Annals of
Clinical Epidemiology and Toxicology

Bajaj A. Ann Clin Epi Toxi: ACET-104

Opinion Open Access

The Furcated Metarteriole-Solitary Fibrous Tumour

Dr. Anubha Bajaj*

Consultant Histopathologist, A.B. Diagnostics, Delhi, India

Solitary fibrous tumour is a neoplasm of fibroblastic origin and obscure aetiology demonstrating innumerable histological configurations. Additionally designated as haemangiopericytoma or giant cell angiofibroma, tumefaction can appropriately be subjected to risk stratification which is indicative of possible prognostic outcomes. Solitary fibrous tumour exemplifies localized tumour reoccurrence or distant metastasis.

An equivalent gender predilection is enunciated. Elderly population is commonly incriminated with a peak of disease emergence between 40 years to 70 years [1,2].

Although no site of tumour emergence is exempt, neoplasm may appear within diverse anatomic locales, superficial and deep-seated soft tissues, visceral organs or bone. In contrast to superficial soft tissues, deep-seated tissues are commonly incriminated. Also, in contrast to pleural neoplasms, extra-pleural tumefaction is common [1,2]. Upper and lower extremities, abdominal cavity, pelvis, retroperitoneum, sites within head and neck as sinonasal tract or orbit and trunk depict neoplastic emergence, in decreasing order of frequency [1,2].

Solitary fibrous tumour exhibits para-centric inversion of chromosome 12q with consequent fusion of NAB2-STAT6 gene. Genomic rearrangement or fusion of NAB2-STAT6 gene can be evaluated with cogent STAT6 immunohistochemistry [1,2].
NAB2 exon 4 STAT6 exon 2 / 4 is a commonly discerned genotype generally associated with thoracic neoplasms occurring within elderly population [1,2].

NAB2 exon 6 STAT6 exon 16 / 18 reflects an aggressive phenotype and is encountered in extra-pleural neoplasms [1,2]. Genetic fusion of NAB2-STAT6 metamorphoses NAB2 from transcriptional repressor to activator. Altered NAB2 function engenders constitutive or deregulated expression of early growth response 1 (EGR1) target genes such as IGF2 or FGFR1[1,2].

Additionally, overexpression of ALDH1, EGFR or JAK2 genetic constituents may be observed along with TERT promoter mutations [1,2]. Typically, solitary fibrous tumour enunciates clinical symptoms contingent to anatomic location of tumefaction [1,2].

Somatic, soft tissue neoplasms manifest as gradually progressive, painless tumefaction [1,2]. Tumours confined to abdominal cavity or pelvic neoplasms exhibit clinical symptoms on account of impingement of diverse organs [1,2]. Tumours confined to pleura are discerned incidentally although exophytic tumour expansion into pulmonary parenchyma may ensue [1,2].
Paraneoplastic syndrome or Doege-Potter syndrome is an extremely exceptional condition arising on account of neoplastic production of insulin-like growth factor 2 (IGF2) [1,2].

Upon gross examination, tumefaction appears un-encapsulated, well circumscribed and occasionally lobulated with magnitude varying from 5 centimetres to 15 centimetres. Tumours situated upon serosal surfaces manifest as exophytic lesions [1,2].

Cut surface of tumefaction is fibrous, firm, grey/white, yellow or red/brown. Foci of cystic, haemorrhagic or myxoid degeneration may be discerned. Malignant metamorphosis is associated with tumour necrosis and focal haemorrhage [1,2].

Frozen section depicts a well circumscribed neoplasm composed of spindle-shaped cells intermixed with staghorn vasculature [1,2].

Cytological assessment exhibits minimally or moderately cellular tumefaction comprised of elliptical to elongated, spherical or stellate cells incorporated with wispy cytoplasm. Circumscribing stroma appears pink and collagenous. Malignant metamorphosis is associated with enhanced cellularity and nuclear pleomorphism [1,2].

Upon microscopy, tumour is composed of ovoid to fusiform, spindle-shaped cells with indistinct cellular perimeter. Tumour cells configure haphazard, abridged, poorly defined fascicles [1,2].

Foci of distended, branching, hyalinised, staghorn-like or haemangiopericytoma-like vascular articulations appear commingled with neoplastic cells. Intervening hyalinised to collagenous stroma with occasional cellular streaming between collagenous fibres is observed. Foci of myxoid alterations can be prominent [1,2].

***Corresponding author:** Dr. Anubha Bajaj, Consultant Histopathologist, A.B. Diagnostics, Delhi, India.
Email: anubha.bajaj@gmail.com*

***Citation:** Bajaj A (2022) The Furcated Metarteriole-Solitary Fibrous Tumour. Ann Clin Epi Toxi: ACET-104.*

***Received Date:** July 10, 2022; **Accepted Date:** July 20, 2022; **Accepted Date:** July 30, 2022*

Characteristically, spindle-shaped to elliptical cells configure a haphazard tumour pattern and appear intermingled with branching, dilated vasculature, prominent, staghorn vascular articulations and variably collagenous intervening stroma [1,2]. Lipomatous variant of solitary fibrous tumour is pervaded with variable quantities of mature adipose tissue (1,2). Giant cell rich variant or giant cell angiofibroma depicts an intermingling of multinucleated giant cells which layer pseudo-vascular spaces confined to tumour parenchyma [1,2].

Hyper-cellular neoplasms composed of spherical or anaplastic tumour cells with cellular atypia, nuclear pleomorphism, elevated mitotic activity and tumour cell necrosis are denominated as 'malignant solitary fibrous tumour' [1,2].
Dedifferentiated or anaplastic solitary fibrous tumour exhibits focal transition into high grade, sarcoma-like foci admixed with or devoid of heterologous components as cartilage, bone, mature adipose tissue or skeletal muscle [1,2].

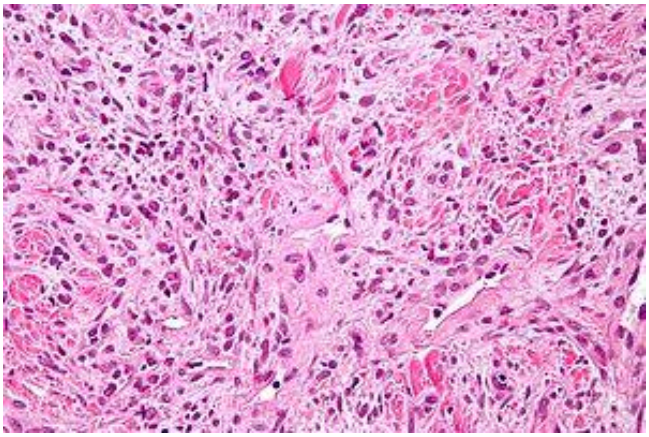

Figure 1: Solitary fibrous tumour exhibiting spherical to elliptical cells with indistinct cytoplasm intermingled with prominent staghorn vasculature and circumscribing collagenous stroma [5].

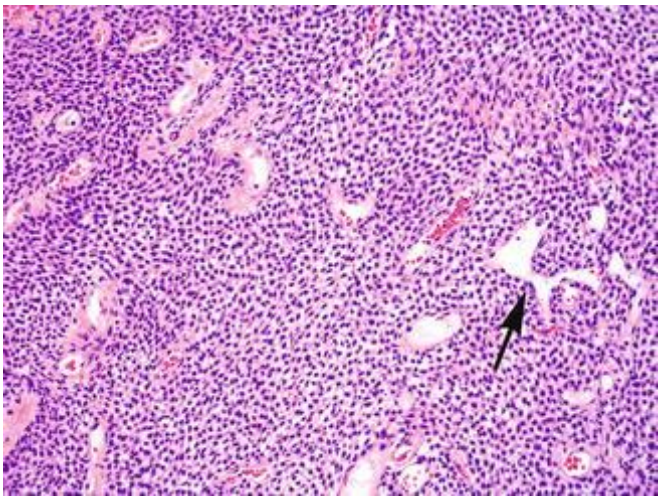

Figure 2: Solitary fibrous tumour demonstrating spherical to spindle-shaped tumour cells with wispy cytoplasm commingled with branching, staghorn vasculature and surrounding collagenous stroma [6].

Table: Risk assessment in Solitary Fibrous Tumour [4].

Risk factor	Score
Age	
<55 years	0
≥55 years	1
Tumour magnitude(cm)	
<5	0
5 to<10	1
10 to<15	2
≥15	3
Mitotic count(/10 high power fields)	
0	0
1-3	1
≥4	2
Tumour necrosis	
<10%	0
≥10%	1
Risk gradation	
Low risk	0-3
Intermediate risk	4-5
High risk	6-7

Ultrastructural examination enunciates fibroblast-like cells imbued with an expansive rough endoplasmic reticulum. Circumscribing collagen fibres can be delineated [3,4].

Solitary fibrous tumour is immune reactive to CD34, STAT6, EMA, actin, bcl2 or CD99. Tumour cells are intensely immune reactive to CD34. Solitary fibrous tumour is immune non-reactive to S100 protein, SOX10, cytokeratin, CD31 and desmin [3,4].

Solitary fibrous tumour requires a segregation from neoplasms such as synovial sarcoma, mesenchymal chondrosarcoma, deep fibrous histiocytoma, mammary type myofibroblastoma, myopericytoma, dermatofibrosarcoma protuberans, gastrointestinal stromal tumour and well differentiated or dedifferentiated liposarcoma [3,4].

Solitary fibrous tumour exhibits nonspecific features on imaging. Cogent histological features are required for appropriate disease discernment. Computerized tomography (CT) demonstrates a well-defined, lobulated tumefaction which is iso-dense as compared to skeletal muscle [3,4]. Heterogeneous image enhancement is observed upon contrast administration [3,4].

Magnetic resonance imaging (MRI) exhibits intermediate image density upon T1 weighted imaging. Upon T2 weighted imaging, tumefaction appears hypo-intense within cellular or fibrous areas and hyper-intense within myxoid zones [3,4]. Multiplexed parallel sequencing assays can optimally detect chromosomal fusion of NAB2-STAT6 gene [3,4]. Cogent discernment of genetic fusion by conventional cytogenetic techniques can be challenging due to proximal situation of NAB2 and STAT6 upon chromosome 12q [3,4].

Employment of PCR techniques for detecting diverse genetic fusions and variants appears inefficacious (3,4). Surgical extermination of solitary fibrous tumour is an optimal treatment strategy. However, extended surveillance

following surgical intervention is recommended. Amalgamation of surgical resection and radiation therapy can be adopted in order to ameliorate localized tumour containment [3,4].

Adoption of chemotherapy appears debatable and therapeutic capacity of chemotherapeutic agents appears limited [3,4]. Factors contributing to inferior prognostic outcomes emerge as mitotic figures > 4 per 10 high power fields, tumour magnitude > 10 centimetres, tumour necrosis, hyper-cellular areas, cellular atypia, dedifferentiation, tumours confined to thoracic cavity, abdominal cavity or pelvis and occurrence of TERT promoter genetic mutations [3,4].

References

1. Teranishi Y, Hongou H et al. Solitary fibrous tumor/hemangiopericytoma. No Shinkei Geka. 2022 ;50(1):141-149.
2. Mudie LI, Echalier EL et al. An intraocular solitary fibrous tumor/hemangiopericytoma with extrascleral extension: Case report and review of literature. Am J Ophthalmol Case Rep. 2022. 26:101513; doi: 10.1016/j.ajoc.2022.101513.
3. Wu H, Du J et al. Aberrant expression of thyroid transcription factor-1 in meningeal solitary fibrous tumor/hemangiopericytoma. Brain Tumor Pathol. 2021 ;38(2):122-131.
4. Demicco E, Wagner M et al. Risk assessment in solitary fibrous tumors: validation and refinement of a risk stratification model. Mod Pathol. 2017.30; 1433–1442.
5. Image 1 Courtesy: Libre Pathology
6. Image 6 Courtesy: Springer link