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Editorial

Motley and Callow-Pulmonary Blastoma

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Pulmonary blastoma emerges as a component of exceptionally discerned, heterogeneous group of pulmonary neoplasms characteristically configured of immature epithelial cells and mesenchymal elements. Tumefaction simulates embryonic pulmonary tissue, especially foetal lung with gestational maturity of 10 weeks to 15 weeks. Contingent to preponderant tissue subtype, pulmonary blastoma is categorized as ~well differentiated foetal adenocarcinoma or monophasic pulmonary blastoma which is singularly comprised of epithelial cellular component. ~classic biphasic pulmonary blastoma which is constituted of epithelial and mesenchymal components, is commonly exemplified and contemplated as a spectrum of sarcomatoid carcinomas. ~pleuropulmonary blastoma which characteristically delineates singular mesenchymal component.

The contemporary World Health Organization (WHO) classification segregates well differentiated foetal adenocarcinoma comprised singularly of epithelial component and pleuropulmonary blastoma constituted singularly of mesenchymal component from the biphasic pulmonary blastoma.

The infrequently encountered pulmonary blastoma configures ~1% of malignant primary pulmonary carcinomas. Tumefaction exemplifies biphasic cellular components of admixed well differentiated glandular tissue and blastematous stroma which appear as commingled epithelial cells and primitive stroma [1,2]. Apart from epithelial element, pulmonary blastoma is configured of mesenchymal stroma which may articulate foci of chondrosarcoma, rhabdomyosarcoma, osteosarcoma or yolk sac tumour. Neoplasm manifests as a sarcomatoid pulmonary carcinoma with an aggressive clinical course [1,2]. Tumefaction represents as a solitary, enlarged, well circumscribed, non-encapsulated tumour mass confined to peripheral pulmonary lobes. Majority (80%) of lesions are discerned in adults although no age of disease emergence is exempt. Generally, subjects within fourth decade to fifth decade are incriminated. An equivalent gender predilection is encountered. Alternatively, an intense female predominance may be observed, possibly due to oestrogen receptors which are expressly activated by β-catenin Mean tumour diameter upon primary neoplastic discernment emerges between 7 centimetres to10 centimetres. Pulmonary blastoma commonly arises as a unilateral neoplasm and incriminates superior pulmonary lobes, although no lobes are exempt from disease emergence [2,3].

Epithelial and mesenchymal components configuring pulmonary blastoma are posited to be derived from singular clone of pluripotent, precursor cells [2,3]. Neoplasm depicts chromosomal mutations within TP53 and CTNNB1 genes. Exceptionally, genomic mutations within EGFR, MET, BRAF and DICER1 genes may ensue [2,3]. Pulmonary blastoma exhibits exon 3 of CTNNB1 missense genetic mutation, a feature which is associated with configuration of morule. Around ~40% neoplasms exemplify chromosomal mutation within TP53.

Of obscure aetiology, majority (~80%) of neoplasms are associated with cigarette smoking [2,3].

An estimated 40% neoplasms are asymptomatic and discovered incidentally upon plain radiography. Clinical symptoms appear non specific. Commonly encountered clinical symptoms emerge as cough, haemoptysis, chest pain, dyspnoea or pyrexia of unknown origin [2,3].

Grossly, majority of neoplasms incriminate peripheral pulmonary parenchyma. Tumour appears enlarged, well circumscribed and non encapsulated. Average tumour magnitude is articulated at 9.1 centimetres. Cut surface exhibits a cystic, lobulated tumefaction demonstrating foci of necrosis and haemorrhagic degeneration [3,4].

Upon microscopy, neoplasm is configured of glandular articulations and branching tubules, simulating foetal lung. Tubules and glandular structures are layered with pseudostratified, columnar epithelial cells pervaded with translucent cytoplasm, hypo-nuclear vacuoles or sub-nuclear and supra-nuclear glycogen vacuoles. Besides, tubular glandular cells may be discerned [3,4]. Epithelial component is preponderantly composed of tubules articulated of non ciliated, glycogen rich cells. Aforesaid tubules appear reminiscent of pseudo-glandular stage of foetal lung and is characteristically immune non reactive to cytokeratin. Nuclear atypia is minimal. Circumscribing stroma is loose and comprised of undifferentiated mesenchymal cells demonstrating variable cellular atypia along with foci of heterologous differentiation as skeletal muscle, cartilage or bone [3,4]. An estimated two thirds (~67%) neoplasms exhibit morules and neuroendocrine cells. Besides, tumefaction may enunciate a component of germ cell tumour or malignant melanoma. Ultrastructural examination exemplifies nuclear filaments and elements rich in biotin, which contribute to nuclear clearing [3,4].

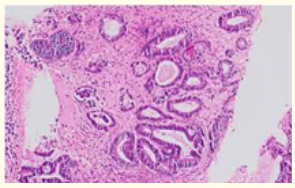

Figure 1: Pulmonary blastoma composed of epithelial and mesenchymal components with tubular structures lined by pseudostratified epithelium surrounded by undifferentiated stroma [7].

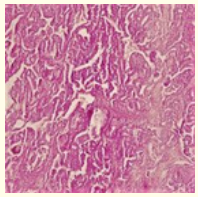

Figure 2: Pulmonary blastoma constituted of biphasic epithelial and mesenchymal constituents with glandular articulated layered by pseudostratified epithelium encompassed by undifferentiated mesenchyme and fibrotic stroma [8].

	CTNNB1 mutation	p53 mutation	DICER1 mutation
Foetal adenocarcinoma	+	-	-
Pulmonary blastoma	+	+	Exceptional
Carcinosarcoma	-	+	-
Pleuropulmonary blastoma	-	+	+

Table 1: Genetic mutation within pulmonary carcinoma [3,4].

Neoplastic cells configuring glandular component appear immune reactive to nuclear β catenin, CK7, AE1/AE3, thyroid transcription factor-1 (TTF-1) carcinoembryonic antigen (CEA), synaptophysin and focally immune reactive to chromogranin. Blastemal stromal component appears immune reactive to nuclear β catenin, vimentin and smooth muscle actin (SMA). Additionally, tumour cells appear immune reactive to muscle specific actin (MSA), vimentin, neuron specific enolase, α-fetoprotein (AFP), carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA) [5,6]. Tumour cells configuring the glandular component appear immune non reactive to vimentin. Blastemal stromal component appears immune non reactive to CAM5.2 and thyroid transcription factor-1(TTF-1) [5,6].

Pulmonary blastoma requires segregation from neoplasms such as foetal type adenocarcinoma, pleuropulmonary blastoma, biphasic synovial sarcoma, pleural fibroma, hamartoma, diverse primary pulmonary carcinomas, distant pulmonary metastases from vari-

ous primaries or carcinosarcoma comprised of non small cell carcinoma as a squamous cell carcinoma or adenocarcinoma admixed with heterologous sarcomatous component. Pulmonary blastoma can be appropriately diagnosed upon histological evaluation of surgical tissue samples and cogent immunohistochemistry. Preoperative tissue samples exhibit a neoplasm composed of dual cellular articulations [5,6]. Optimally, trans-bronchial biopsy may be employed to obtain cogent tissue samples in ~25% neoplasms as pulmonary blastoma is predominantly located within peripheral pulmonary lobes. Serum α-fetoprotein (AFP) and carcinoembryonic antigen (CEA) levels may be elevated [5,6].

Upon radiography, pulmonary blastoma exemplifies an enlarged, solitary, well circumscribed tumour mass confined to peripheral pulmonary lobes or displacing the mediastinum. Neoplasm may exhibit variable image enhancement. Tumour necrosis may be encountered. Computerized tomography (CT) exhibits dense, vesical constituents delineating variable contrast uptake. An estimated 25% neoplasms represent with endobronchial neoplastic growth. Pleural invasion is exceptional.

Surgical extermination is preferentially adopted to alleviate the typically well demarcated, peripheral pulmonary blastoma [5,6]. Outcomes of surgical intervention are contingent to ~tumour magnitude ~pleural invasion ~regional lymph node metastasis ~associated comorbid conditions [5,6]. Adoption of neoadjuvant chemotherapy or radiation therapy is accompanied by limited response to therapy. Adjuvant chemotherapy with cisplatin in combination with etoposide may ameliorate disease associated survival. Besides, multi-kinase inhibitor therapy appears efficacious [5,6]. Pulmonary blastoma is associated with inferior prognostic outcomes wherein 2 year survival appears at ~34% and 5 year survival emerges at ~16%. Average survival following surgical intervention is 33 months. Surgical manoeuvers as limited lobectomy are associated with superior proportionate survival, in contrast to procedures as pneumonectomy, possibly on account of decimated primary tumour burden [5,6]. Factors contributing to inferior prognostic outcomes are configured of ~tumour reoccurrence ~occurrence of biphasic tumefaction ~tumour magnitude > 5 centimetres ~preliminary tumour relapse within 12 months of commencement of therapy ~regional lymph node involvement ~distant metastasis upon initial disease representation. Distant metastasis upon initial disease representation appears in ~43% subjects and is preponderantly confined to brain, pleura, mediastinum, diaphragm or hepatic parenchyma [5,6].

Bibliography

1. Xie Y., *et al*. "Pulmonary blastoma treatment response to anti-PD-1 therapy: a rare case report and literature review". *Frontiers in Oncology* 13 (2023): 1146204.

2. Budzik MP., *et al*. "Pulmonary blastoma in a pregnant woman: a case report and brief review of the literature". *BMC Pulmonary Medicine* 22 (2020): 8.

3. Pooniya S., *et al*. "Classic biphasic pulmonary blastoma (CBPB): a rare primary pulmonary malignancy". *BMJ Case Report* 14.8 (2020): e244151.

4. Bu X., *et al*. "Epidemiological features and survival outcomes in patients with malignant pulmonary blastoma: a US population-based analysis". *BMC Cancer* 20.1 (2020): 811.

5. Le Caer H., *et al*. "Classic biphasic pulmonary blastoma: A case report and review of the literature". *Critical Reviews in Oncology/Hematology* 125 (2018): 48-50.

6. Zhao YY., *et al*. "A retrospective analysis of the clinicopathological and molecular characteristics of pulmonary blastoma". *OncoTargets and Therapy* 9 (2016): 6915-6920.

7. Image 1 Courtesy: Turkish Journal of Pathology.

8. Image 2 Courtesy: BMC pulmonary medicine.