Case Report

Solitary fibrous tumor arising from pericardium - Case report of a rare entity

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Abstract
Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm mainly originated in the pleural cavity. We report here an unusual case of a large SFT arising from the pericardium. A 40 year female with complaints of left sided chest pain was admitted to our hospital with a diagnosis of pericardial mass. CT showed 11x7 cm mass arising from pericardium showing enhancement and peripheral calcification. Excised mass showed a grey brown well circumscribed nodular mass measuring 8x6x5cm along with attached lobe of lung measuring 6x4x3 cm. Microscopic examination showed a predominant population of monotonous bland looking ovoid to spindle cells arranged in patternless pattern or interlacing bundles with areas of necrosis, hemorrhage and myxoid change, mild nuclear atypia and no mitotic activity. Immunohistochemistry revealed that the tumor cells were positive for CD34, BCL-2, Vimentin and Desmin (focally) and negative for EMA, SMA and Calretinin. Based on histomorphology and immunohistochemistry, the final diagnosis of solitary fibrous tumor was made.

Keywords: solitary fibrous tumor, pericardium

1. Introduction
Solitary fibrous tumors (SFTs) are rare ubiquitous spindle cell neoplasm having their origin most commonly from pleura.1,2 SFTs were first recognized as a distinct entity by Klemperer and Rabin in 1931.2 Initially thought to be of mesothelial in origin, these tumors are now considered to be derived from mesenchymal cells owing to their occurrence in extrathoracic sites devoid of mesothelial cells. Peak incidence occurs in the sixth and seventh decades of life, with both sexes equally affected.3 Solitary fibrous tumors occurring at sites other than pleura is relatively uncommon.4 Very few cases of solitary fibrous tumors of pericardium are reported in the literature reviewed.

2. Case report
A 40 years female presented with complaint of left sided chest pain since 6 months which was constricting and non-radiating in nature. There was no other complaint. Physical examination along with chest and CVS examination was within normal limits. Bronchoscopy revealed bulge in left main bronchus with edematous changes on right sided macosa. ECG showed sinus tachycardia with low voltage graph. CECT showed a mass measuring 11x7 cm arising from pericardium and pushing heart to the right side with left bronchus compressed, mass showed enhancement and peripheral calcification with presence of minimal pleural effusion. Air brochogram sign was present in mid zone. CT guided transthoarcic FNA performed from mass showed low grade spindle cell neoplasm with closest resemblance to spindle cell carcinoid tumor.

Excision biopsy was performed through a median sternotomy approach. Excised mass showed a grey brown well circumscribed nodular mass measuring 6x4x3cm along with attached left lobe of lung measuring 8x6x5 cm. External surface was congested. Cut section showed variegated appearance revealing necrotic and hemorrhagic areas. Microscopic examination showed a predominant population of monotonous bland looking ovoid to spindle cells arranged in patternless pattern or interlacing bundles with mild nuclear atypia and no mitotic activity. Cellular areas showed fibroblast like cells arranged in bundles and other areas showed predominant collagen fibres with few cells. Areas of necrosis, hemorrhage and myxoid change were also seen. Neoplastic cells showed high degree of cohesion, naked nuclei with inconspicuous nucleoli and scant cytoplasm. Immunohistochemistry revealed that the tumor cells were positive for CD34, BCL-2, Vimentin and Desmin (focally) and negative for EMA, SMA and Calretinin. Based on histomorphology and immunohistochemistry, the final diagnosis of solitary fibrous tumor was made. Two years later the patient is asymptomatic, with no evidence of local or distant recurrence.

Figure 1 – grey brown well circumscribed nodular mass measuring 8x6x5 cm with attached left lobe of lung

Figure 2 – H&E (100x) predominant population of monotonous bland looking ovoid to spindle cells arranged in patternless pattern

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Solitary fibrous tumor is a well recognized entity occurring in serosal surfaces seen most commonly arising from pleura. SFTs are usually benign. Atypical and malignant variants; reported in both thoracic and extrathoracic locations are rare and tend to recur and metastasize. The increasing use of immunohistochemistry in recent years has proven a fibroblastic origin of solitary fibrous tumor occasionally along with myofibroblastic differentiation. The description of solitary fibrous tumors in extrathoracic sites devoid of mesothelial cells further reinforces the fibroblastic origin of solitary fibrous tumor. Extrathoracic solitary fibrous tumors and those arising from sites other than pleura appear to have similar histological, immunohistochemical and ultrastructural features as their counterparts arising from pleura. Solitary fibrous tumors of the pericardium are extremely rare neoplasms with only few cases reported in literature so far.

Clinical features are related to the pericardial mass effect. Grossly, solitary fibrous tumors tend to be well circumscribed, firm, fleshy tumors; few cases with diffuse mesothelial involvement have also been reported. Histological variability is the rule and multiple growth patterns have been described. Most tumors have a monomorphic spindle cell pattern resembling low grade fibrosarcoma although broad tumor cell fascicules are rare. Areas of hypercellularity alternate with areas of low cellularity. Less cellular areas can be myxoid or contain abundant collagen. A hemangiopericytoma like vascular pattern may be conspicuous, present in small portion of the lesion or absent. Solitary fibrous tumors are CD 34 and bcl-2 positive. They are consistently negative for epithelial markers, muscle specific actin, desmin, CD 31, CD117, S-100 protein, calretinin and inhibin. Differential diagnosis of solitary fibrous tumor of pericardium include other monomorphic spindle cell tumors, including neurogenic tumors, spindle cell mesotheliomas, monophasic synovial sarcoma and fibrosarcoma. Complete local excision of the tumor is the treatment of choice of solitary fibrous tumor, however, wide local excision of the tumor can be problematic in heart. The prognosis of SFTs is generally good, although recurrences and local spread have been reported. Criteria for malignancy of pleural tumors include necrosis and a mitotic count of greater than four per ten high power fields, but the applicability of these criteria to tumors in pericardium remains unknown. The treatment of choice for SFT is complete local surgical excision, however, it is a surgery which requires skill and caution due to its anatomical location.

References