Mini Review

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Wiry and Weeping-Sclerosing Pneumocytoma Lung

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ABSTRACT

Abbreviations: WHO: World Health Organization; CT: Computerized Tomography; EMA: Epithelial Membrane Antigen; TTF-1: Thyroid Transcription Factor-1

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Sclerosing pneumocytoma appearing within pulmonary parenchyma emerges as a tumefaction composed of dual population of cells. Characteristically, neoplasm is constituted of cuboidal surface cells simulating type II pneumocytes and spherical stromal cells. Alongside, variable foci of haemangioma-like, papillary, sclerotic or solid tumour configurations may be discerned. Initially scripted by Liebow and Hubbell in 1956 as sclerosing haemangioma, tumefaction is associated with an indolent clinical course [1]. Although nomenclated as pulmonary sclerosing pneumocytoma by the World Health Organization (WHO), the terminology is contemplated as obsolete. The characteristic bland cytological features, disparate tumour configurations and dual cell population composed of cuboidal surface cells and spherical stromal cells may be appropriately delineated by cogent immunohistochemistry. Appropriate tumour discernment upon evaluation of miniature tissue samples, cytological smears or frozen section may be challenging. Therefore, examination of permanent formalin fixed, paraffin embedded sections is recommended. Sclerosing pneumocytoma exhibits a female predominance with female to male proportion of ~3.5:1. The mean age of disease emergence is 46 years although no age of disease occurrence is exempt. Neoplasm exhibits a predilection for Eastern Asians and is exceptionally encountered with European population.

Nonsmokers are commonly incriminated [2,3]. Typically, neoplasm manifests as a solitary lesion confined to peripheral pulmonary parenchyma. Lower lobes of lung are commonly implicated. Exceptional, the condition may represent with multifocal, endobronchial tumour nodules or tumefaction confined to pulmonary hilum, visceral pleura or mediastinum [2,3]. Of obscure aetiology, sclerosing pneumocytoma is posited to arise from primitive respiratory tract epithelium. Spherical stromal and surface epithelial cells emerge as clonal cells. Sclerosing pneumocytoma preponderantly delineates pathognomonic mutations within AKT1 gene [2,3]. The typically asymptomatic tumefaction may be incidentally discovered upon radiographic imaging or Computerized Tomography (CT). Nevertheless, symptoms are contingent to tumour localization and magnitude. Nonspecific respiratory symptoms as cough, sputum, haemoptysis or chest pain may ensue. Generally, clinical history of nonsmoker, young subject demonstrating a well circumscribed, peripheral pulmonary lesion is exemplified. Regional lymph node metastasis may concur, a manifestation which is devoid of adverse prognostic outcomes [3,4]. Cytological features of sclerosing pneumocytoma may concur with pulmonary adenocarcinoma. Cytological smears are moderately to markedly cellular with configuration of papillary articulations and discernible, enlarged sheets of tumour cells.

Tumefaction is comprised of miniature to intermediate cells permeated with moderate to abundant cytoplasm, bland spherical to elliptical nuclei and inconspicuous nucleoli. Cellular components are commingled with haemorrhagic foci comprised of red cell extravasation, siderophages and disseminated hemosiderin pigment [3,4]. Upon intra-operative frozen section, macroscopic appearance appears reminiscent of sclerosing pneumocytoma. A distinct dual cell population may be observed. Definitive architectural articulations as papillary, sclerotic, solid or haemorrhagic pattern may be enunciated. Notwithstanding, categorization of lesions demonstrating specific cytological features as predominant solid pattern, hyper cellular smears, occurrence of glandular spaces, desmoplasia-like sclerosis, cellular atypia or coagulative necrosis may be challenging [3,4]. Upon intraoperative assessment, distinction from focal malignant metamorphosis or histological assessment upon frozen section may be challenging. Minimally invasive procedures may be employed for categorizing sclerosing pneumocytoma intraoperatively upon frozen section examination [4,5]. Grossly, a well circumscribed tumefaction with variably grey/tan or yellow surface is observed. Besides, tumefaction may be hard and dark red with distinct haemorrhagic foci. Foci of cystic degeneration and calcification may be enunciated. Tumour magnitude is variable and ranges from one centimetre to 8 centimetres.

However, majority of lesions are < 3.5-centimetre diameter [5,6]. Upon microscopy, pathognomonic dual cell component is comprised of surface cuboidal epithelial cells and spherical stromal cells. Superficial cuboidal epithelial cells simulate type II pneumocytes (Figure 1) Spherical stromal cells appear bland, monomorphic, delineate well defined cellular perimeter and are pervaded with fine nuclear chromatin. Sclerosing pneumocytoma exhibits distinct histologic configurations as ~papillary pattern comprised of superficial epithelial cells superimposed upon hyper-cellular, fibro-vascular core constituted of spherical stromal cells. ~sclerotic pattern composed of hyalinised collagen, hemosiderin pigment deposited within macrophages, cholesterol clefts and foci of dystrophic calcification. ~solid pattern constituted of sheets of spherical cells articulating tubular or incomplete, adenoid configurations encompassed by surface epithelial cells. ~haemorrhagic pattern permeated with blood filled spaces layered by cuboidal epithelial cells. Majority of neoplasms manifest minimally three of aforementioned tumour configurations in varying proportions [5,6]. Sclerosing pneumocytoma appears immune reactive to Epithelial Membrane Antigen (EMA) and Thyroid Transcription Factor-1(TTF-1), pancytokeratin (AE1/AE3), CAM5.2, CK7 or Napsin A. Neoplasm is immune non-reactive to neuroendocrine markers as chromogranin A and synaptophysin [5,6].

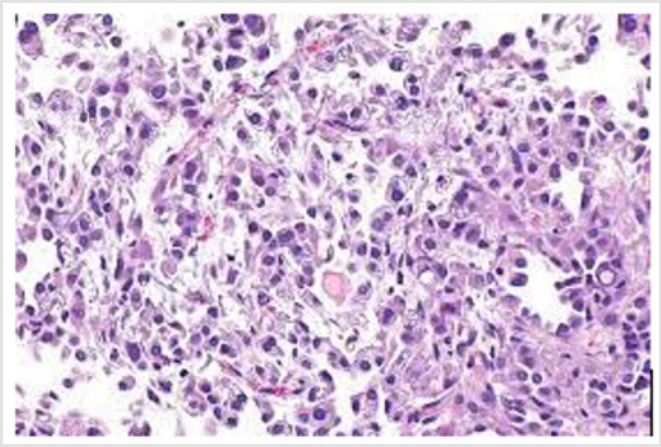

Figure 1: Sclerosing pneumocytoma demonstrating papillary configurations composed of surface cuboidal cells and bland, monomorphic, spherical stromal cells imbued with fibro-vascular cores [8].

Sclerosing pneumocytoma requires segregation from neoplasms such as papillary adenocarcinoma and carcinoid or neuroendocrine tumour of lung [5,6]. Sclerosing pneumocytoma can be appropriately discerned by assessment of cogent clinical features, histological examination, features discerned upon plain radiography or Computerized Tomography (CT) and concordant adoption of ancillary immunohistochemistry (Figure 2) Upon preoperative determination, tumefaction may appear as a solitary pulmonary nodule. Nevertheless, appropriate histological categorization upon miniature tissue samples, cytological smears or cell blocks may be challenging [6,7]. Upon imaging, neoplasm manifests with variable magnitude and ranges from one centimetre to 8-centimetre diameter. However, lesions are preponderantly < 3.5 centimetres. Plain radiographs demonstrate a solitary, well defined, nodular tumefaction with an elliptical or spherical shadow. Foci of calcification may ensue. Besides, a crescent shaped radiolucent zone confined to the periphery is observed, thereby configuring the pathognomonic 'air crescent' sign [6,7]. Computerized Tomography (CT) enunciates a well-defined, intra-parenchymal tu-

mour nodule. Frequently, tumefaction is juxta pleural. Besides, neoplasm may demonstrate focal calcification, in-homogenous image enhancement and an 'air crescent' sign. Occasionally, tumefaction may represent as a lobulated mass delineating 'pure' or 'mixed', ground glass nodule (Table 1).

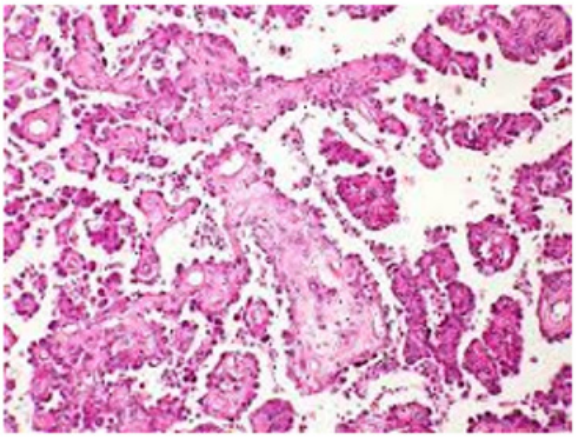

Figure 2: Sclerosing pneumocytoma delineating papillary articulations constituted of surface cuboidal cells and bland, monotonous, spherical stromal cells incorporated with fibro-vascular cores [9].

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

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%)

Additionally, tumour nodules may be associated with pleural traction, thereby simulating a malignant pulmonary neoplasm [6,7]. Surgical resection of the neoplasm is an optimal mode of therapy. Generally, manoeuvers such as sub-lobar wedge resection and regional lymph node dissection for neoplasms demonstrating lymph node metastasis appear adequate for alleviating miniature, peripheral tumours [6,7]. Limited surgical resection of the neoplasm is recommended along with preservation of lung function, especially within neoplasms amenable to surgical extermination with an optimal resection margin [6,7]. Sclerosing pneumocytoma can be appropriately detected in pulmonary nodules exceeding > 1.0-centimetre magnitude. Majority of lesions appear benign and are associated with excellent prognostic outcomes. However, malignant metamorphosis or tumour reoccurrence may ensue. Besides, regional lymph node metastasis, pleural metastasis or distant metastasis are documented. Exceptionally, tumour associated mortality may emerge [6-9].

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8. Image 1 Courtesy: Wikipedia.
9. Image 2 Courtesy: Flickr.com.

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