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Commentary

Sinewy and Tendinous-Sclerosing Rhabdomyosarcoma

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Received: February 21, 2024; **Published:** March 08, 2024

Sclerosing or spindle cell rhabdomyosarcoma emerges as an exceptional subtype of rhabdomyosarcoma associated with specific genetic mutations and focal sclerosis. Characteristically, tumefaction is constituted of spindle shaped cells configuring fascicles admixed with foci of stromal sclerosis. Additionally designated as spindle cell rhabdomyosarcoma, subcategorization of the neoplasm is contingent to occurrence of definitive genetic alterations which concur with prognostic outcomes.

Although morphological concordance is observed within various subtypes, the preponderantly heterogeneous neoplasm displays diversity within genetic and clinical segments, especially occurring within paediatric lesions.

Sclerosing rhabdomyosarcoma exhibits a male preponderance. Neoplasm commonly incriminates adult subjects although may arise within children [1,2].

Sclerosing rhabdomyosarcoma demonstrates a predilection for upper or lower extremities or head and neck region whereas spindle cell variant is commonly observed within the parathesis or head and neck, in decreasing order of frequency. Paediatric neoplasms are commonly confined to paratesticular region. Neoplasm expounds a proportionate ~10% of rhabdomyosarcomas [1,2].

Tumefaction demonstrates multiple genetic anomalies.

Congenital or infantile spindle cell subtype exhibits genomic fusion within VGLL2, SRF, TEAD1, NCOA2, CITED2 genes. Typically, tumefaction delineating VGLL2, NCOA2 and CITED2 genetic rearrangements occur within infants and demonstrate favourable prognosis. Infantile spindle cell rhabdomyosarcoma delineating VGLL2::NCOA2 associated genetic fusion is associated with superior prognostic outcomes [2,3].

Spindle cell or sclerosing rhabdomyosarcoma arising in adolescents or adults expounds MYOD1 genetic mutations. Neoplasms depicting MYOD1 chromosomal mutations implicate adolescents or adults and delineate an unfavourable prognostic outcome [2,3].

Intraosseous spindle cell or epithelioid sclerosing rhabdomyosarcoma exemplifies EWSR1 or FUS genetic fusion with TFCP2 or MEIS1::NCOA2 genomic fusion. Also, lesions exemplifying TFCP2::NCOA2 genomic rearrangements articulate intraosseous neoplasms.

Generally, tumour cells are devoid of PAX genetic fusion [2,3].

Grossly, tumefaction exhibits a firm, nodular countenance with variable circumscription and grey/white to tan hue. Cut surface may occasionally be whorled [3,4].

Upon microscopy, fascicles of spindle shaped cells configure a 'herringbone' pattern, thereby simulating leiomyosarcoma or fibrosarcoma. Sclerosing rhabdomyosarcoma exhibits prominent foci of hyalinization or stromal sclerosis. Tumour cells configure cords, nests, trabeculae or expound a distinct pseudo-vascular pattern [3,4].

Focal areas of primitive, undifferentiated tumour zones composed of spherical cells pervaded with hyperchromatic nuclei may be encountered. Few neoplasms exemplify 'tadpole' cells or 'strap' cells admixed with accumulates of rhabdomyoblasts.

Apart from characteristic areas composed of spindle shaped cells, tumefaction confined to bone demonstrates epithelioid cells configuring sheets and fascicles [3,4].

Histological subtype	Score
Atypical lipomatous tumour/Well differentiated liposarcoma	1
Well differentiated leiomyosarcoma	1
Malignant neurofibroma	1
Well differentiated fibrosarcoma	1
Myxoid liposarcoma	2
Conventional leiomyosarcoma	2
Conventional fibrosarcoma	2
Myxofibrosarcoma	2
High grade myxoid (round cell) liposarcoma	3
Pleomorphic liposarcoma	3
Dedifferentiated liposarcoma	3
Pleomorphic rhabdomyosarcoma	3
Poorly differentiated/ pleomorphic leiomyosarcoma	3
Biphasic/monophasic/poorly differentiated synovial sarcoma	3
Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Extraskeletal Ewing's sarcoma	3
Malignant rhabdoid tumour	3
Undifferentiated pleomorphic sarcoma	3
Undifferentiated sarcoma, not otherwise specified	3

Table 1: *Differentiation of soft tissue tumours [2,3].*

Prognosis	Subtype of rhabdomyosarcoma
Superior prognosis	Botryoid, spindle cell
Intermediate prognosis	Embryonal
Inferior prognosis	Alveolar, undifferentiated

Table 2: *Prognostic outcomes of rhabdomyosarcoma [3].*

Tumour cells are variably immune reactive to desmin and focally express myogenin. Neoplasms associated with MYOD1 genetic mutations display significant immune reactivity to MyoD1. Intraosseous lesions appear immune reactive to keratin and anaplastic lymphoma kinase (ALK) [5,6].

Sclerosing rhabdomyosarcoma requires segregation from neoplasms as alveolar rhabdomyosarcoma, angiosarcoma, mesenchymal chondrosarcoma, metastatic carcinoma, osteosarcoma, sclerosing epithelioid fibrosarcoma, synovial sarcoma, leiomyosarcoma, spindle cell carcinoma, spindle cell melanoma, fibrosarcoma, malignant peripheral nerve sheath tumour or malignant triton tumour [5,6].

Spindle shaped cells configuring cellular fascicles or tumour cells enmeshed within a sclerotic or collagenous stroma appear immune reactive to desmin, myogenin or MyoD1.

Ascertainment of MYOD1 genetic mutation or diverse genetic rearrangements may be beneficially adopted for subcategorizing the neoplasm [5,6].

Upon magnetic resonance imaging (MRI), tumefaction manifests as a heterogeneous soft tissue mass with image enhancement.

Upon T1 weighted magnetic resonance imaging, neoplasm exemplifies hypo-intense to isointense signal intensity.

Upon T2 weighted imaging, tumefaction appears hypo-intense to hyper-intense [5,6].

Neoplasm is devoid of optimal, recommended consensus of applicable therapeutic guidelines.

Congenital, localized tumours may be subjected to comprehensive surgical eradication, a procedure associated with favourable prognostic outcomes.

Majority of neoplasms are optimally managed with a combination of surgical intervention, radiotherapy or chemotherapy [5,6].

Tumour relapse and neoplastic progression are commonly observed.

Neoplasms arising in adults are accompanied by tumour reoccurrence and distant metastasis in up to ~ 50% instances.

Congenital or infantile spindle cell variant with concomitant genetic fusions expounds superior prognostic outcomes.

Spindle cell or sclerosing rhabdomyosarcoma devoid of specific genetic anomalies is associated with favourable prognosis.

Lesions delineating chromosomal mutations within MYOD1 or TFCP2 associated genetic fusion enunciate adverse prognostic outcomes [5,6].

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6. Image 1 Courtesy: Academic oup.
7. Image 2 Courtesy: Wikipedia.

Volume 23 Issue 3 March 2024