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Review Article

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The Sore, Metamorphosed Sinew-Inflammatory Leiomyosarcoma

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Inflammatory leiomyosarcoma is an extremely exceptional, mesenchymal, malignant neoplasm demonstrating smooth muscle differentiation and a predominant inflammatory cell infiltrate infiltrating the tumour parenchyma. Characteristically, inflammatory leiomyosarcoma represents as a low grade neoplasm of smooth muscle origin exemplifying chronic inflammatory cell infiltrate comprised of lymphoid cells and plasma cells. Initially scripted by William Merchant, Eduardo Calonje and Christopher DM Fletcher, inflammatory leiomyosarcoma is established as a distinct entity by World Health Organization (WHO) in 2020. Inflammatory leiomyosarcoma is appropriately discerned with cogent histological evaluation and pertinent immunohistochemistry. Characteristically, inflammatory leiomyosarcoma is comprised of atypical spindle shaped cells permeated with variably eosinophilic cytoplasm. Tumefaction is variably infiltrated with lymphoid cells or histiocytic cells commingled with xanthomatous inflammatory cells. Mitotic activity is variable. Tumour cells appear immune reactive to diverse smooth muscle antigens. Inflammatory leiomyosarcoma predominantly incriminates adults. Peak age of disease emergence is encountered within 3rd decade or 4th decade. Tumefaction may be discerned within adolescents. A male predisposition is encountered [1,2]. Inflammatory leiomyosarcoma manifests as a gradually progressive tumefaction of variable duration which appears confined to deep seated soft tissues. Certain instances delineate clinical symptoms pertaining to an inflammatory and neoplastic origin [1,2]. Of obscure aetiology, inflammatory leiomyosarcoma predominantly exhibits a near-haploid genotype, a feature which appears significant for enunciating precise disease pathogenesis. Inflammatory leiomyosarcoma commonly incriminates deep seated soft tissues of lower limb, trunk or retroperitoneum. Neoplasm may exceptionally be confined to diverse visceral sites [2,3]. Upon gross examination, inflammatory leiomyosarcoma manifests as a well defined neoplasm with tumour magnitude varying from 3 centimetres to 12 centimetres. Cut surface is fleshy and manifests with yellowish, tan or reddish hue [2,3]. Upon microscopy, inflammatory leiomyosarcoma exhibits characteristic histological features as spindle shaped cells incorporated with abundant, eosinophilic cytoplasm. Tumour cells configure a fascicular or storiform pattern of evolution. Cellular aggregates are infiltrated with diffuse inflammatory infiltrate comprised of small lymphocytes, plasma cells and a preponderant component of histiocytic cells. Few neoplasms may depict focal areas of psammomatous calcific bodies. Cellular and nuclear atypia, pleomorphism and mitotic activity is variable [2,3].

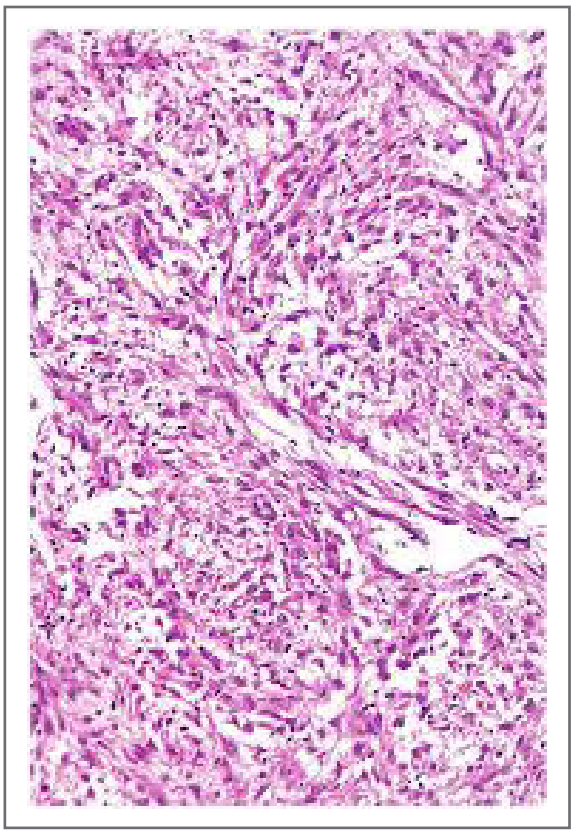

Figure 1: Inflammatory leiomyosarcoma delineating fascicles of spindle shaped cells imbued with abundant, eosinophilic cytoplasm and an infiltration by inflammatory cells as small lymphocytes, plasma cells and innumerable histiocytic cells (6).

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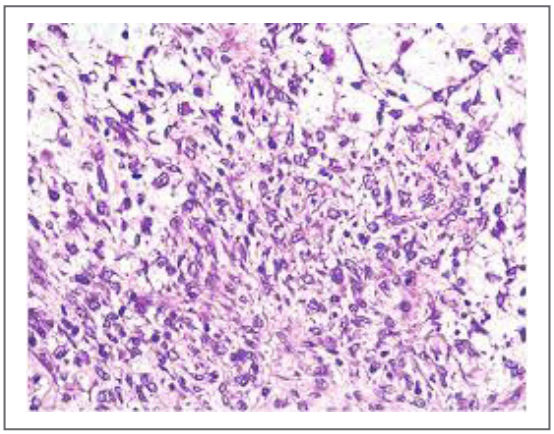

Figure 2: Inflammatory leiomyosarcoma exhibiting bundles of spindle shaped cells permeated with abundant, eosinophilic cytoplasm and an infiltration of inflammatory cells as small lymphocytes, plasma cells or histiocytic cells (7).

TNM staging of uterine leiomyosarcoma as per American Joint Committee on Cancer 8th edition and International Federation of Gynaecology and Obstetrics (FIGO) [3,4].

Primary Tumour

- TX: primary tumour cannot be assessed
- T0: no evidence of primary tumour
- T1(IA): tumour confined to the uterus ~T1a (IA): tumour magnitude ≤ 5 centimetres in greatest dimension ~T1b (IB): tumour magnitude > 5 centimetres in greatest dimension
- T2 (II): tumour extends beyond uterine cavity and appears confined within the pelvis ~T2a (IIA): tumour incriminates adjoining uterine adnexa ~T2b (IIB): tumour incriminates diverse pelvic anatomical tissues
- T3 (III): tumour infiltrates various abdominal soft tissues and visceral structures ~T3a (IIIA): tumour infiltrates adjoining abdominal soft tissues confined to singular site ~T3b (IIIB): tumour infiltrates adjoining abdominal soft tissues within > singular site
- T4 (IVA): tumour infiltrates adjoining viscera as urinary bladder or rectum

Regional Lymph Nodes

- NX: regional lymph nodes cannot be assessed
- N0: regional lymph node metastasis absent
- N0(i+): isolated tumour cells disseminated within regional ymph node(s) ≤ 0.2 millimetre magnitude
- N1 (IIIC): regional lymph node metastasis present

Distant Metastasis

- M0: distant metastasis absent
- M1 (IVB): distant metastasis present excluding sites such as uterine adnexa, pelvic soft tissue or abdominal soft tissues.

International Federation of Gynaecology and Obstetrics (FIGO) staging and grouping of uterine leiomyosarcoma [3,4].

- stage I: T1, N0, M0
- stage IA: T1a, N0, M0
- stage IB: T1b, N0, M0
- stage II: T2, N0, M0
- stage IIIA: T3a, N0, M0
- stage IIIB: T3b, N0, M0
- stage IIIC: T1, T2 or T3, N1, M0
- stage IVA: T4, any N, M0
- stage IVB: any T, any N, M1

Inflammatory leiomyosarcoma appears immune reactive to smooth muscle actin (SMA), desmin or caldesmon [4,5]. Inflammatory leiomyosarcoma requires segregation from several benign and malignant soft tissue tumours principally emerging within deep seated soft tissues or retroperitoneum such as leiomyoma of deep seated soft tissue, conventional leiomyosarcoma, pleomorphic liposarcoma or undifferentiated sarcoma [4,5]. Upon computerized tomography (CT), inflammatory leiomyosarcoma emerges as a well circumscribed neoplasm with homogenous image enhancement [4,5]. Magnetic resonance imaging (MRI) manifests a lobulated neoplasm. Upon T1 weighted magnetic resonance imaging, tumefaction represents with mildly hyper-intense image enhancement. Upon T2 weighted magnetic resonance imaging, neoplasm manifests with significant image enhancement. Upon administration of gadolinium contrast with T1 weighted imaging, diffuse image enhancement is encountered. Upon diffusion weighted magnetic resonance imaging (DWI), moderately restricted image enhancement is observed [4,5]. The exceptionally discerned inflammatory leiomyosarcoma appears devoid of precise elucidation regarding disease monitoring and prognostic outcomes. Few neoplasms are associated with distant metastases [4,5]. Tumefaction delineating near-haploidization manifest with minimally aggressive biological behaviour and superior prognostic outcomes, in contrast to diverse variants of leiomyosarcoma [4,5].

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