Abstract: Solitary fibrous tumours (SFT) are rare spindle cell neoplasm that commonly involve lung, pleura and mediastinum. They are believed to be mesenchymal in origin. Though originally thought to arise from pleura increasing numbers of extra pulmonary SFT are reported nowadays because of increased awareness of histology of the tumour. The tumour may be benign or malignant. Here we report a case of extra pulmonary SFT in inguinal region which presented as inguinal hernia.

Keyword: Solitary fibrous tumour, extra pulmonary, mesenchymal

Introduction: Extra pulmonary SFT are rare and share the same histological features as pleural SFT. The most common extra pulmonary site is subcutis. Histologically, SFT are characterized by a typical morphologic appearance of alternating hypo and hypercellular areas of spindle shaped cells, dense bands of collagen, and a hemangiopericytomatous vascular pattern. CD34 immunoreactivity appears useful as a positive marker for distinguishing SFT from other tumours that may enter into the differential diagnosis. Here we report a case of solitary fibrous tumour in inguinal region which had an unusual presentation.

Case report: A 38yr old male presented with complaints of swelling in right inguinal region for 7 months. Examination revealed a swelling of size 5 x 8cm in right inguinal region which was partially reducible with cough impulse. A provisional diagnosis of right irreducible inguinal hernia was made. Planned for right hemioplasty. Intraoperatively a swelling of size 6 x 4 x 3cm was found in subcutaneous plane at the level of external ring. Swelling was excised in toto. Associated Indirect inguinal hernia was present for which hemioplasty was done. Specimen was sent for histopathological examination.
DISCUSSION:
Solitary fibrous tumours (SFT) are rare spindle cell neoplasm. They are mesenchymal in origin. More common in 60 – 70 yrs of age and equal in both sexes. 78% - 88% are benign and 12% - 22% are malignant. SFT was first mentioned in the scientific literature by Wagner. The first discussion of its clinical and pathological properties was by Klemperer and Rabin. Most commonly involve pleura and mediastinum. About 80% of SFTs originate in the visceral pleura, while 20% arise from parietal pleura. Approximately 800 cases of SFT of Pleura have been reported in the literature. Extra pleural sites: Subcutis (40%), deep soft tissue, arm, back, head and neck, peritoneum, retroperitoneum, small intestine, liver, gall bladder, skin, kidney, adrenal, periosteum, spinal cord, testis, prostate, seminal vesical, bladder, vulva, cervix. Over the last 20 years, immunohistochemical studies have provided strong evidence for a mesenchymal origin of these tumours. Solitary fibrous tumours of the pleura express CD34. CD34 is a transmembrane cell surface glycoprotein (originally described as a marker of human hematopoietic stem cells) that is ubiquitously observed in a novel family of interstitial spindle cells involved in antigen presentation and characterized by slender dendritic prolongation of their cytoplasm. van de Rijn and Rouse have described this distinctive group of cells as dendritic interstitial cells and have raised the possibility that solitary fibrous tumours seen in the lung and in other sites may originate from such cells.

Clinical features:
SFT of pleura (STFP):
Malignant SFT are usually symptomatic. Symptoms include cough, chest pain, dyspnea, Hemoptysis and obstructive pneumonitis occur due to airway obstruction. Digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre-Marie-Bamberg syndrome) (10% to 20%) could be respectively due to abnormal production of hepatocyte growth factor or an excessive release of hyaluronic acid by the tumour. These features resolve 2 to 5 months or longer after resection of tumour. Refractory hypoglycemia (Doege-Potter syndrome) (5%) due to insulin-like growth factor II secreted by the tumour which return to normal values within 3 to 4 days after resection of the tumour.

SFT of extrapleural site:
Well circumscribed slow growing painless tumour. Symptoms are related to compression rather than infiltration of adjacent structures.

Radiological features:
Benign and malignant SFTs of pleura usually appear as a well-dened, homogeneous, and rounded mass on the initial chest radiograph. Rarely, a pleural effusion is associated with malignant SFT of pleura. Computed tomographic scan usually demonstrates a well-delineated, homogeneous, and occasionally lobulated mass of soft tissue attenuation. Heterogeneity may be observed with benign and malignant variants of SFT of pleura because of myxoid degeneration, hemorrhage, or necrosis. Magnetic resonance imaging is helpful in differentiating the tumour from other structures and in commencing intrathoracic localization when the tumour abuts the diaphragm. Computed tomographic scan showing a large solitary fibrous tumour of the pleura with heterogeneous zones due to hemorrhage.

AND NECROSIS OF THE TUMOUR

MRI SHOWING A LARGE SOLITARY FIBROUS TUMOUR OF THE PLEURA ON THE LEFT HEMIDIAPHRAGM

Macroscopically, benign and malignant tumours appear as rm, smoothly lobulated masses. Most of them are encapsulated by a thin, translucent membrane, containing a reticulated vascular network. The cut surface appears gray-white to tan with a whorled pattern and may show areas of hemorrhage and necrosis. ROUND, PALE, WELL DELIMITED TUMOUR SURROUNDED BY SMOOTH REDDISH CAPSULE

Microscopically, SFT are characterized by a proliferation of uniform elongated spindle cells intimately intertwining with various amounts of connective tissue. Zones of hypercellularity may alternate with hypocellular or brous areas within the same tumor. Typically, broblasts and connective tissue are arranged in a so called patternless pattern or storiform pattern that is characterized by a haphazard distribution of spindle cells and collagen brils. Occasionally, an increased amount of blood vessels within the tumour causes a hemangiopericytoma-like pattern. More rarely, other patterns, such as herringbone and neural palisading, are also observed inside the tumor.

PATTERNLESS PATTERN

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities
INTERSECTING BUNDLES OF SPINDLE CELLS WITH LITTLE ATYPIA

Histologic signs of malignancy include:
- high mitotic counts, defined as more than four mitoses per 10 highpower fields;
- mild to marked pleomorphism based on nuclear size, irregularity, and nuclear prominence;
- bundles of high cellularity with crowding and overlapping of nuclei;
- presence of necrotic or hemorrhagic zones;
- andstromal or vascular invasion

Differential diagnosis:
- Monophasic fibrous synovial sarcoma,
- Nerve sheath tumours,
- Leiomyosarcoma,
- Malignant fibrous histiocytoma,
- Mesenchymal chondrosarcoma,
- Hemangiopericytoma

Immunohistochemistry:

Immunohistochemistry has been an extremely useful tool to differentiate SFT from mesotheliomas and other sarcomas over the last few years. Indeed, SFT by definition is vimentin positive and keratin negative. In addition, CD34 is positive in most benign and malignant tumour. Though hemangiopericytomas are CD34 positive tumors, the antiapoptotic protooncogene bcl-2 is strongly expressed in SFTP, whereas it is not expressed or is only poorly expressed by hemangiopericytomas.

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Cytogenetics:
Cytogenetic analyses have shown anomalies such as trisomy 8, trisomy 21, or more complex translocations in solitary fibrous tumours may help to distinguish them from mesothelioma and other sarcomas. Chromosomal anomalies were present mainly in tumors larger than 10 cm. Hence, the relationship between size and chromosomal anomalies may suggest that genetic changes may promote tumor growth. Staging: Stage 0 Pedunculated tumor without signs of malignancy Stage I Sessile or inverted tumor without signs of malignancy Stage II Pedunculate tumor with histologic signs of malignancy Stage III Sessile or inverted tumor with histologic signs of malignancy Stage IV Multiple synchronous metastatic tumor

Surgical treatment:
Complete en bloc surgical resection is the mainstay of therapy for all benign and malignant SFT (SFT of pleura and extra pulmonary SFT). A distance of 1 to 2 cm from the tumor is usually recommended. Whereas pedunculated tumors can be safely resected with a wedge resection of the lung, large sessile tumors can be difficult to resect because of extensive adhesions and may occasionally require a lobectomy or a pneumonectomy in order to achieve complete resection.

Adjuvant therapy:
The role of adjuvant therapy in SFT of pleura has not been systematically explored because of the limited number of patients. However, some indices suggest that radiotherapy and chemotherapy could be beneficial in some patients. Suter and colleagues have reported 1 patient who is alive with no evidence of disease more than 20 years after subtotal resection of the tumor followed by radiotherapy, and Veronesi and colleagues have observed significant reduction of an inoperable recurrent SFT of pleura with ifosfamide and Adriamycin.

Prognosis:
*Intra thoracic solitary fibrous tumours*
Benign – 2-8% recurrence Malignant – 14-63% recurrence 22% metastasis

*Extra thoracic solitary fibrous tumour*
Recurrence is more when size of the tumor is >10 cm Presence of malignant areas

Follow up:
Risk of recurrence is more after excision of malignant SFT. Review of the literature has shown that the majority of the recurrences after resection of malignant sessile tumors occur within the first 24 months after the initial resection and that approximately half of the recurrences were the cause of death during that period. Hence, half-yearly radiologic control with chest x-ray or computed tomographic scan during the initial 2 years after the resection and yearly thereafter seems warranted and may help to reduce the mortality from malignant SFT. In case of recurrence, aggressive surgical resection remains the treatment of choice and may lead to long-term survival. Adjuvant therapy should be considered if the tumor appears histologically malignant.

Conclusion: Solitary fibrous tumour are rare pleural tumors. Its extrapulmonary location is very rare. So far only five cases have been reported in the inguinal region in literature. Peculiarity of this case is, it presented as inguinal hernia and this is the first case reported in our hospital. Histologic feature of our case was consistent with benign tumour. Follow up was done at regular interval for this case. Recurrence has not occurred so far.

REFERENCES: