Cancer Association of South Africa (CANSA)

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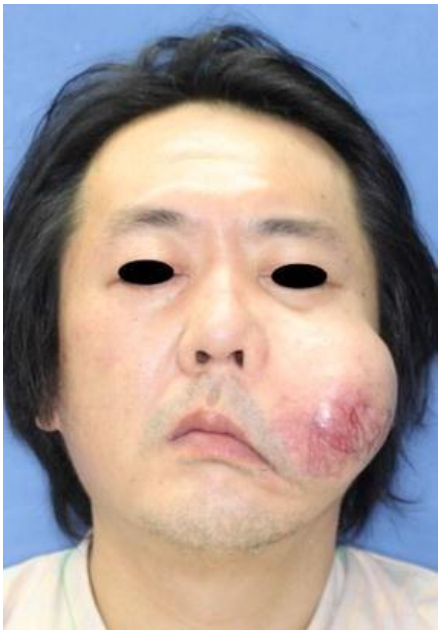
Fact Sheet
on
Solitary Fibrous Tumour

Introduction

Solitary fibrous tumour is an uncommon form of soft tissue tumour which, although seen predominantly within the lung pleura, can occur throughout the body in sites such as the peritoneum, mediastinum and head and neck.



Solitary fibrous tumour (SFT) was first described in 1870 by Wagner and further established in 1931 by Klemperer and Rabin as a pleural neoplasm. The term emangiopericytoma (HPC) was first used by Stout and Murray in 1942 to describe a distinct neoplasm of pericytic origin. However, over time, the staghorn-branching vascular pattern representative of HPC was found to be present at least focally in 15% of all soft-tissue tumours and was more of a characteristic histopathologic pattern than a specific clinicopathologic entity.

Today, the diagnosis of HPC is primarily reserved to neuropathologists. The term SFT is favoured by soft-tissue pathologists to describe a rare, heterogeneous group of benign and malignant neoplasms along a morphologic continuum.

Solitary Fibrous Tumour

Most solitary fibrous tumours are noncancerous (benign), but in rare cases, solitary fibrous tumours can be cancerous (malignant). Solitary fibrous tumours tend to grow slowly and may not cause signs and symptoms until they become very large.

Solitary fibrous tumours are a rare neoplasm of mesenchymal origin that comprise less than 2% of all soft tissue tumours. The majority are benign, although up to 20% may be malignant.

The only way to be certain if a tumour is benign or malignant is with a pathology examination.

Researched and Authored by Prof Michael C Herbst
[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer; Medical Ethicist]
Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

The majority of solitary fibrous tumours occur in an intrathoracic location but up to one third may occur in extrathoracic locations, and as such, they may be encountered essentially anywhere, including:

- pleura
- spinal cord
- dura
- head and neck
- extremities
- abdominal parenchymal organs
- retroperitoneum
- peritoneum
- pelvic organs

Huang, S-C. & Huang, H-Y. 2019.

"Solitary fibrous tumor (SFT) is a distinct fibroblastic neoplasm of intermediate biological potential, prototypically presenting as a pleura-associated tumor characterized by patternless proliferation of generally banal oval to spindle cells with hemangiopericytoma-like staghorn vessels in fibrocollagenous stroma. Over the past decades, the clinicopathological spectrum of SFT has been ever-expanding with the incorporation of cases exhibiting myxoid, giant cell-containing, and fat-forming histology, as well as those from extrathoracic sites, including the meninx. Atypical, frankly malignant and even dedifferentiated variants have also been recognized in a subset of SFTs. Notably, the recent groundbreaking discovery of the disease-defining NAB2-STAT6 gene fusion, resulting from intrachromosomal inversion involving 12q13.3, has largely unified tumors with the aforementioned variations. The derived immunohistochemical detection of nuclear STAT6 expression has high diagnostic value in distinguishing SFTs from histologic mimics, although some relevant pitfalls have been proposed as a precaution. NAB2-STAT6 fusions yield numerous transcript subtypes associated with the clinicopathological variations. Despite mostly following a favorable course, SFT is notoriously difficult for prognostication because of the propensity for late relapse or even metastases in 10-40% of cases, which prompts several proposed schemes incorporating age, size, mitosis, and/or necrosis as factors for risk stratification. Mitotic figures >4/10 HPFs, TERT promoter and/or TP53 mutations have been considered as variables that are better correlated with aggressiveness. Although radiotherapy and chemotherapy provide unsatisfactory responses, a better understanding of SFT tumorigenesis may pave the way for new treatment modalities. In this review, we comprehensively discuss the recent advances of SFTs in diagnostic and molecular pathology."

Incidence of Solitary Fibrous Tumour

The south African National Cancer Registry (2017) does not provide any information regarding the incidence

Signs and Symptoms of Solitary Fibrous Tumour

Painless and slow growth are the main sign/symptoms. Solitary fibrous tumour can also cause hypoglycaemia due to insuline-like growth factor production (called paraneoplastic hypoglycaemia), or local compression symptoms in case of large tumour (nasal cavity, the orbit and meninges).

Diagnosis of Solitary Fibrous Tumour

To confirm a suspected solitary fibrous tumour, a doctor may remove a small piece of tissue for examination under a microscope (biopsy). An experienced pathologist can confirm the diagnosis and determine whether the tumour is cancerous.

Imaging tests may include:

- X-rays
- Ultrasound
- Computerized tomography (CT) scan
- Magnetic resonance imaging (MRI)
- Positron emission tomography (PET)

Treatment of Solitary Fibrous Tumour

Due to the paucity of randomized control trials there is no established, globally accepted treatment strategy. As a result, SFT should be approached in a manner similar to other soft tissue sarcomas, by a multidisciplinary team consisting of surgeons, medical oncologists, radiation oncologists, and additional ancillary support. This strategy has proven to improve outcomes in soft tissue sarcomas.

Treatment may include:

- Surgery
- Radiation Therapy
- Chemotherapy
- Oncological surveillance

Salimov, Z.M., Pikin, O.V., Ryabov, A.B., Popov, M.I., Kononets, P.V., Volchenko, N.N., Blinov, D.A., Nikulin, K.A. 2020.

Objective: Optimization of diagnosis and treatment of patients with solitary fibrous tumor of pleura, analysis of overall survival and disease-free survival, predictors of recurrence.

Material and methods: There were 66 patients with solitary fibrous tumor of pleura (26 men and 40 women) aged 57.6 years (range 26-80 years). Asymptomatic course was found in 29 (44%) patients, various symptoms - in 37 (56%) patients. Thoracotomy was applied in 36 patients, thoracoscopy - in 30 patients. Immunohistochemical examination included analysis of definition of Stat6 expression.

Results: Benign variant of SFT was diagnosed in 50 (75.7%) patients, malignant variant - in 16 (24.3%) patients. STAT6 expression was observed in all cases. Postoperative morbidity was 9%, mortality - 1.6%. Recurrence was diagnosed in 2 (4%) patients with benign variant of disease and in 5 (31.2%) patients with malignant variant (2 of them died from progression of disease). Progression-free survival was 89.4%, overall survival - 95.4%. Predictors of recurrence are tumor dimension over 10 cm, necrosis and/or hemorrhagic component of tumor, mitotic count of at least four per 10 high-power fields.

Conclusion. s: Olitary fibrous tumor of pleura is a rare mesenchymal fibroblastic neoplasm growing from submesothelial layer. Differential and preoperative morphological diagnosis of SFT is difficult and demands a special immunohistochemical examination with analysis of Stat 6 expression. Surgery is preferred for tumor de novo and recurrent neoplasm.

Mansilla Fernández, B., Román de Aragón, M., Paz Solís, J.F., García Feijoo, P., Roda Frade, J. & Regojo Zapata, M.R. 2020.

Introduction: Solitary fibrous tumor (TFS) is a rare tumor of mesenchymal origin, located mainly in the pleura. It is extraordinarily infrequent find it at the intraespinal level, being the thoracic region the most frequent.
Case presentation: We present the case of a 48-year-old patient with progressive ascending lower limb and myelopathy of one month of evolution, with intraspinal location at the D3-D4 level. It was surgically operated by posterior dorsal approach and D3-D4 laminoplasty, with an intradural tumor with an intramedullary component of approximately 18×12mm. The resection was complete and the pathological anatomy gave the diagnosis of solitary fibrous tumor. The patient is currently asymptomatic.
Discussion: Complete tumor resection and histopathological features are the main prognostic factors. Surgery have a main role in this type of neoplasia.
Conclusion: There are few case published of solitary fibrous tumor with intraspinal localization. We apport another case to the literature.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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