MEDWIN PUBLISHERS
Committed to Create Value for Researchers

Cell & Cellular Life Sciences Journal
ISSN: 2578-4811

The Unctuous Unconformity-Pleomorphic Liposarcoma

Anubha B*

AB Diagnostics, Panjab University, India

***Corresponding author:** Anubha Bajaj, A.B. Diagnostics, A-1, Ring Road , Rajouri Garden , New Delhi - 110027, India, Email: anubha.bajaj@gmail.com

Editorial

Volume 8 Issue 1

Received Date: April 24, 2023

Published Date: May 03, 2023

DOI: 10.23880/cclsj-16000178

Keywords: Pleomorphic Liposarcoma; Epithelioid Cells; Myxoid Liposarcoma; Ultrasonography

Abbreviations: WHO: World Health Organization; CT: Computerized Tomography; MRI: Magnetic Resonance Imaging.

Editorial

Liposarcoma was initially scripted by Rudolph Virchow in 1857. Subsequently, Franz Michael Enzinger and Donald J Winslow described pleomorphic liposarcoma and diverse subtypes of liposarcoma in 1962. Epithelioid pleomorphic liposarcoma was pre-eminently denominated by Markku Miettinen and Franz Michael Enzinger in 1999. Pleomorphic liposarcoma represents as an exceptional, high grade, malignant neoplasm emerging from adipocytic tissues. Characteristically, the high grade pleomorphic liposarcoma is constituted of variably quantifiable pleomorphic lipoblasts with absent foci of well differentiated liposarcoma or diverse lineages of cellular differentiation. The neoplasm is devoid of areas which simulate atypical lipomatous tumour / well differentiated liposarcoma or diverse foci of mesenchymal differentiation.

As per contemporary World Health Organization (WHO) classification of soft tissue and bone tumours, variable quantities of pleomorphic lipoblasts configuring pleomorphic liposarcoma delineate distinctive tumour morphology as

- pleomorphic spindle shaped cell sarcoma
- neoplasm with myxofibrosarcoma-like morphology
- epithelioid cell morphology constituted of carcinoma-like epithelioid cells
- Singular composition of pleomorphic lipoblasts [1,2].

Cytogenetic and molecular evaluation of the neoplasm exhibits lack of MDM2 genetic alterations. Of obscure aetiology, pleomorphic liposarcoma commonly emerges as a pleomorphic spindle cell sarcoma or a variant pleomorphic epithelioid cell sarcoma configured of variable quantities of pleomorphic lipoblasts. Pleomorphic liposarcoma manifests < 5% of lip sarcomas and ~20% of pleomorphic sarcomas [1,2]. Peak age of disease incidence is encountered within seventh decade. Incrimination of paediatric subjects is extremely exceptional. A mild male predominance is observed [1,2]. Majority (~66%) of tumours arise within lower extremities followed in frequency by upper extremities. Sites such as trunk, thoracic or abdominal wall, retroperitoneum and spermatic cord are infrequently incriminated [1,2].

Neoplasm may be exceptionally confined to mediastinum, heart, pulmonary parenchyma, pleura, breast, scalp, colon or orbit. Pleomorphic liposarcoma manifesting as a primary bone tumour is extremely infrequent. However, singular dermal lesions are documented. Generally, neoplasm is confined to deep seated soft tissue or subcutaneous adipose tissue [1,2].

Pleomorphic liposarcoma is devoid of pathognomonic, reoccurring molecular alterations. An absence of supernumerary ring chromosomes with amplification of MDM2 / CDK4 genes is characteristic and aids in distinction from high grade, dedifferentiated liposarcoma with pleomorphic features [2,3]. Tumour cells display complex karyotypes. Molecular profile of pleomorphic liposarcoma simulates profile of various pleomorphic sarcomas as high grade myxofibrosarcoma or undifferentiated pleomorphic sarcoma, in contrast to well differentiated or dedifferentiated liposarcoma, thereby indicating a distinctive molecular pathway of neoplastic emergence [2,3]. Majority of somatic genetic mutations occur within TP53 or NF1 gene. Amplification of MDM2 nuclear gene is absent. Myxoid liposarcoma may enunciate epithelioid neoplastic cells demonstrating pleomorphic morphological features.

Nevertheless, characteristic reoccurring chromosomal translocation t (12; 16) (q13; p11) and FUS-DDIT3 genetic rearrangement may be encountered [2,3]. Pleomorphic liposarcoma frequently manifests as a rapidly progressive, painless tumefaction. However, pain or clinical symptoms contingent to tumour location or organ displacement may be uncommonly encountered [2,3]. Upon gross examination, an enlarged, firm, well demarcated, non-encapsulated, multinodular, white/yellow tumour is encountered. Alternatively, poorly defined, infiltrative, yellowish white tumours may be encountered. Median tumour magnitude varies from 8 centimetres to 10 centimetres. Cut surface depicts foci of myxoid alterations, tumour necrosis and haemorrhage. Focal or disseminated zones of cystic degeneration may be encountered [2,3]. Upon frozen section, morphological features may be typically non diagnostic. Generally, high grade sarcoma can be expounded on degree of cytological atypia, mitotic activity and presence or absence of tumour necrosis [2,3]. Upon cytological examination, a high grade neoplasm is composed of pleomorphic spindle shaped which appear admixed with occasional lip oblasts [2,3]. Upon microscopy, variable proportion of pleomorphic lipoblasts appear disseminated within a high grade, pleomorphic, undifferentiated sarcoma. The well circumscribed, non-encapsulated tumour with infiltrative perimeter may display significant morphological overlap with neoplasms as myxofibrosarcoma, undifferentiated pleomorphic sarcoma and high grade dedifferentiated lip sarcoma [3,4]. Pleomorphic liposarcoma is composed of high grade neoplastic cells commingled with varying quantities of pleomorphic, bizarre, multinucleated tumour cells [3,4]. Morphologically, pleomorphic liposarcoma demonstrates infiltrative tumour perimeter, varying proportion of pleomorphic lipoblasts, an intermingling of non lipogenic, undifferentiated, pleomorphic sarcoma comprised of spindle shaped cells and multinucleated giant cells, myxofibroma-like areas in ~50% neoplasms, focal tumour necrosis in~50% tumefaction and epithelioid tumour cells in ~ 25% tumours. Tumour cells delineate subtle to prominent cytoplasmic vacuolization [3,4]. Classic lipoblasts with scalloped nuclei may be discerned. Besides, signet ring lipoblasts appear disseminated within the tumour parenchyma in concurrence with malignant cells [3,4]. Epithelioid pleomorphic liposarcoma (~25%) requires differentiation from diverse high grade tumours demonstrating epithelioid morphology as vascular tumours, malignant melanoma, carcinoma or various epithelioid sarcomas, which may be challenging [3,4]. Tumour necrosis is frequently encountered. Mitotic activity is significant with a median of 25 mitotic figures/10 high power fields. Phagocytosed neutrophils within giant cells (emperipolesis-like) and nonspecific, extracellular or intracellular, eosinophilic hyaline droplets may be discerned [3,4]. Upon ultrastructural examination, tumour cells depict abundant, coalescing lipid droplets along with numerous cytoplasmic organelles Figures 1 & 2, [5].

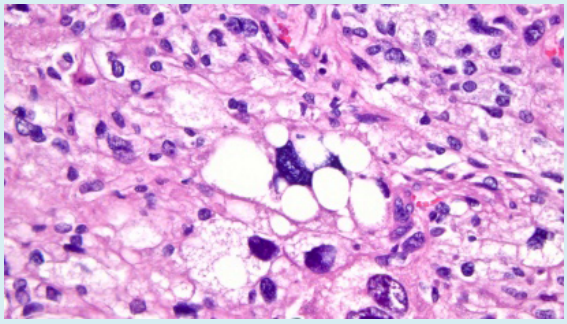

Figure 1: Pleomorphic liposarcoma demonstrating variable quantities of pleomorphic lipoblasts admixed with high grade, undifferentiated, pleomorphic sarcoma composed of spindle shaped cells with hyperchromatic nuclei. Foci of necrosis and prominent mitotic activity are seen [6].

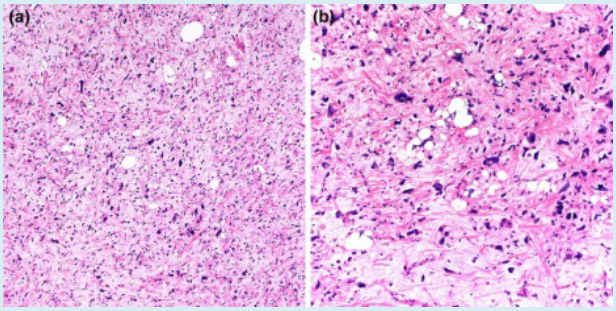

Figure 2: Pleomorphic liposarcoma enunciating variable quantities of pleomorphic lipoblasts commingled with high grade, undifferentiated, pleomorphic sarcoma composed of spindle shaped cells with hyperchromatic nuclei. Foci of necrosis and prominent mitotic activity are observed [7].

TNM Staging of Pleomorphic Liposarcoma [3,4]

Primary Tumour

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

Regional Lymph Nodes

- N0:Regional lymph node metastasis absent
- N1:Regional lymph node metastasis present

Distant Metastasis

- M0: Distant metastasis absent
- M1: Distant metastasis present

Histologic Grade of Pleomorphic Lipsarcoma [3,4]

- GX: Tumour grade cannot be assessed
- G1: Tumour differentiation, mitotic count and necrosis quantified between 2 and 3
- G2: Tumour differentiation, mitotic count and necrosis quantified between 4 and 5
- G3: Tumour differentiation, mitotic count and necrosis quantified between 6,7 or 8

Tumour Stage of Pleomorphic Sarcoma [3,4]

- Stage IA: T1, N0,M0, GX, G1
- Stage IB: T2, N0, M0, GX,G1 OR T3, N0, M0, GX, G1 OR T4, N0,M0,GX,G1
- Stage II: T1, N0, M0,G2, G3
- Stage IIIA: T2, N0, M0, G2,G3
- Stage IIIB: T3,N0,M0,G2,G3 OR T4, N0,M0, G2,G3 OR any T, N1, M0, any G
- Stage IV: any T, any N, M1, any G.

Pleomorphic liposarcoma appears immune reactive to vimentin, S100 protein, smooth muscle actin, CD34, keratin or desmin. Epithelioid variant of pleomorphic liposarcoma is immune reactive to diverse keratins and Melan A. Pleomorphic liposarcoma appears non-reactive to MDM2 and CDK4 [4,5]. Pleomorphic liposarcoma requires segregation from neoplasms such as dedifferentiated liposarcoma, metastatic carcinoma, undifferentiated pleomorphic sarcoma and various high grade pleomorphic sarcomas, myxoid pleomorphic liposarcoma, myxoid liposarcoma, fibrosarcoma, myxofibrosarcoma, pleomorphic lipoma / spindle cell lipoma or pleomorphic rhabdomyosarcoma [4,5]. Pleomorphic liposarcoma is appropriately discerned with cogent histological examination of tissue samples obtained with core needle biopsy or surgical excision [4,5].

Specific biochemical or haematological abnormalities are absent. Imaging is optimal in discerning

- configuration, location and tumour magnitude
- tumour perimeter
- quantifiable solid and lipoid component
- Spatial arrangement of neoplasm in concurrence with diverse organs and/or anatomic structures [4,5].

Upon imaging, pleomorphic liposarcoma manifests as a nonspecific soft tissue tumefaction with heterogeneous foci of tumour necrosis and haemorrhage. In contrast to diverse liposarcomas, mature adipose tissue component is minimal or absent. Displacement of adjacent organs or abutting soft tissue may ensue. Ultrasonography depicts a multi-lobulated neoplasm with irregular to minimal echogenicity [4,5]. Computerized tomography (CT) exhibits a nonspecific, heterogeneous mass with soft tissue density simulating or isodense to surrounding skeletal muscle and streaky foci of image enhancement. Magnetic resonance imaging (MRI) displays a soft tissue tumefaction with an image enhancing, fibrous component [4,5]. Upon T1 weighted magnetic resonance imaging, a heterogeneous, hypo-intense neoplasm is encountered. Upon T2 weighted magnetic resonance imaging, a heterogeneous, hyper-intense tumour with signal intensity minimally decreased or akin to circumscribing adipose tissue is enunciated. Upon T1 weighted magnetic resonance imaging with gadolinium contrast, heterogeneous image enhancement is delineated [4,5]. Pleomorphic sarcoma is appropriately treated with radical surgical resection and extermination of wide, tumour free tissue perimeter. Alternatively, amputation of incriminated limb may be adopted. Postoperative radiotherapy and adjuvant chemotherapy appear beneficial [4,5]. Cogent surgical resection of pleomorphic liposarcoma may be amalgamated by precise chemotherapy with combined ifosfamide and doxorubicin. Alternatively, neoadjuvant chemotherapy may be optimally employed [4,5]. Pleomorphic sarcoma demonstrates an aggressive clinical course. Neoplasm is frequently associated with localized tumour reoccurrence in ~30% to 50% instances. Distant metastasis ensues in ~50% tumours. Tumefaction exhibits a 5 year overall survival of ~60%. Commonly, distant metastasis ensues into pulmonary parenchyma. However, the pleura, hepatic parenchyma and bone may be incriminated [4,5].

Factors contributing to inferior prognostic outcomes emerge as

- centric tumour location
- enhanced depth of neoplasm
- elevated tumour magnitude
- Elevated mitotic count [4,5].

References

1. Gjeorgjievski SG, Thway K, Dermawan JK, John I, Fisher C, et al. (2022) Pleomorphic Liposarcoma: A Series of 120 Cases with Emphasis on Morphologic Variants. Am J Surg Pathol 46(12): 1700-1705.

2. Ciliberti V, Cretella P, Zeppa P, Caputo A (2022) Pleomorphic liposarcoma with liver metastasis diagnosed by combined fine-needle aspiration cytology and core-needle biopsy. Diagn Cytopathol 50(1): 28-31.

3. Robles Tenorio A, Solis Ledesma G (2022)

Undifferentiated Pleomorphic Sarcoma. Stat Pearls.

4. Gruber KM, Vasudevan SS, Janus JR (2022) Pleomorphic Liposarcoma of the Parapharyngeal Space: A Rare Case Report. Ear Nose Throat J 1455613221148312.

5. Gencarelli P Jr, Hali A, Aurup J (2022) Use of Ultrasound for Evaluation of a Large Undifferentiated Pleomorphic Sarcoma in the Lower Extremity of an Elderly Male. Cureus 14(10): 30153.

6. Image 1 Courtesy: Pathology outlines.

7. Image 2 Courtesy: Research gate.