vascularity and abundant mast cells [5]. The diagnosis of SFT is almost always confirmed with an immunohistochemical analysis because the tumor cells are immunoreactive for CD34 (90–95%), CD99 (70%), and Bcl-2 (35%) [5, 17].

Chan et al. [8] have summarized the essential diagnostic criteria for the diagnosis of SFT which was fulfilled by our case and this was augmented by strong immunohistochemical CD34 positivity which is a consistent finding in most SFT's confirming the diagnosis [3, 5].

Malignant SFT's are very rare and account to 10–15% of all cases in pleura while in oral cavity there are only two reported cases [6, 7]. Furthermore, there are no unifying histologic criteria that appear to reliably and consistently predict malignancy in SFT [5]. Various histologic criteria has been put forward which include lack of circumscription, nuclear atypia, hypercellularity, a mitotic count greater than or equal to four per 10 HPF and presence of necrosis. Additional criteria, like clinical aggressiveness, large size (>50 cm) resectability, invasiveness, infiltrative margins have also been proposed [5, 8, 18, 19].

Thus, it has been suggested that the morphologic features and clinical growth pattern must be evaluated in the categorization of these tumors into benign and malignant [5, 11]. In the present case, the presence of high cellularity, atypia, mitosis, and areas of necrosis coincidental with clinical aggressive behavior lead to the diagnosis of malignant SFT. The tumor could be graded as grade 2/3 based on the French Federation scheme [20].

The wide morphologic spectrum exhibited by SFT generates a broad differential diagnosis like hemangiopericytoma, fibrosarcoma, desmoplastic fibroma, nerve sheath tumors, malignant fibrous histiocytoma, fibrosarcoma, leiomyosarcoma, synovial sarcoma, etc. SFT's stain positive for vimentin and CD34 and are usually negative for S-100, keratin, EMA, and muscle markers [5]. Leiomyosarcoma, malignant fibrous histiocytoma, and sclerosing epitheloid fibrosarcoma may be confused with malignant SFT but they can be distinguished by lack of CD34 expression. In addition, leiomyosarcoma also exhibits positive immunoreactivity for actin and desmin [2, 21, 22]. Synovial sarcoma are known to express CK which is usually negative in SFT, while desmoplastic fibroma have a more monotonous appearance and lack the haemangiopericytic vascular pattern and CD34 positivity seen in SFT. Peripheral nerve sheath tumors may share some histologic

similarities but unlike SFT they exhibit moderate to strong positivity for S-100 [5, 16]. Thus, immunohistochemistry plays a pivotal role, in diagnosis, especially CD34 positively although not specific but along with compatible histopathological background is very helpful in differentiating SFT from other lesions [1, 3, 13, 23].

We would like to mention that, hemangiopericytoma (HPC) is probably the lesion most difficult to distinguish from SFT due to their morphologic similarities and CD34 positivity. But, HPC is generally more cellular and morphologically homogenous and is not known to express CD-99. However, there is currently a debate as to whether or not HPC simply represents a histologic pattern or is a true biological entity [5, 16]. As a result, there is an increased trend to diagnose most potential HPC's as SFT's as most believe that they represent the spectrum of same entity [5, 16].

Complete surgical excision is the treatment of choice. Although no recurrences have been reported in the head and neck region, a long term follow up of patient is recommended regardless of the anatomic location; if malignancy is suspected in the final pathology [3, 6, 12, 14]. Factors that predispose to local recurrence in non head and neck SFT's include tumor diameter larger than 10 cm, presence of malignant component in histology and microscopic positive surgical margins [11].

In summary, we report a rare case of SFT exhibiting clinical aggressiveness and histopathological features of malignancy which contradicts the general belief that head and neck SFT are usually non aggressive [17]. This case report emphasizes the inclusion of this rare entity in the differential diagnosis of soft tissue tumors of head and neck as well as proper clinicopathologic correlation to categorize it as benign or malignant.

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