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Editorial

Sinewy and Intramural-Intramuscular Myxoma

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Intramuscular myxoma emerges as an exceptionally discerned, benign, gelatinous lesion reminiscent of foetal umbilical cord. Frequently, lesion is embedded within deep seated skeletal muscle of extremities.

Intramuscular myxoma commonly arises within thigh, shoulder or upper arm.

Generally, intramuscular myxoma manifests as a solitary lesion. Commonly, adults between 40 years and 70 years are implicated. A mild female predilection is observed.

Genomic mutations within GNAS1 are significantly encountered within intramuscular myxomas. However, tumefaction is devoid of cogent cytogenetic aberrations.

Multiple myxomas are associated with conditions as Carney' complex or fibrous dysplasia configuring as Mazabraud syndrome. Tumefaction is posited to be engendered from modified fibroblasts.

Cytological examination expounds pauci-cellular smears composed of bland, spindle shaped cells admixed with foamy macrophages disseminated within a finely granular, myxoid stroma and viscous, gelatinous substance.

Grossly, a mucoid or gelatinous, inadequately circumscribed neoplasm appears to infiltrate surrounding tissue perimeter. Tumour magnitude is variable and may extend up 13 centimetres.

Upon microscopy, the hypo-cellular neoplasm is constituted of bland, spindle shaped cells admixed with focal aggregates of histiocytic cells. Vascular articulations are minimal. Lipoblasts are absent. Mitotic figures are absent. Neoplasms may appear as cellular lesions circumscribed by a peripheral collagen capsule. Besides, tumefaction may hyper-cellular or exhibit enhanced vascular structures.

Intervening matrix may be minimally basophilic and pervaded with few spindle shaped cells with ovoid nuclei.
Neoplasm may demonstrate a centric mucinous cyst.

Lesion periphery may infiltrate circumscribing striated muscle or concur with atrophy of skeletal muscle.

Ultrastructural examination exhibits fibroblast-like cells with prominent Golgi bodies, endoplasmic reticulum or intracytoplasmic filaments.

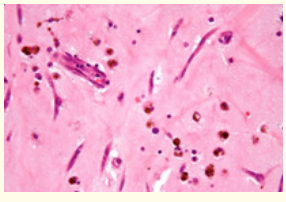

Figure 1: Intramuscular myxoma.

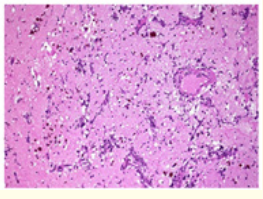

Figure 2: Intrmuscular myxoma.

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].

TNM staging of soft tissue tumours [3,4].

Primary tumour

Head and neck

- TX: Tumour grade cannot be assessed
- T1: Tumour magnitude ≤ 2 centimetre
- T2: Tumour magnitude > 2 centimetres to ≤ 4 centimetres
- T3: Tumour magnitude exceeding > 4 centimetres
- T4: Tumour associated with invasion of adjoining structures
  - T4a: Tumour demonstrating infiltration of orbit, base of skull, dura, central compartment viscera, facial skeleton or pterygoid muscles
  - T4b: Tumour associated infiltration of brain parenchyma, encasement of carotid artery, prevertebral muscle or central nervous system via perineural dissemination

Trunk and extremities

- TX: Tumour grade cannot be assessed
- T0: No evidence of primary tumour
- T1: Tumour magnitude ≤ 5 centimetres in greatest dimension
- T2: Tumour magnitude > 5 centimetres and ≤ 10 centimetres in greatest dimension
- T3: Tumour magnitude > 10 centimetres and ≤ 15 centimetres in greatest dimension
- T4: Tumour magnitude > 15 centimetres in greatest dimension

Abdomen and thoracic viscera

- TX: Tumour grade cannot be assessed
- T1: Tumour is confined to organ of origin
- T2: Tumour extension into circumscribing tissue beyond organ of origin
  - T2a: Tumour infiltrates serosa or visceral peritoneum
  - T2b: Tumour extension beyond serosa or into mesentery
- T3: Tumour infiltrates adjacent organ
- T4: Tumour demonstrates multifocal visceral involvement
  - T4a: Tumour is multifocal and confined to two sites.
  - T4b: Tumour is multifocal and confined to three to five sites
  - T4c: Tumour is multifocal and implicates > 5 sites

Retroperitoneum

- TX: Tumour grade cannot be assessed
- T0: No evidence of primary tumour
- T1: Tumour ≤ 5 centimetres in greatest dimension
- T2: Tumour > 5 centimetres and ≤ 10 centimetres in greatest dimension
- T3: Tumour magnitude > 10 centimetres and ≤ 15 centimetres in greatest dimension
- T4: Tumour magnitude > 15 centimetres in greatest dimension

Orbit

- TX: Tumour grade cannot be assessed
- T0: No evidence of primary tumour
- T1: Tumour magnitude ≤ 2 centimetres in greatest dimension
- T2: Tumour magnitude > 2 centimetres in greatest dimension in the absence of invasion of bony walls or globe
- T3: Tumour of variable magnitude along with invasion of bony walls

- T4: Tumour of variable magnitude along with invasion of globe or periorbital structures, eyelids, conjunctiva, temporal fossa, nasal cavity, paranasal sinuses or central nervous system.

Regional lymph nodes

- NX: Regional lymph nodes cannot be assessed
- N0: Regional lymph node metastasis absent
- N1: Regional lymph node metastasis present

Distant metastasis

- MX: Distant metastasis cannot be assessed
- M0: Distant metastasis absent
- M1: Distant metastasis present
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Relevant prefixes

- m: multiple
- y: adoption of preoperative radiotherapy or chemotherapy
- r: recurrent tumour stage

Spindle shaped cells configuring intramuscular myxoma appear immune reactive to vimentin or CD34. Intervening mucoid matrix may be highlighted by Alcian blue, mucicarmine or colloidal iron stains. Tumour cells appear immune non reactive to S100 protein and desmin. Intramuscular myxoma requires segregation from neoplasms as low grade myxofibrosarcoma, nerve sheath myxoma, focal mucinous degeneration of soft tissue or cutis due to various conditions as cysts, ganglion, mucinosis, myxoedema or nodular fasciitis. Besides, neoplasms with myxoid features as aggressive angiomyxoma, chondrosarcoma, embryonal rhabdomyosarcoma, leiomyoma, leiomyosarcoma, liposarcoma or neurofibroma.

Morphological features as cellular or nuclear atypia, mitotic activity or frequently discerned vascular articulations may not be observed within intramuscular or juxta-articular myxoma and cogent neoplastic discernment may be challenging.

Intramuscular myxoma may be appropriately subjected to comprehensive surgical extermination of the neoplasm. Tumour reoccurrence is exceptional.

Cellular lesions managed with comprehensive surgical eradication demonstrate indolent biological behaviour.

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8. Image 1 Courtesy: Wikipedia.
9. Image 2 Courtesy: Symbiosis online publishing.