A Case of Sclerosing Pneumocytoma: A Rare Lung Tumor

Nadir Bir Akciğer Tümörü olan Sklerozan Pnömositoma Olgusu

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Abstract

Sclerosing pneumocytoma is a rare, benign neoplasm which may be in solid, papillary, sclerosing or hemorrhagic patterns histologically, and its papillary surfaces are covered with hyperplastic type 2 pneumocytes. It is often seen in middle-aged adults and women with no recurrence and no reported disease-related deaths. Herein, we present a 50-year-old female case of sclerosing pneumocytoma case who was diagnosed incidentally during acute bronchitis episode.

Key words: Sclerosing pneumocytoma, TTF-1, EMA.

This tumor previously defined as a vascular tumor and named as a sclerosing hemangiomata due to its significant angiomatoid properties, which derived from the primitive respiratory epithelium in the ultrastructural and immunohistochemical studies (1,2). It is common in the mid-aged adults (range, 11 to 80 years, median=46) (3,4). About 80 % of the cases are women (5). It frequently shows solitary or peripheral localization and is more than one in 4% of the cases (6). It is mostly asymptomatic (80%). However, cough, chest pain, or hemoptysis may present (2).

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CASE

A 50-year-old woman was admitted to our emergency unit with fever, cough, and expectoration complaints and was incidentally diagnosed with a tumor during acute bronchitis episode. Her medical history revealed no smoking habit. A detailed examination was performed with a preliminary diagnosis of acute bronchitis. A lesion was detected in her direct pulmonary radiography and pulmonary computed tomography (CT) was obtained following treatment for an infection. The CT images revealed an 11x9.5 mm nodular lesion of the soft tissue density in the fissure which was adjacent to the inferior lobe anterobasal segment of the right lung (Figure 1A). The mass was removed through wedge resection. The macroscopic evaluation showed 1 cm round lesion, which had a harder structure, no capsule, contained brown-black hemorrhagic foci, and separated from the surrounding pulmonary tissues through regular borders (Figure 1B).

The microscopic evaluation of the tumor with hematoxylin and eosin staining included cystic and solid stromal structures. The cystic fields were covered with flat epithelial cells including erythrocytes. Papillary projections were present in some of the cysts. Stromal cells were round-shaped, had spindle-shaped elongated nuclei, and wide eosinophilic cytoplasm with no atypia. Stroma had a rich vascular structure and included hyalinized sclerotic areas and scattered chronic inflammatory cells (Figure 2A-C).
According to the immunohistochemical staining, flat epithelial cells lining the cyst and stromal cells were TTF-1 (+), vimentin (+), EMA (+), chromogranin A (-), S-100 (-), desmin (-), actin (-) and CD34(-). CD34 was (+) in only vascular structures. Flat epithelial cells lining the cyst were pancytokeratin (+), where stromal cells were (-). Ki-67 proliferation index was evaluated to be 1-2% (Figure 3A-5B).

Based on these findings, the patient was diagnosed with sclerosing pneumocytoma. The patient was discharged without any complication two weeks later.

Figure 3a, b and c: Immunohistochemical staining showing that the both stromal and cyst lining epithelial cells are positive for TTF-1 (A), EMA (B), and vimentin (C) [original magnification 100×]

Figure 4a, b and c: Negative for Chromogranin-A (A), SMA (B) and CD34 (C) [original magnification 100×]

Figure 5a and b: Pancytokeratin positivity in only the cyst lining epithelial cells (A). Intratumoral inflammatory cells, but not tumor cells, are positive for Ki67 (B) [original magnification 100×]
DISCUSSION

Sclerosing pneumocytoma is a benign pulmonary tumor observed rarely in Western countries and more frequently in the Eastern Asia (7). Radiologically, the tumor is well-bounded, frequently cystic, rarely calcified in plain radiographs, significantly contrasted and well-bounded in CT (8). Its hemorrhagic component helps to discriminate it from other lesions in magnetic resonance imaging (MRI) (9).

They are macroscopically 0.3-8 cm in-size, well-bounded solid masses with no capsule and lobar distribution, and they contain grey-yellow hemorrhagic foci. Cystic or calcific changes can be also observed (10). Microscopically, they are formed by two different types of cells which are round stromal cells and surface cells (11). Round stromal cells have central localized nuclei without significant nucleoli. Their mitotic index is low (1/10 BBA). Surface epithelial cells has the morphologic properties of type 2 pneumocytes, they are clear, vacuolated, multinucleated, cuboidal flat epithelial cells. In the majority of the patients, different combinations of the papillary, solid, sclerotic, and hemorrhagic patterns are observed. In general, one of these patterns is dominant. Hemorrhagic pattern includes wide spaces filled with blood, covered by flat epithelial cells and a stromal component. Papillary pattern includes round cells in the papillary stalk and complex papilla lined through a cuboidal surface epithelium demonstrating sclerosing and sometimes myxoid changes.

In the sclerotic pattern, co-existence of dense hyalinization and papillary and solid structures is present around the hemorrhagic areas. Solid pattern includes cuboidal surface epithelium which formed small tubules between the round stromal cell clusters (11,12). In our case, hemorrhagic and solid pattern areas were significant. There were also hyalinized sclerotic areas, although less significant.

Immunohistochemically, round stromal cells were TTF-1 and EMA (+), and pancytokeratin (-). The surface epithelium was TTF-1, EMA, and pancytokeratin (+), and CEA, S100, smooth muscle actin, chromogranin-a, and CD34 negative (2,7,13).

Vascular tumors, carcinoid tumor, and pulmonary hamartoma were considered in the discriminative diagnosis. Pulmonary hamartoma are the mixture of different tissues such as cartilage, muscular and fat tissues, and myxoid stroma. A glomus tumor was initially suspected, since the flat cells lining the cystic spaces filled with blood resembled endothelial cells and stromal round cells were present in our case. However, CD34 and SMA negativity and TTF-1 and EMA positivity in both two components are in the favor of sclerosing pneumocytoma. Chromogranin A negativity excluded the possibility of a carcinoid tumor.

In the prognosis, it is a benign tumor with no reported disease-related death and shows no recurrence. Lymph node involvement has been reported in 1% of the cases. Surgical excision is usually sufficient for the treatment. Lymph node dissection may be necessary in case of metastasis suspicion (14).

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS


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REFERENCES


5. Devouassoux-Shisheboran M, Hayashi T, Linnoila RI, Koss MN, Travis WD. A clinicopathologic study of 100 cases of pulmonary sclerosing hemangioma with immunohistochemical studies: TTF-1 is expressed in both round and surface cells, suggesting an origin from primi-
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