ACTA SCIENTIFIC CLINICAL CASE REPORTS

Volume 5 Issue 2 February 2024

Editorial

Jejune and Eclectic-Pleuropulmonary Blastoma

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Received: November 09, 2023

Published: January 01, 2024

Pleuropulmonary blastoma is an exceptionally encountered, primary pulmonary neoplasm which may incriminate diverse thoracic subsites. Frequently, tumefaction is associated with cystic pulmonary lesions. Initially scripted as a distinct entity in 1988, the primitive neoplasm may emerge within pulmonary parenchyma, mediastinum or pleura. Pleuropulmonary blastoma is additionally designated as neuroblastoma, mesenchymal cystic hamartoma, cystic mesenchymal hamartoma, pulmonary rhabdomyosarcoma, rhabdomyosarcoma in lung cyst or paediatric pulmonary blastoma. Commonly, tumefaction is enunciated within paediatric population.

Pleuropulmonary blastoma configures distinct subtypes representing a morphological continuum from minimally malignant to predominantly malignant lesions denominated as

- Type I demonstrates a multi-cystic lesion with attenuated walls wherein the lesion appears concordant with congenital pulmonary airway malformation (CPAM) type IV
- Type II exhibits thickened areas or nodules amalgamated within the cystic lesion
- Type III is constituted of solid tumour mass.

Neoplasm emerges within the paediatric population, predominantly (90%) within subjects up to 2 years of age. Pleuropulmonary blastoma type I frequently arises in infancy up to 2 years whereas type II and type III may appear beyond 2 years. Lesions commonly incriminate pulmonary parenchyma, mediastinum or pleura [1,2].

Pleuropulmonary blastoma is posited to arise from primitive mesenchymal cells, reminiscent of stem cells.

Majority of incriminated subjects delineate genetic mutation within DICER1 gene. Pleuroplumonary blastoma is accompanied by pleuropulmonary blastoma family tumour and dysplasia syndrome in nearly one third (33%) of tumefaction [1,2].

Incriminated paediatric population exhibits cogent clinical symptoms as dyspnoea. Diverse respiratory conditions as persistent pneumonitis, cough or atelectasis of implicated lung may concur. An estimated 10% lesions may represent with multi-locular, cystic nephroma like lesions. Nevertheless, Wilms tumour like lesions are extremely exceptional [2,3].

Cytological examination expounds cellular smears composed of disseminated singular cells and cohesive cellular aggregates. Tumour cells appear uniform, miniature and spherical with a bluish hue. Tumour cells appear devoid of cytoplasm and characteristically exemplify spherical to elliptical nuclei with nuclear moulding and dark, fine nuclear chromatin [2,3].

Malignant tumour giant cells are permeated with abundant cytoplasm, dispersed nuclear chromatin and exhibit an indistinct cellular perimeter. Few malignant tumour cells appear spindle shaped, are pervaded with eosinophilic cytoplasm and demonstrate irregular nuclear membrane with elevated nucleocytoplasmic ratio. Cellular rosettes or glandular articulations appear absent [3,4].

Grossly, pleuropulmonary blastoma type I appears multi-cystic with attenuated walls. Lesion is situated within peripheral pulmonary parenchyma.

Pleuropulmonary blastoma type II articulates an admixture of solid and cystic tumour areas characteristically delineating variably thickened or nodule-like zones.

Pleuropulmonary blastoma type III is typically comprised of a solid, well circumscribed, mucoid, grey/white or tan tumefaction adherent to superimposed pleura. An entire lobe or lung may be incriminated. Friable tumour zones delineate foci of necrosis and haemorrhage [4,5].

Upon microscopy, lesion denominates distinct architecture as

- Pleuropulmonary blastoma type I is a multi-cystic structure with attenuated walls and appears confined to peripheral parenchyma.
- Pleuropulmonary blastoma type II configures an admixture of solid and cystic tumour pattern along with variably thickened or nodule-like zones.
- Pleuropulmonary blastoma type III emerges as a heterogeneous tumefaction composed of singular element or an amalgamation of
  - Primitive, blastema-like miniature cells pervaded with hyperchromatic nuclei with elevated nucleocytoplasmic ratio. Mitotic figures are abundant.
  - Spindle shaped or ovoid cells embedded within a myxoid stroma nodules of immature, malignant chondroid constituents

- Isolated, singular cells or clusters of enlarged anaplastic cells pervaded with pleomorphic nuclei or eosinophilic hyaline bodies may be discerned. Atypical mitotic figures are exemplified [4,5].

Neoplastic cells appear intensely immune reactive to vimentin and focally immune reactive to CD117or alpha 1 antitrypsin. Weak immune reactivity to CD99 is observed. Surface epithelium appears immune reactive to cytokeratin. Rhabdom oblasts and primitive cells appear immune reactive to muscle specific actin (MSA) and desmin [6,7]. Pleuropulmonary blastoma appears immune non reactive to epithelial membrane antigen (EMA), myogenic, S100 protein, glial fibrillary acidic protein (GFAP), neuron specific enolase (NSE), α fetoprotein (AFP), chromogranin and synaptophysin [6,7].

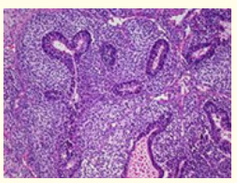

Figure 1: Pleuropulmonary blastoma exhibiting solid areas and multiple cystic structures with attenuated walls surrounded by a primitive, blastema-like stroma with hyperchromatic nuclei and enhanced nucleocytoplasmic ratio [8].

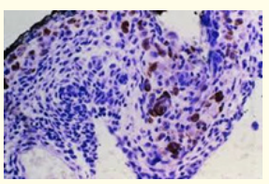

Figure 2: Pleuropulmonary blastoma delineating solid areas commingled with multi-cystic structures with attenuated walls and a circumscribing primitive, blastema-like stroma with hyperchromatic nuclei [9].

Gene	Molecular alteration
EGFR	Mutation (~35%)
KRAS	Mutation (25%)
HER2	Mutation (~6.7%), amplification (~22%), overexpression (~23%)
ALK	Chromosomal rearrangement (~8%)
MET	Amplification (~4%), mutation (~4%)
BRAF	Mutation (~5%)
RET	Chromosomal rearrangement (~2%)
ROS1	Chromosomal rearrangement (~1.7%)
NTRK	Gene fusions (~1%)

Table 1: Driver mutations within non small cell carcinoma lung [5].

Pleuropulmonary blastoma requires segregation from neoplasms such as foetal lung interstitial tumour (FLIT), malignant peripheral nerve sheath tumour, malignant teratoma, mesothelioma, monophasic synovial sarcoma, peripheral neuroectodermal tumour (PNET), undifferentiated sarcoma or congenital cystic adenomatoid malformations. Besides, type III pleuropulmonary blastoma necessitates distinction from embryonal rhabdomyosarcoma. Type I pleuropulmonary blastoma mandates demarcation from enlarged bronchogenic cyst/pulmonary cyst [6,7].

Computerized tomography (CT) or magnetic resonance imaging (MRI) may be suitably adopted to discern pleuropulmonary blastoma. However, surgical tissue sampling with cogent histopathological evaluation is optimal and recommended for appropriate neoplastic discernment.

Upon radiography, tumour mass appears situated upon pleural surface or incriminates peripheral pulmonary parenchyma. Commonly, right lung is implicated [6,7].

Pleuropulmonary blastoma may be treated by multimodal therapeutic strategies. Surgical intervention may be beneficially adopted. Additionally, chemotherapy or radiation therapy appears advantageous.

Appropriate amalgamation of precise therapeutic strategies is contingent to subtype of lesion and proportionately aggressive clinical course. Prognostic outcomes of pleuropulmonary blastoma, especially type II and type III appear adverse. Disease reoccurrence is frequent. Distant metastasis are commonly encountered within brain and bone [6,7].

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8. Image 1 Courtesy: UPMC Pathology.
9. Image 2 Courtesy: American cancer society journals.