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A rare tumor of the lung: Pulmonary sclerosing hemangioma (pneumocytoma)

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Summary
A 67-year-old woman was referred to our department for further evaluation of her abnormal, chest radiogram. Thorax computed tomography revealed a well-circumscribed, round mass in the middle lobe of the right lung. A thoracotomy was performed and pulmonary sclerosing hemangioma was diagnosed. We herein present a rare tumor of the lung.

Introduction

Sclerosing hemangioma (pneumocytoma) is a rare benign tumor of the lung with uncertain histogenesis that is composed of two major cell types: surface and round cells. Despite the implication by its name of a vascular neoplasm, sclerosing hemangioma is considered by most authorities to be an epithelial tumor, possibly related to the pulmonary epithelium (type II pneumocyte). Because of this, some investigators call this tumor "pneumocytoma".

It is a well-circumscribed lung parenchymal lesion. This tumor is usually seen in the fifth decaded females. Most of the patients are asymptomatic. They have usually an incidental lung mass on chest radiograms. Computerized tomography is characterized with well defined juxtapleural mass. Calcification may be seen often (41% of the tumor have calcification on microscopic examination). We herein report a case of a rare lung tumor called sclerosing hemangioma or "pneumocytoma".

Case

A 67-year-old woman who presented with a one-month history of non productive cough and upper back pain was referred to local the Department of Chest Diseases of State Hospital. The chest X-ray on admission showed a round

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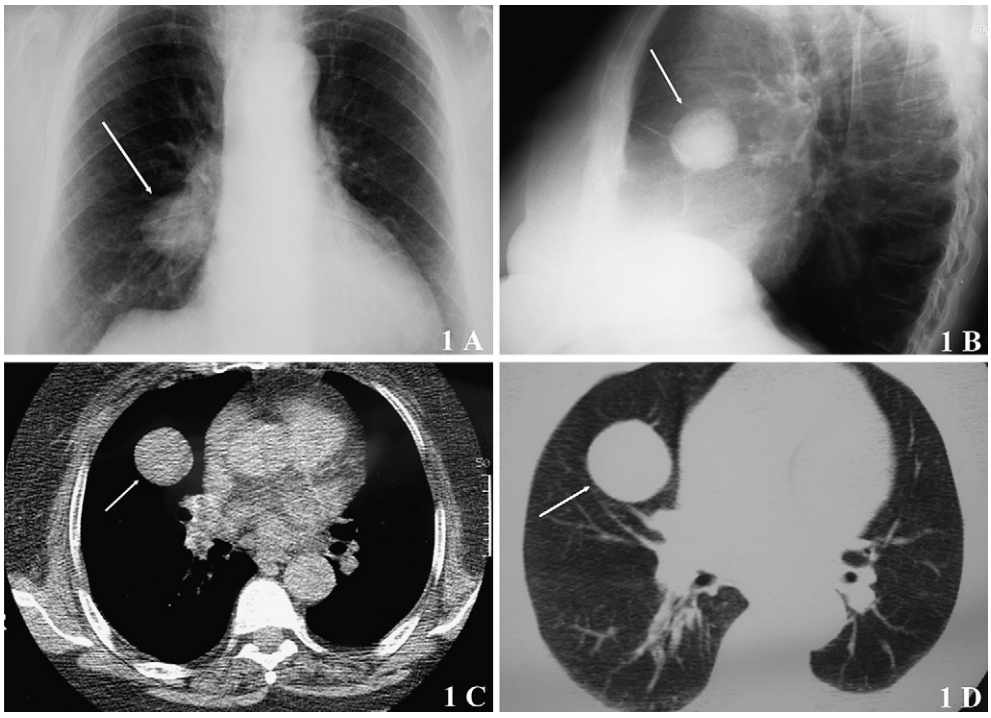

Figure 1 A, B: The mass lesion localized infrahilar and paracardiac area and eventration of the right diaphragma in the chest X-ray. C, D: A round lesion located in the middle lobe of the right lung in thorax CT.

mass lesion on the right paracardiac area. For further investigation, she was referred to Ege University Medical School, Chest Diseases Department. She had a history of 20 pack/years and worked as afarmer for many years. She has been taking antihypertensive drugs for 10 years. Physical examination was normal on admission. Laboratory examinations including total blood count, bleeding time, prothrombin time (PT), and activated partial thromboplastin time (APTT), urine analysis, arterial blood gas analysis were within normal ranges except the erthrocyte sedimentation rate (60 mm/h). In biochemical analysis, the blood urea and creatinin were moderately high. The chest

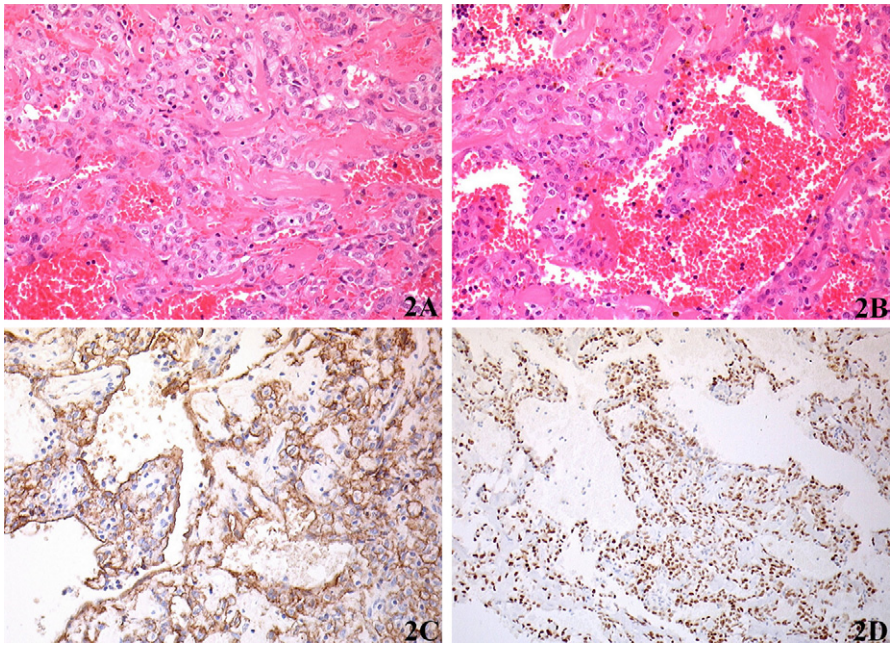

Figure 2 A, B: Strands of sclerotic stroma separating blood-filled spaces lined by flattened cells; pathcy sheets of small polygonal cells. C, D: Both the lining and the polygonal cells were positive for TTF-1 and EMA, weakly positive for AE1/AE3.

X-ray on admission showed a round, well circumscribed mass lesion localized infrahilar and paracardiac area and eventration of the right diaphragma (Fig. 1A, B). A thorax computed tomography (CT) revealed a round lesion located in the middle lobe of the right lung. The tumor was 3.5 cm in diameter (Fig. 1C, D).

Abdominal ultrasonography was normal. The fiberoptic bronchoscopic examination was normal with no malignant cell histology. She underwent a right posterolateral thoracotomy and enucleation of the tumor was performed. She was discharged without any complications on the seventh postoperative day. The pathological examination of the specimen revealed a lung architecture which was replaced by strands of sclerotic stroma separating blood-filled spaces lined by flattened cells. There were pathcy sheets of small polygonal cells (Fig. 2A, B). Both the lining and the polygonal cells were positive for TTF-1 and EMA, weakly positive for AE1/AE3 (Fig. 2C, D). In the immunohistochemical examination, CD1a and S-100 staining were negative and there was no hemosiderin deposition. A Congo Red stain for amyloid was negative. The pathologic appearances were consisted with sclerosing hemangioma.

Discussion

Sclerosing hemangioma is a relatively rare benign tumor of the lung. It is first described by Liebow and Hubell[1] and termed sclerosing hemangioma of the lung owing to prominent sclerotization and vascularization of the tissue. Pathologically, SH is typically composed of solid, papillary, sclerotic or hemangiomatous components.[2,4] The proportions of these four components in the tumor typically vary, although one of them tends to predominate.[4,7] Ultrastructural and immunohistochemical studies have shown that pulmonary sclerosing hemangioma is likely to be the manifestation of the primary proliferation of epithelial cells, probably type II pneumocytes, which may show a neuroendocrine differentiation.[4] The epithelial origin of the tumor was also supported by TTF-1 and EMA positivity in both the surface cells and round cells in the majority of the cases.[8] Pancytokeratin (AE1/3) usually reacts with surface cells, and not with round cells.[8] Tumor is well circumscribed but has not a definite capsule. It grows expansively and compresses the neighboring parenchyma. The tumor is clinically asymptomatic and occurs in young as well as in old people. The peak age incidence is the 5th decade. Women are affected by this tumor more frequently (F:M = 5:1). Based on the findings of higher incidence in females, both estrogen receptors (ER) and progesterone receptors (PR) were studied immunohistochemically. Most patients were positive for both ER and PR, suggesting a relationship between this tumor and female sex hormones.[8] SH is often detected incidentally as a round, well-defined homogenous mass on routine chest radiograph.[9] The greatest diameter of the tumor on chest radiograph has ranged from 13 to 82 mm. Sometimes, in thorax computerized tomography, air-trapping zone surrounding SH or air meniscus sign, which is most commonly seen in aspergilloma, is observed.[6] The tumor is rarely bilateral. Calcification has been reported in 41% of sclerosing hemangiomas.[4] Lymph node metastasis is extremely rare.[5] Tanaka and colleagues in 1986 reported the first case of SH with lymph node metastasis.[8] Contralateral lung metastasis is also rarely seen.[7] Thorax CT and bronchoscopic examinations could not make accurate diagnosis preoperatively. A thoracotomy is usually indicated for definite diagnosis and treatment.[10] The diagnosis is based on histo-pathohistologic examination of the biopsy material. Intraoperative frozen section histology may prevent unnecessary resection of lung tissue. Extracapsular enucleation is the treatment of choice for SH.[11] After surgery, the prognosis of patients is excellent.[3] Lymph node dissection may be necessary to detect lymph node metastasis in cases with a large SH or with enlarged lymph nodes that may be due to metastasis.[7]

Authors contribution

Aysegul Baysak: Design and writing of the article.

Adnan Tolga Oz: Last control before submission and discussion.

Nesrin Moğulkoç: Scientific evaluation.

William Bishop: Pathological evaluation and discussion.

Kenan Can Ceylan: Surgical procedure and discussion.

Conflict of interest statement

None.

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