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Solitary fibrous tumor with atypical histological features occurring in the palatine tonsil: an uncommon neoplasm in an uncommon site

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Solitary fibrous tumor (SFT) is an uncommon mesenchymal neoplasm that usually arises in the pleura. Although this tumor has been described in other sites, including the head and neck area, in the oropharynx it is extremely rare. We report the first case of a SFT arising from the palatine tonsil of a 62-year-old man. The tumor consisted of spindle-shaped cells distributed in a haphazard pattern and presented atypical histological features such as hypercellular areas and high mitotic count. Immunohistochemical studies showed strong positivity for CD34 and bcl-2, and weak positivity for desmin. Smooth muscle actin, S-100 protein and cytokeratines were negative. The patient was well without disease 1 year after surgery.

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A 62-year-old male presented with a 1 month lasting history of odinophagy and foreign-body sensation in the throat. Past medical history revealed heavy smoking during 48 years, chronic alcoholism and previous treatment for Paracoccidioidomycosis. At physical examination a right palatine tonsil enlargement was detected. The hemogram showed normal parameters for the leukocytes and erythrocytes. A clinical hypothesis of low-grade lymphoma was raised and right tonsillectomy was performed for diagnostic purpose.

Macroscopically, the specimen measured $4.5 \times 4 \times 3$ cm. It had an ovoid form and was covered by mucosal membrane. The cut section showed a whitish, firm, well-delimited mass, measuring $4.3 \times 3.9 \times 2.8$ cm, substituting the tonsil parenchyma. Microscopically, the encapsulated tumor was composed of spindle to epithelioid cells showing pale to eosinophilic cytoplasm, oval or elongated nuclei with inconspicuous nucleoli (Fig. 1, 2). Such cells were arranged in a haphazard pattern with the extra cellular matrix varying from dense collagenized (Fig. 3) to loose-myxoid. There were hypercellular areas in which the nuclei of the cells were hyperchromatic with irregular contours (Fig. 4) and in which the mitotic count achieved seven mitosis per 10 high power fields (Fig. 5). No necrotic foci were detected. There were no signs of residual tonsil lymphoid tissue but the superficial squamous epithelium was intact. The surgical margins were free from neoplasm. Immunohistochemically, the tumor cells were strongly positive for CD34 (Fig. 6), BCL-2, vimentine and focally positive for desmin and CD99. They were negative for S-100 protein, smooth muscle actin, muscle specific actin, EMA and pan-cytokeratine. Based on these findings the diagnosis of solitary fibrous tumor (SFT) with atypical histological features was rendered. No complementary treatment was performed. The patient was followed for 1 year without relapse.

Comments

Solitary fibrous tumor is an uncommon spindle cell neoplasm that was first recognized as a distinctive pleural lesion in 1931 (1). The numerous terms used to describe it reflect the initial controversies regarding its histogenesis. Klemperer and Rabin (1) considered SFT submesothelial in origin, while Stout and Murray (2) later suggested mesothelial genesis, and then terms like solitary fibrous mesothelioma, benign fibrous mesothelioma and localized fibrous mesothelioma were used to refer to this neoplasia. Nowadays SFT is considered a mesenchymal tumor with features of myopericytic, fibroblastic and myofibroblastic differentiation (3, 4). As cellular areas of SFT overlap histologically with hemangiopericytoma (HPC) and the immunoprofile of these entities is similar, they are considered closely related if not a spectrum of the same entity, according to the recent WHO's classification of soft tissues tumors (3).

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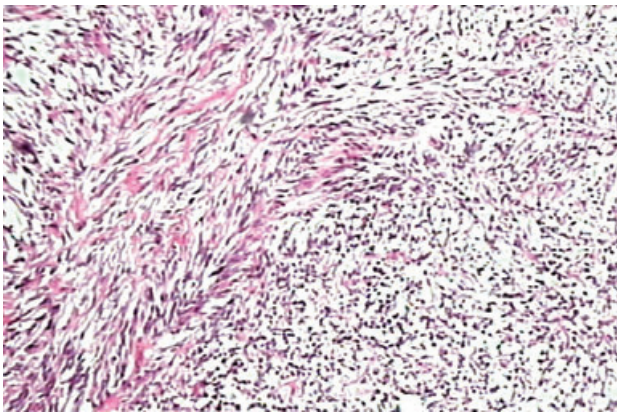

Figure 1 Spindle and epithelioid cells in a 'patternless pattern' arrangement. Collagen fibers are more prominent at the left.

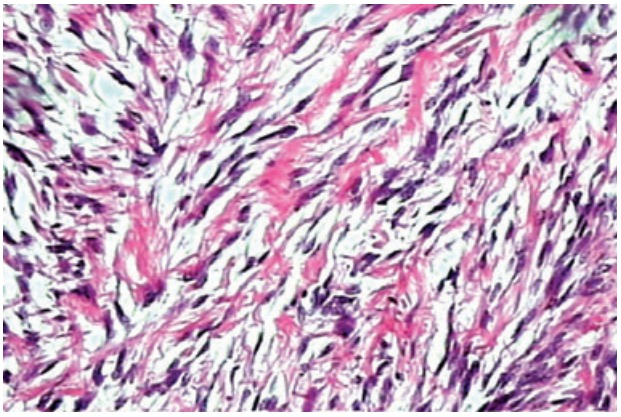

Figure 2 Collagen fibers intermingled with spindle cells that show homogen nuclear chromatin and inconspicuous cytoplasm.

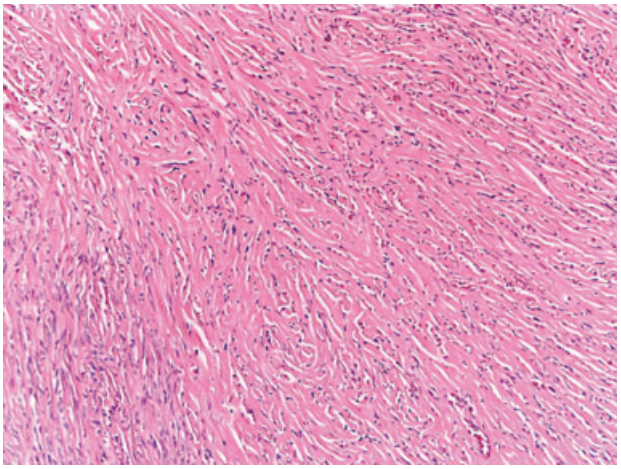

Figure 3 This low power magnification field highlights hypocellular area with abundant collagenized extra cellular matrix.

SFT was originally thought to occur exclusively in the pleura (1), but later extrapleural intrathoracic lesions, including intrapulmonary and mediastinal tumors were described, as well as extrathoracic SFTs have been recognized in numerous sites such as liver, kidney, spinal cord and soft tissues. Of note, extrapleural SFTs may have been reported as HPCs.

In the upper aerodigestive tract SFTs have been reported in the nose and paranasal sinuses (5), nasopharynx (6), larynx (7), tongue and oral cavity (8, 9). Although rare HPCs have been reported in the pharynx (10), to our knowledge no cases of either SFT or HPC have been described in the palatine tonsils so far.

SFT may be difficult to diagnose outside the pleura because of its histological variability. Characteristic microscopic features are described as a 'patternless' growth pattern, with bland spindle cell cytology, alternating hyper and hypocellular areas, keloid-like hyalinization, neural-type palisading and a frequently prominent branching vasculature often described as 'HPC-like'. Irrespective of anatomic location, most SFTs exhibit diffuse and moderate to strong positivity for CD34, bcl-2 and CD99 (9). In the current case CD34 and bcl-2 were both diffusely positive whereas CD99 was focally positive.

Complete surgical removal is curative in most cases of SFT. Despite of its atypical histological signs, our case showed neither recurrence nor metastasis during 1 year after a free margin tonsillectomy. Features alleged to be associated with local or distant recurrence of SFTs include high cellularity, mitotic activity ($>4/10$ HPF), nuclear pleomorphism, and necrosis, but prediction of behavior based on histological criteria alone is not possible, as SFTs at all sites may recur or metastasize after complete resection in the absence or presence of atypical histological features (8). Long-term follow-up therefore is recommended for all SFTs and it is better not to regard any such lesion as definitely benign.

The differential diagnosis in the present case includes leiomyoma, leiomyosarcoma, HPC, myofibroblastic inflammatory tumor (inflammatory pseudotumor), myofibroma/myofibromatosis, fibroma, sarcomatoid carcinoma, melanoma, low grade myofibrosarcoma (LGM) and spindle cell rhabdomyosarcoma (SCR). Leiomyoma and leiomyosarcoma, in contrast to SFTs, are characterized by eosinophilic spindle cells arranged more uniformly in anastomosing intersecting fascicles, and expressing diffuse positivity for desmin and smooth muscle actin. HPC, as already mentioned, can overlap histologically and immunohistochemically with SFT. Indeed most HPCs show CD34 immunoreactivity although in a more focal or weaker fashion than SFTs. The present case was considered a SFT because the overall vascular pattern seen in HPC was not predominant and CD34 was diffusely expressed. Inflammatory pseudotumor (inflammatory myofibroblastic tumor) consists of an admixture of spindle cells resembling SFT and aggregates of plasma cells or lymphocytes. The tumor cells, however, are usually positive for smooth muscle actin and desmin. Unlike SFT, myofibroma/myofibromatosis and fibroma are often strongly and diffusely positive for smooth muscle actin and muscle

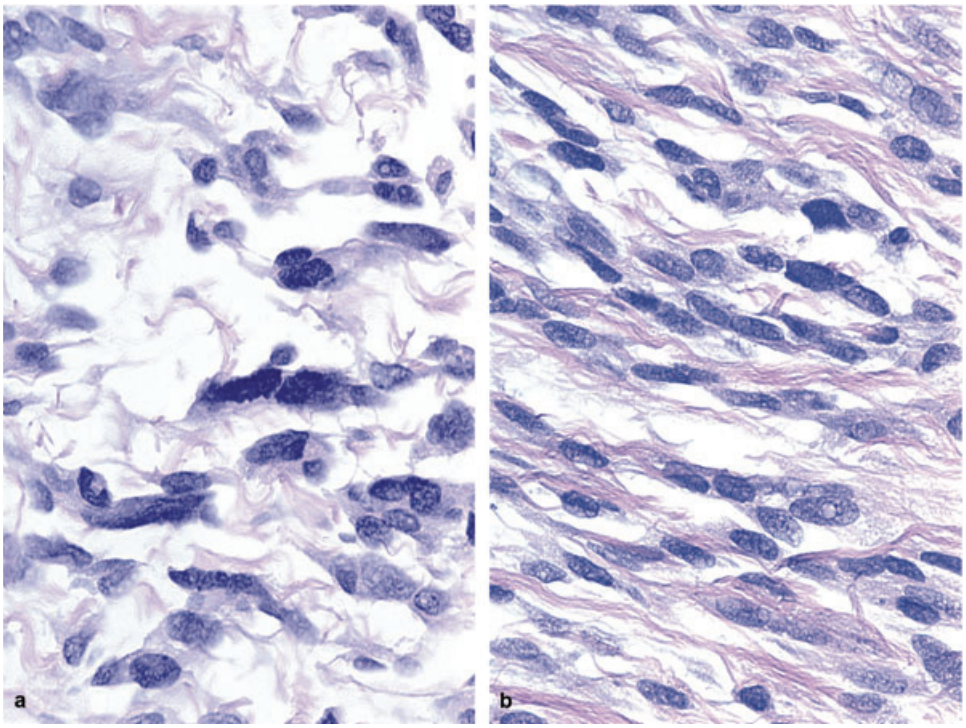

Figure 4 (a) High power magnification field demonstrates a hypercellular area composed of enlarged and hyperchromatic nuclei with irregular contours. (b) Another area showing nuclei with similar features and prominent nucleoli.

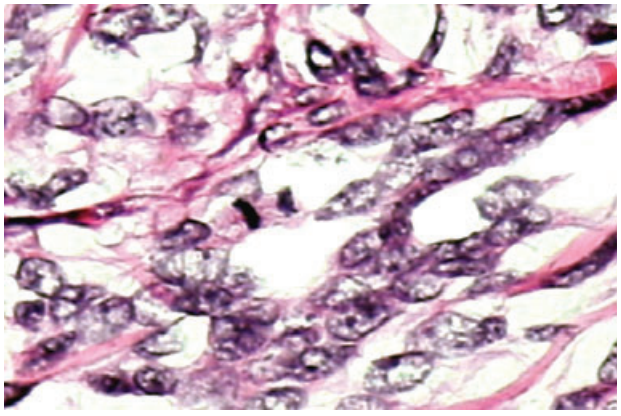

Figure 5 Mitotic figure in a hypercellular area. The highest mitotic count detected was seven mitosis per 10 high power fields.

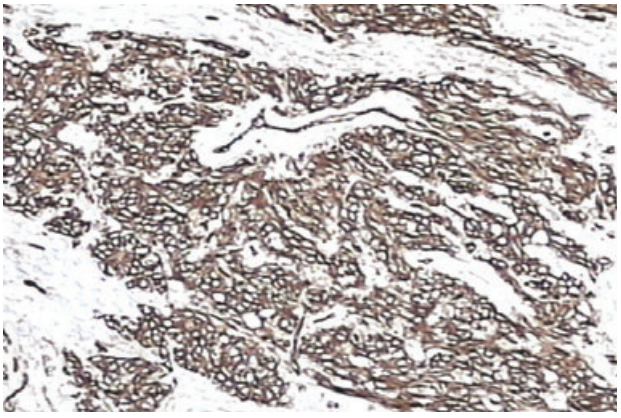

Figure 6 Diffuse and strong immunohistochemical expression of CD34 by the neoplastic cells.

specific actin, whereas CD34 is consistently negative. LGM is an uncommon spindle cell sarcoma that presents predilection for the head and neck area and limbs, it may occasionally show positivity for anti-CD34 antibodies, but unlike the present case LGM has infiltrative growth pattern, fibromatosis-like histology and consistently expresses desmin and/or muscle actins. Most sarcomatoid carcinomas of the upper aerodigestive tract have a greater degree of cellular atypia and mitotic activity than SFTs; some cases show morphologic evidence of epithelial differentiation and most of them express cytokeratines, while SFT usually does not. Although spindle cell melanoma is morphologically in the differential diagnosis list, it can be ruled out in this case by the negativity for S100 protein. SCR is most likely detected in the paratesticular region of children, but recently has been described in adults with predilection for the head and neck area in this age group, including the oral cavity (11). Despite of its spindled morphology, it differs from SFTs by the presence of scattered spindled or polygonal shaped rhabdomyoblasts and by its diffuse expression of desmin and myogenin.

In conclusion, we present the first description of a SFT occurring in the palatine tonsil. At the same time, it also represents a rare example of SFT arising in the

pharynx. It behaved in a benign fashion during 1-year period after local surgical resection.

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