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Editorial

Inveterate and Girdling-Congenital Peribronchial Myofibroblastic Tumour

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Congenital peribronchial myofibroblastic tumour emerges as an extremely exceptional pulmonary tumefaction incriminating neonates and infants. Initially documented as a 'hamartoma' in 1949 by Jones, tumefaction was designated as congenital peribronchial myofibroblastic tumour preliminarily in 1993 by Mcginnis., *et al*. Neoplasm encompasses airways and exhibits peribronchial expansion. The predominantly solid tumefaction is composed of cartilage and interlacing fascicles of spindle shaped cells and elliptical cells. Pre-eminently, neoplasm is composed of bland myofibroblasts configuring neoplastic fascicles and bundles while delineating significant mitotic activity. As per current World Health Organization (WHO) classification of thoracic tumours, congenital peribronchial myofibroblastic tumour articulates a solid fibroblastic or myofibroblastic tumour. Previously designated as bronchopulmonary fibrosarcoma, leiomyosarcoma, hamartoma or mesenchymal tumour, congenital peribronchial myofibroblastic tumor develops in utero or is represented within foetal and neonatal period. Frequently asymptomatic, the pulmonary neoplasm of variable magnitude may be appropriately managed with surgical excision wherein subsequent therapeutic intervention remains superfluous. A male predilection is encountered with male to female proportion of ~17:8. Majority of incriminated infants appear as preterm births as the neoplasm is configured within early gestation. Tumour magnitude varies from 3.3 centimetres to 12 centimetres [1,2]. Of obscure aetiology, neoplasm may occur during early gestation period. Tumefaction is posited to arise from pluripotent mesenchymal cells circumscribing proximal branches of various bronchi. Aforesaid pluripotent mesenchymal cells encompassing proximal bronchial branches appear pre-destined to differentiate into cartilage and myofibroblasts which configure the bronchial wall [1,2]. Janus activated kinase 2 (JAK2), a member of JAK protein family is involved within activation of JAK2/STAT3 signalling pathway. Aforesaid pathway is frequently incriminated in oncogenesis of several neoplasms along with angiogenesis and subsequent distant metastasis. Activating chromosomal mutations within Smoothened (SMO) appear tumorigenic and concur with cellular expansion [2,3]. Upon cytogenetic analysis, neoplasm denominates complex genetic rearrangements confined to chromosome 4, 8 and 10. Neoplastic karyotype emerges as 46XX, t (8;10) (p11.2; p15), lns(10;4) (p15; q12iq21) or 46XX, ins (8;4) (p11.2; q12q21), t (8;10) (p11.2; p15) [2,3]. Congenital peribronchial myofibroblastic tumour delineates cogent clinical symptoms associated with respiratory distress, foetal hydrops and intrauterine foetal death. Characteristically, neoplasm is associated with rapid progression and may be misinterpreted as a malignant neoplasm. The essentially benign, rapidly progressive neoplasm may delineate specific clinical symptoms as respiratory distress, polyhydramnios, foetal hydrops or intrauterine foetal death on account of tumour localization or magnitude [4,5].

Grossly, pulmonary parenchyma exhibits a solid, non encapsulated tumefaction of variable magnitude. Tumefaction appears to circumscribe the airways and configures a peribronchial tumefaction adherent to superimposed pleura [4,5]. Upon microscopy, the cellular tumefaction is comprised of immature cartilage and exhibits spindle shaped cells and elliptical cells configuring interlacing fascicles. Tumour cells are permeated with eosinophilic cytoplasm and uniform nuclei with inconspicuous nucleoli. Tumefaction is devoid of cellular and nuclear anaplasia or pleomorphism. Tumour necrosis is absent. Mitotic figures are exceptional and appear as one per 10 high power fields [4,5].

Tumour cells appear intensely immune reactive to vimentin, smooth muscle actin (SMA) or S100 protein. Neoplasm is immune non-reactive to CD34, pan-cytokeratin, epithelial membrane antigen (EMA), anaplastic lymphoma kinase 1 (ALK1), desmin and OCT3/4. Ki67 proliferative index is ~8% [6,7]. Congenital peribronchial myofibroblastic tumour requires segregation from various congenital anomalies or pulmonary tumours as pulmonary sequestration, congenital pulmonary airway malformation, chondromatous hamartoma, teratoma, pleuropulmonary blastoma, foetal lung interstitial tumour (FLIT), inflammatory myofibro-

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].

blastic tumour, congenital infantile fibrosarcoma, monophasic fibrous type or synovial sarcoma [6,7]. Next generation sequencing (NGS) may enunciate JAK2 chromosomal mutation or deletion and chromosomal mutations or deletions within sites ranging from 780 to 879 (c.780-879del) with consequent frameshift mutation confined to exon 7. SMO gene may exemplify point mutation upon acceptor splicing site of exon 5 (c.1140 + 1G > A). Fluorescent in situ hybridization (FISH) may exhibit ETV6-NTRK3 genetic fusion or ALK genetic rearrangement [6,7]. Although diverse pulmonary lesions can be appropriately discerned with plain radiographs, ultrasonography or computerized tomography (CT), cogent imaging features of congenital peribronchial myofibroblastic tumour appear as nonspecific. Pulmonary lesions confined to infants can be appropriately discerned by plain radiography of chest or ultrasonography. As congenital peribronchial myofibroblastic tumour is accompanied by non specific features on imaging, precise histological evaluation of excised surgical tissue samples appears diagnostic [6,7]. Besides, congenital pulmonary lesions comprised of anomalous pulmonary development as pulmonary sequestration, congenital pulmonary airway malformation and primary pulmonary neoplasms as pleuropulmonary blastoma, congenital infantile fibrosarcoma or foetal lung interstitial tumour (FLIT) may be appropriately discerned with prenatal ultrasonography [6,7]. Surgical eradication of the neoplasm is an optimal and recommended mode of therapy. Lobectomy or pneumonectomy of incriminated pulmonary parenchyma is frequently adopted. Additional therapeutic intervention remains superfluous [6,7]. Neoplasm is associated with superior prognostic outcomes. Clinical or radiographic

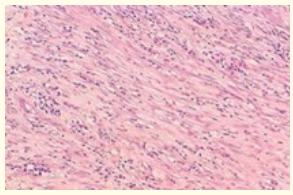

Figure 1: Congenital peribronchial myofibroblastic tumour delineating fascicles of spindle shaped cells and elliptical cells permeated with eosinophilic cytoplasm, uniform nuclei and prominent nucleoli. Few mitotic figures are seen. Cellular atypia or necrosis is absent [8].

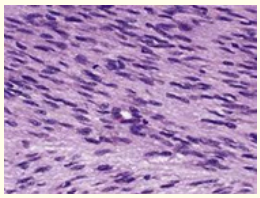

Figure 2: Congenital peribronchial myofibroblastic tumour exhibiting fascicles of spindle shaped cells pervaded with eosinophilic cytoplasm and uniform nuclei. Few mitotic figures are seen. Cellular atypia and necrosis is absent [9].

evidence of tumour reoccurrence or distant metastasis is absent. Tumour associated mortality appears absent. However, extended monitoring is recommended [6,7].

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8. Image 1 Courtesy: Diagnostic pathology, biomed central

9. Image 2 Courtesy: Medical library.com