

Transitional and Amalgamated-Mesenchymal Hamartoma of Chest Wall

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Mesenchymal hamartoma of chest wall configures as an extremely exceptional, benign neoplasm confined to the chest wall. Alternatively, lesion may be contemplated as a non neoplastic, developmental anomaly.

Mesenchymal hamartoma of chest wall is preponderantly confined to the ribs wherein few lesions represent with multifocal disease [1]. Characteristically, neoplasm occurs within neonates or infants [2,3].

Clinically, tumefaction represents as a mass or deformity confined to thoracic wall. Additionally, symptoms as respiratory distress may ensue [2,3].

Grossly, a well circumscribed, lobulated lesion of tan to reddish hue is observed [2,3].

Upon microscopy, neoplasm expounds alternating cellular zones constituted of islands of cartilage admixed with foci of primitive, proliferating mesenchymal cells. Pre-eminently heterogeneous, mesenchymal hamartoma is comprised of cartilaginous component commingled with mesenchymal cells [3,4].

Tumefaction appears constituted of an admixture of diverse mesenchymal tissues as adipocytes, smooth muscle fibres, segments of nervous system or ganglion cells. Entangled smooth muscle tissue represents with myofibrillar disarray [3,4].

Focal cystic change, calcification and ossification may be encountered. Generally, tumour expansion is observed. Focal bone formation may occur. Cystic spaces pervaded with haemorrhagic content may be discerned, recapitulating foci of secondary aneurysmal bone cyst-like areas [3,4].

Nasal glands may be observed within site specific tumours [3,4].

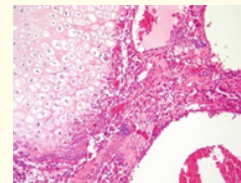


Figure 1: Mesenchymal hamartoma of chest wall composed of alternating cartilaginous areas and primitive, proliferating mesenchymal cells. Cavities filled with haemorrhagic content are encountered [11].

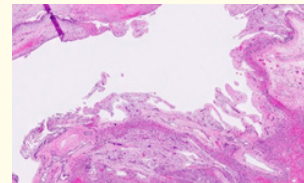


Figure 2: Mesenchymal hamartoma of chest wall displaying well defined cartilaginous areas intermingled with primitive, proliferating mesenchymal cells. Cavities filled with haemorrhagic content are enunciated [12].

- T3a: Tumour is ≤ 8 centimetre magnitude
- T3b: Tumour is > 8 centimetre magnitude
- T4: Tumour is confined to three pelvic foci or extends beyond sacroiliac joint
 - T4a: Tumour invades sacroiliac joint and incriminates sacral neuro-foramina
 - T4b: Tumour encases regional vasculature or impedes vascular outflow

Regional lymph nodes

- NX: Regional lymph nodes cannot be assessed
- N0: Regional lymph node metastasis absent
- N1: Regional lymph node metastasis present which is exceptional in a primary bone sarcoma

Distant metastasis

- M0: Distant metastasis absent
- M1: Distant metastasis present
 - M1a: Metastasis into pulmonary parenchyma
 - M1b: Metastasis into various bones or viscera

Mesenchymal hamartoma appears immune reactive to CD117. Constituent smooth muscle tissue may be suitably immuno-stained with anti-c-KIT antibodies [5,6].

Mesenchymal hamartoma of chest wall requires segregation from neoplasms as primary Ewing sarcoma or distant metastasis into thoracic wall from malignant conditions as neuroblastoma, leukaemia or lymphoma. Besides, benign rib lesions as fibrous dysplasia, haemangioma and Langerhans cell histiocytosis or histiocytosis X, devoid of internal ossification require demarcation [5,6].

A concordance of histological features and imaging manifestations is necessitated for appropriate neoplastic discernment [7,8].

Upon radiography, tumour manifests as a well circumscribed lesion constituted of alternating solid and cystic components. Neoplasm may depict multiple fluid-fluid levels. Destruction of singular or multiple ribs may be exemplified [7,8].

Plain radiographs may expound a well defined, partially calcified tumour mass confined to singular or multiple ribs [7,8].

T1 and T2 weighted magnetic resonance imaging (MRI) depicts heterogeneous signal intensity. Cavities impregnated with haemorrhagic content, simulating secondary aneurysmal bone cyst-like zones may be frequently encountered [7,8].

Mesenchymal hamartoma of chest wall emerges as a self limited lesion. However, rapid tumour progression may ensue. The pre-eminently non neoplastic lesion may infrequently undergo malignant transformation. Surgical manoeuvres as en bloc tumour resection appear curative [9,10]. However, complications as significant residual chest wall defect may frequently induce scoliosis [9,10].

Symptomatic mesenchymal hamartoma of chest wall may be appropriately alleviated by surgical eradication. Additionally, conservative management may be adopted for managing miniature, asymptomatic mesenchymal hamartoma of chest wall which may undergo spontaneous resolution. Prognostic outcomes are superior [9,10].

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11. Image 1 Courtesy: Springer link.
12. Image 2 Courtesy: Pathology outlines.