Fine needle aspiration cytology of solitary fibrous tumor of the orbit

ABSTRACT
Solitary fibrous tumor (SFT) is a distinct and a rare spindle cell neoplasm, commonly known to occur in the pleura and other serosal sites. With the advent of immunohistochemistry, varied extraserosal sites are being recognized as common locations for this rare tumor. We report a case of SFT in a 50-year-old male patient who presented with multiple swellings in the eyelid, with emphasis on the cytological features.

Key words: Fine needle aspiration cytology; orbit; solitary fibrous tumor

Introduction
Solitary fibrous tumor (SFT) is an uncommon tumor first described in 1931. Although the tumor frequently presents as a pleural-based mass lesion, it has been reported from almost every site in the body.[1] With disputed histogenesis, the tumor is now considered to be of mesenchymal origin, which explains the divergent location of this lesion.[2] Since the initial description of orbital SFT in 1994, approximately 70 cases of orbital SFTs have been reported.[3] However, literature search shows that cytological features of the lesion have been described in very few cases.[4] Upon consideration of SFT in the differential diagnosis of spindle cell lesions on the basis of typical cytological features, definitive management can be formulated preoperatively in order to prevent its recurrence and an aggressive course following the recurrence.

Case Report
A 50-year-old male patient presented with multiple swellings over the right eye since 2 years. Larger swelling was a firm, pedunculated, and lobulated mass measuring 4 × 3 cm over the medial aspect of left upper eyelid involving the palpebral conjunctiva. The second swelling was on the forehead just above the medial aspect of the left eyebrow measuring 3 × 3 cm in the subcutaneous plane. The third was on the medial aspect of the bulbar conjunctiva of 1 × 1.5 cm size. Cornea was clear. Anterior chamber, lens, and visual acuity were normal. Fine needle aspiration cytology (FNAC) of all the three lesions was done. All the smears were cellular and showed spindle cells in cohesive clusters and in singles lacking polarity. Loose cell aggregates of haphazardly arranged spindle cells in a background of amorphous material were also seen. The cells had oval-to-spindle nuclei with bland chromatin and scanty cytoplasm. Many naked nuclei of cells were also noted. Dense ropy collagen fragments intimately admixed with tumor cells was conspicuous [Figure 1]. A diagnosis of spindle cell tumor with the possibilities of SFT and neural tumor were considered.
The patient later underwent lid construction surgery, during which complete excision of all the three lesions was carried out. On histopathology, a spindle cell tumor was seen with cells arranged in bundles and fascicles. Hypo and hypercellular areas, hyalinized blood vessels, and dense collagen were prominent at places [Figure 2]. Infiltration of adipose tissue and skeletal muscle fibers was seen. The tumor margins were clear. On immunohistochemistry (IHC), the tumor cells were diffusely positive for CD34 and negative for S-100 protein. A final diagnosis of SFT was made.

Discussion

SFTs are rare soft tissue neoplasms commonly recognized in the pleura, mediastinum, and other serosal sites. Because the usual origin is from the pleural surfaces, SFT was originally thought to be derived from mesothelial cells. However, occurrence of the lesion at various extraserosal sites, reactivity of neoplastic cells for vimentin, and non-reactivity for cytokeratin favored mesenchymal origin. CD34 reactivity of SFT is thought to be consistent with an origin in a mesenchymal progenitor cell.

The first case of SFT of the orbit was reported in 1994 by Dorfman et al. Since then, approximately 70 cases of SFTs in the orbit have been reported. They may arise from any of the orbital spaces such as medial/superomedial, lateral/superolateral, retrobulbar soft tissues, lacrimal caruncle, and lacrimal gland fossa. As in the present case, soft tissues of the eyelid can also be the site of origin of SFT.

FNAC is considered to be the first-line diagnostic technique for soft tissue swellings. However, due to their complex heterogeneous constituents, soft tissue tumors are a significant diagnostic challenge for the cytopathologists. The cytological features of SFT include scant-to-moderately cellular aspirates composed of oval-to-spinde cells in a background of irregular ropy fragments of collagen and a few inflammatory cells. Most of the cells are dispersed singly or are present in irregular, loose clusters enmeshed in an eosinophilic collagenous matrix. The tumor cells have uniformly bland nuclei with even finely granular chromatin. Most consistent features are the presence of stripped nuclei in the background and thick ropy bands of matrix material. Aspiration smears of nodular fasciitis will be highly cellular and have metachromatic myxoid background. The cells are mainly myofibroblasts with plump spindle-shaped/stellate cells having pale, bland, evenly distributed chromatin and prominent nucleoli. However, SFTs have monotonous spindly cells without myxoid background. Smears from benign peripheral nerve sheath tumors such as schwannoma contain clusters of spindle cells in interlacing bundles against myxoid background. The diagnostic verocay bodies may be present.

On cytology, the cellular features of SFT can be found in nodular fasciitis, benign peripheral nerve sheath tumor, smooth muscle tumor, synovial sarcoma, low grade malignant peripheral nerve sheath tumor (MPNST), malignant fibrous histiocytoma (MFH), dermatofibrosarcoma protuberance (DFPS), and hemangiopericytoma (HPC). Aspiration smears of nodular fasciitis will be highly cellular and have metachromatic myxoid background. The cells are mainly myofibroblasts with plump spindle-shaped/stellate cells having pale, bland, evenly distributed chromatin and prominent nucleoli. However, SFTs have monotonous spindly cells without myxoid background. Smears from benign peripheral nerve sheath tumors such as schwannoma contain clusters of spindle cells in interlacing bundles against myxoid background. The diagnostic verocay bodies may be present.

Smooth muscle tumors show variable cellularity on aspiration, and unlike SFT, isolated cells are not common. In contrast to the cells of SFT, smooth muscle tumor cells will have abundant dense cytoplasm and the nuclei are regular, central, and elongated.

Though pleomorphic and bizarre cells can be observed in both SFT and MFH, the classic smear of MFH shows highly
cellular pleomorphic spindle cells along with atypical histiocyte-like and giant cells.\textsuperscript{[2]} MPNST is characterized by tumor cells with tapering ends and are variably S-100 positive.\textsuperscript{[7]} Synovial sarcomas are usually cellular and show a pericapillary arrangement of oval-to-round cells along with the presence of mast cells. Low grade fibromyxoid sarcoma and fibrosarcoma may also mimic SFT and clinicoradiological details may be of help.\textsuperscript{[7]}

The importance of IHC for confirming cytological diagnosis has been emphasized in the present case. However, irregular ropy collagen and presence of stripped nuclei in the background are the prominent and distinctive features in most of the reported cases of SFT,\textsuperscript{[9]} including the present case which prompted us to consider SFT in the differential diagnosis on the basis of cytology alone.

In general, SFT behaves in a benign manner and does not metastasize. Cytology of malignant cases shows hypercellular smears and single cells with moderate degree of nuclear pleomorphism and prominent nucleoli.\textsuperscript{[1]} Recurrent SFT can reveal more mitotic activity than the original tumor. Therefore, the most important prognostic factor of SFT is considered to be the complete resectability of the lesion and not the histological appearance.\textsuperscript{[9]}

In the appropriate clinical setting, it is possible to make a correct diagnosis of SFT on cytology and IHC markers can be judiciously used to establish the diagnosis. However, it is necessary to consider the cytological overlaps to avoid false positive diagnosis. Correct preoperative diagnosis on typical cytological appearance permits a wide aggressive resection to avoid recurrence and an aggressive course following recurrence.

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\section*{Conflicts of interest}
There are no conflicts of interest.

\section*{References}
\begin{enumerate}
\item Gerhard R, Fregnani ER, Falzoni R, Siqueira SA, Vargas PA. Cytologic features of solitary fibrous tumor of the parotid gland. A case report. Acta Cytol 2004;48:402-6.
\item Shidham VB, Weiss JP, Quinn TJ, Grotnkowski CE. Fine needle aspiration cytology of gastric solitary fibrous tumor: A case report. Acta Cytol 1998;42:1159-66.
\item Feuerman JM, Fint A, Elner VM. Cystic solitary fibrous tumor of the orbit. Arch Ophthalmol 2010;128:385-7.
\item Clayton AC, Salomão DR, Keeney GL, Nascimento AG. Solitary fibrous tumor: A study of cytologic features of six cases diagnosed by fine-needle aspiration. Diagn Cytopathol 2001;25:172-6.
\item Dorfman DM, To K, Dickersin GR, Rosenberg AE, Pilch BZ. Solitary fibrous tumor of the orbit. Am J Surg Pathol 1994;18:281‑7.
\item Furusato E, Valenzuela IA, Fanburg-Smith JC, Auerbach A, Furusato B, Cameron JD, \textit{et al.} Orbital solitary fibrous tumor: Encompassing terminology for hemangiopericytoma, giant cell angiofibroma, and fibrous histiocytoma of the orbit: Reappraisal of 41 cases. Hum Pathol 2011;42:120-8.
\item Gupta N, Barwad A, Katamuthu K, Rajwanshi A, Radotra BD, Nijhawan R, \textit{et al.} Solitary fibrous tumor: A diagnostic challenge for the cytopathologist. Cytopathology 2012;23:250-5.
\item Chhieng DC, Cohen JM, Cangiarella JF. Fine-needle aspiration of spindle cell and mesenchymal lesions of the salivary glands. Diagn Cytopathol 2000;23:253-9.
\item Krishnakumar S, Subramanian N, Mohan ER, Mahesh L, Biswas J, Rao NA. Solitary fibrous tumor of the orbit: A clinicopathologic study of six cases with review of the literature. Surv Ophthalmol 2003;48:544-54.
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