Respiratory Medicine Case Reports 17 (2016) 20–23

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case report

Sclerosing hemangioma of the lung showing strong FDG avidity on PET scan: Case report and review of the current literature

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ARTICLE INFO

Article history:
Received 12 December 2015
Accepted 17 December 2015

Keywords:
Sclerosing hemangioma
PET avidity
Lung

ABSTRACT

Sclerosing Hemangioma is a rare lung tumor with polymorphic histologic features that usually occurs in middle aged women. Based on many immunohistochemical and ultrastructural studies, it is most probably derived from undifferentiated respiratory epithelial cells. Symptoms are usually due to enlargement of the tumor and compression of the surrounding tissues. Occurrence of multiple lesions or metastasis is extremely rare although some authors consider sclerosing hemangioma as a potentially low grade malignancy tumor. It usually presents with low to moderate uptake on FDG PET imaging. We present a case of sclerosing hemangioma with strong FDG avidity on PET scan in a 41 year old lady with history of haemoptysis. A full review of the literature on this topic was performed.

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1. Introduction

Sclerosing hemangioma (SH) of the lung represents a rare neoplasm that usually occurs in middle-aged women with only few case reports in young adults and children. SH has four major histological patterns, which vary in their proportions: solid, papillary, sclerotic and haemangiomatous [1]. Occurrence of multiple lesions or metastasis is extremely rare [2,3]. Various immunohistochemical and ultrastructural studies, suggest that SH is most probably arising from a type II pneumocyte [4]. Because of this, it is also called “pneumocytoma”.

2. Case history

A 41 year old lady working as a designer was presented to our outpatient Clinic complaining of cough for several months associated with episodes of mild haemoptysis. There was no significant previous medical history. Examination of the chest revealed decreased breath sound on the right upper lobe. Chest x-ray was performed and revealed a homogenous mass on the right upper lobe with no evidence of effusion or pneumothorax. Blood tests and tumor markers including carcinoembryonic antigen, CA 199 CA 125 and SCC antigen were all normal. A whole body FDG-CT-PET scan showed in the medial right upper zone, a 3.3 cm well circumscribed mass with almost homogenous increased focal uptake (SUV max 8.9). Mild focal activity at a small adjacent right lower paratracheal node at the tracheobronchial angle, as well as in the right hilum (SUV max up to 2.0). No further suspicious for focal avid pulmonary or pleural abnormality. Lung function tests showed a FEV_1 of 2.58 (88% of predicted) with a KCO of 1,40 (82% of predicted). Intraoperative, the lesion was found in the apical segment of the right upper lobe. Decision was made to perform an apico-posterior segmentectomy followed by systematic mediastinal lymphadenectomy. The procedure was uncomplicated. Postoperative course was unremarkable and the patient was discharged home on the 2nd postoperative day. Gross pathological examination revealed a well circumscribed non encapsulated intrapulmonary tumor measuring 35 × 28 × 22 mm, compressing but not infiltrating the adjacent lung parenchyma and not abutting the pleura. On microscopic examination the lesion showed a variegated appearance with papillaroid, sclerotic, solid and angiomatoid areas (Fig. 1a–e). The cells lining the papillaroid and angiomatoid areas have a cuboidal appearance with moderate amount of pale eosinophilic cytoplasm and round nuclei with inconspicuous nucleoli. These

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http://dx.doi.org/10.1016/j.rmcr.2015.12.005
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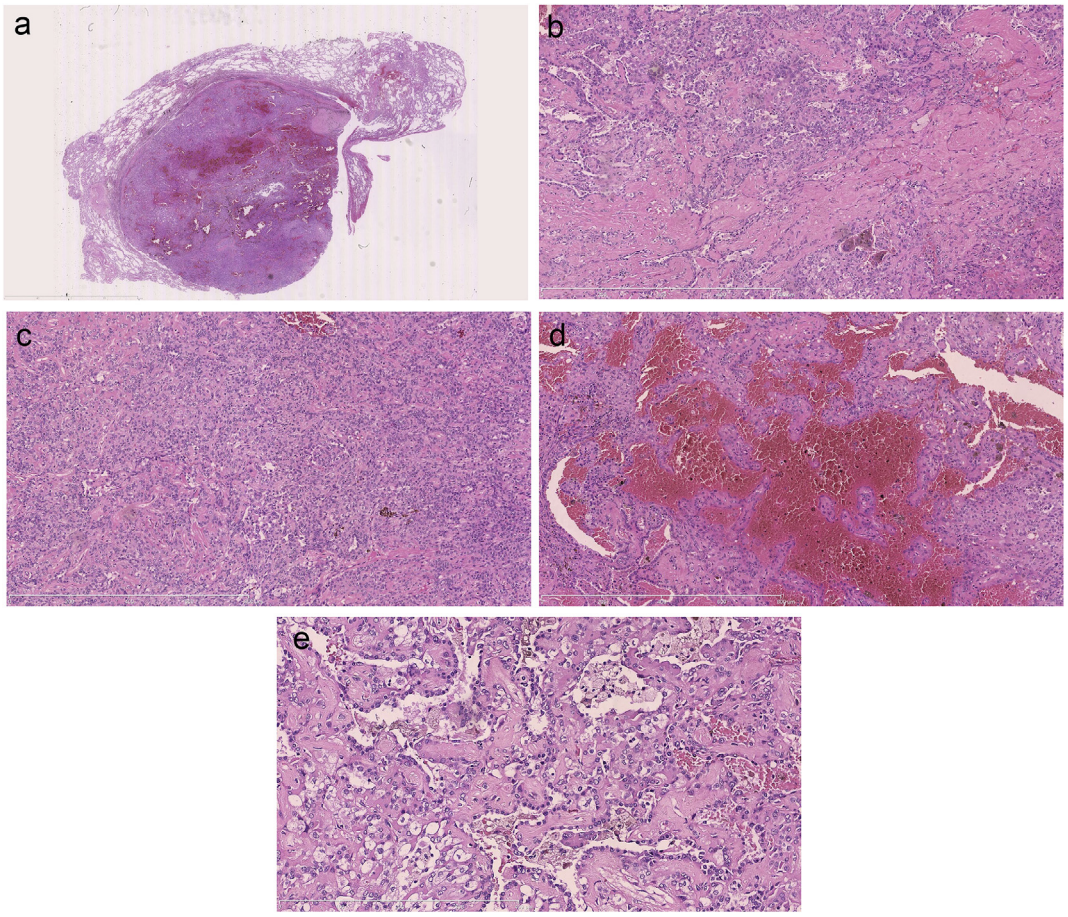

Fig. 1. Sclerosing Pneumocytoma (H&E). Well circumscribed non encapsulated intrapulmonary tumour compressing the adjacent lung parenchyma, showing papillaroid, sclerotic, solid and angiomatoid areas. There are no areas of necrosis. (a) Sclerosing pneomocytoma (H&E X12.5 Magnification) Well circumscribed unencapsulated haemorrhagic intrasclerosing mass. (b) Sclerosing pneumocytoma (H&E X100 Magnification) Sclerotic and papillaroid areas. (c) Sclerosing pneumocytoma (H&E X100 Magnification) Hypercellular areas. (d) Sclerosing pneumocytoma (H&E X100 Magnification) Angiomatoid and haemorrhagic areas. (e) Sclerosing pneumocytoma (H&E X200 Magnification) Papillaroid areas.

cells are noted to express AE1/AE3, EMA and TTF-1 (Fig. 2a–c). A second cell population is observed in the intervening stroma. The latter have a round uniform cellular morphology with clear cytoplasm, distinct cell border and centrally placed small nuclei with fine chromatin and inconspicuous nucleoli. These cells express EMA and TTF-1 but not AE1/AE3. None of the cells express CD31 or CD34 (Fig. 2d). Both cell population are negative for HHV8, CD56, CD10, Synaptophysin, Melan A, S100, HMB45 and SMA. No mitotic figures are observed and the mib-1 proliferation index is less than 1% (Fig. 2e). Several foci of haemosiderosis are observed. There are no foci of necrosis. Lymph nodes showed reactive lymphoid hyperplasia with no evidence of malignancy.

3. Discussion

Sclerosing haemangioma (SH) is a rare benign primary pulmonary tumor. The term sclerosing haemangioma was firstly introduced by Liebow and Hubbell in 1956 [5] owing to prominent sclerotization and vascularization of the tissue. Clinically SH presents most frequently in females (male to female ratio is 1:8) with higher incidence in the far East [4]. It is presenting as an incidental, asymptomatic, solitary peripheral coin lesion detected by chest radiograph [6]. Common symptoms of SH include haemoptysis, chronic cough, chest pain, expectoration and fever; symptoms are usually due to enlargement of the tumor and compression of the surrounding tissue [7]. Tumors range in size from 0.3 to 8 cm and are most commonly located at the periphery of lung parenchyma; rarely, the presentation may be bilateral or as an intramediastinal or endobronchial polypoid mass [8–12]. The FDG-PET scan has been used to differentiate malignant from benign pulmonary lesion: SH usually shows low to moderate FDG uptake, which corresponds to its slow growing behaviour. However the degree of FDG uptake may vary. Some SH have moderate FDG uptake which may exceed the cut off value to determine malignant tumors [13]. Usually larger SH (more than 3 cm) may have higher FDG uptake, whereas smaller SH (less than 3 cm) show lower FDG uptake. The mechanism of this enhanced uptake of FDG is unknown: benign and slow growing tumors usually showed low glucose metabolism. However some larger SH have enhanced FDG uptake, even though mitotic figures are rarely seen. Reason for this increased uptake may be the more active cell proliferation in these larger SH; or these larger SH may have more cell components which are responsible for higher FDG uptake. Therefore, larger SH can be misreading as a malignant neoplasm. The tumor is composed of 2 distinct cellular components, the round or polygonal or stroma cells and the cuboidal or surface cells. Variable cystic spaces, filled with blood cells, simulating large vascular spaces are seen. Area of sclerosis may predominate a tumor. Mitotic activity is typically rare [14]. Differential diagnosis of SH include hamartoma, cavernous haemangioma, inflammatory lesions, arteriovenous malformations,

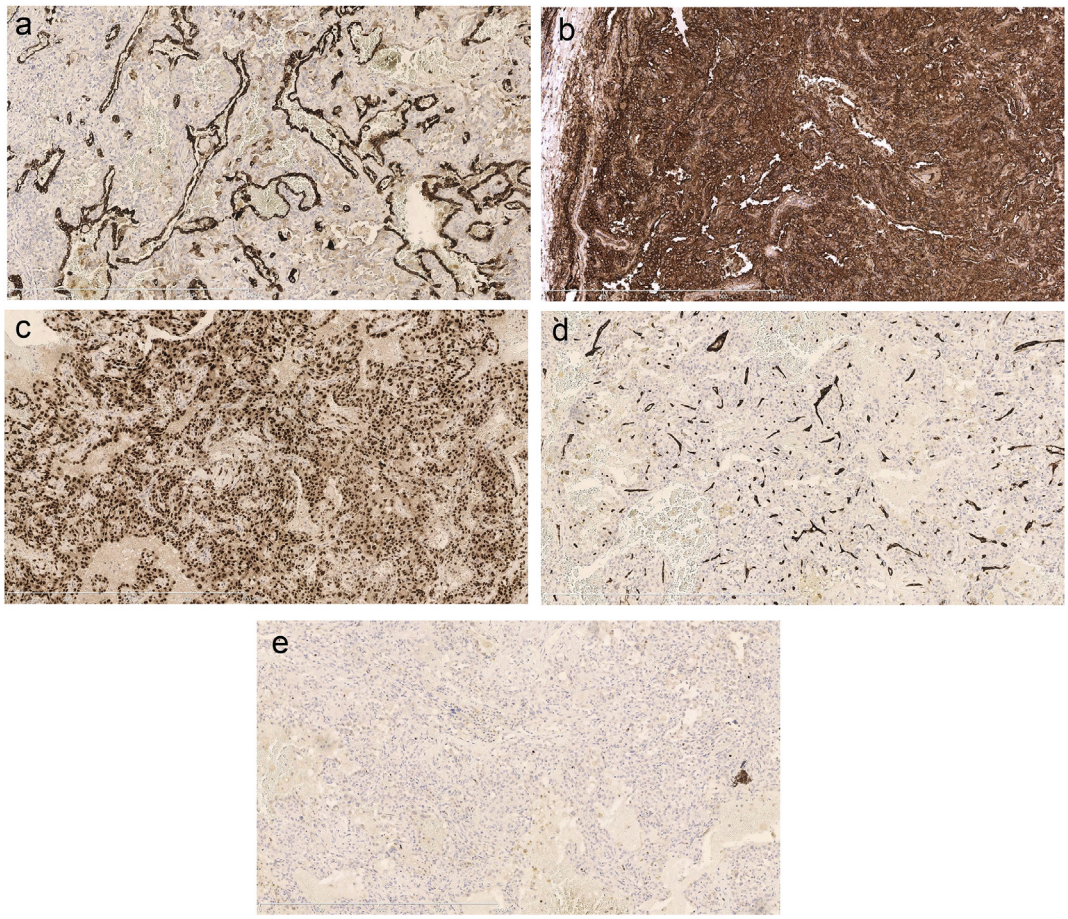

Fig. 2. Sclerosing Pneumocytoma (Immunohistochemistry). AE1/AE3 highlights the presence of a rim of cells lining the angiomatoid and papillaroid areas and a second population of cells found in the intervening spaces which are AE1/AE3 negative. Both cellular components express EMA and TTF-1. (a) Sclerosing Pneumocytoma AE1/AE X100 Magnification. (b) Sclerosing Pneumocytoma EMA X100 Magnification. (c) Sclerosing Pneumocytoma TTF-1×100 magnification. (d) Sclerosing Pneumocytoma CD34×100 Magnification. (e) Sclerosing Pneumocytoma Mib-1×100 Magnification.

malignant teratomas or angiosarcomas. The most important pitfall in the cytological differential diagnosis of SH is well differentiated adenocarcinoma. Immunocytochemistry can aid in distinguishing SH from adenocarcinoma because cells are often negative for keratin. The frequent occurrence of SH in young and middle aged women and the imaging appearance of well circumscribe mass, not common for adenocarcinoma, can be helpful; sometimes calcification is observed: this feature more implies benign than malignant entity Another potential differential diagnosis to consider is well differentiated neuroendocrine carcinoma that shows well defined, hypervascular, and somewhat calcified lesions [15]. In order to better understand the nature of this lesion numerous studies have investigated the immunoprofile of SH [16–19]: both round cells and surface cells have been shown to express EMA and TTF-1. They are negative for factor VIII, S100, smooth muscle actin, calretin, cytokeratin 5/6 and neuroendocrine markers, namely, synaptophysin and chromogranin. The histogenesis of SH has been a matter of controversy since it was originally described. Although an epithelial differentiation for this tumor was postulated by several studies [20,21] other investigators proposed endothelial [10], mesothelial [22], mesenchymal [23], and neuroendocrine [24] origins for this tumor. The clinical features are usually benign and the prognosis after surgical excision is usually excellent. However, some authors consider SH as a potentially low grade malignancy since a few cases of lymph node metastastases have been reported [7]; the first case of SH with lymph node involvement was described by Tanaka and coll. in 1986 [25]. In conclusion SH represents a rare benign pulmonary tumor with significant female predilection. Although considered to be “of uncertain histogenesis” for decades, there is abundant evidence supporting its epithelial derivation and it histogenesis from undifferentiated respiratory epithelium. Its current name, the same term described originally in 1956, is obviously a mismomer. Several names have been proposed, namely “papillary pneumocytoma” [21], “benign sclerosing pneumocytoma” [8], and “sclerotic haemorrhagic alveolar cell tumor of the lung” [26]. Morphologically, SH demonstrates a variety of different patterns. Biologically although capable of metastasis, it has an indolent behaviour with no reported mortality in the literature.

4. Consent

Written informed consent was obtained from the patient for publications of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this Journal.

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

References

[1] K. Sugio, H. Yohohama, S. Kaneko, T. Ishida, K. Sugimachi, Sclerosing hemangioma of the lung: radiographic and pathological study, Ann. Thorac. Surg. 53 (1992) 295–300.

[2] A. Miyagawa-Hayashino, H.D. Tazelaar, D. Langel, T. Colby, Pulmonary sclerosing hemangioma with lymph node metastases, Arch. Pathol. Lab. Med. 127 (2003) 321–325.

[3] J. Hanaoka, M. Ohuchi, S. Inoue, S. Sawai, N. Tezuka, S. Fujino, Bilateral multiple sclerosing hemangioma, Jpn. J. Thorac. Cardiovasc Surg. 53 (2005) 157–161.

[4] B. Chen, J. Gao, H. Chen, Y. Cao, X. He, W. Zhang, M. Luo, S. Zhang, W. Li, Pulmonary sclerosing hemangioma: a unique epithelial neoplasm of the lung (report of 26 cases), World J. Surg. Oncol. 11 (2013) 85.

[5] A.A. Liebow, D.S. Hubbell, Sclerosing hemangioma (histiocytoma, xanthoma) of the lung, Cancer 9 (1956) 53–75.

[6] A.L. Katzenstein, J.T. Gmelich, C.B. Carrington, Sclerosing hemangioma of the lung: a clinicopathologic study of 51 cases, Am. J. Surg. Pathol. 4 (4) (1980) 343–356.

[7] W. Jungraithmayr, S. Eggeling, C. Ludwig, G. Kayser, B. Passlick, Sclerosing Hemangioma of the Lung: a benign tumour with potential for malignancy? Ann. Thorac. Cardiovasc. Surg. 12 (5) (2006) 352–354.

[8] K.W. Chan, A.R. Gibbs, W.S. Lo, et al., Benign Sclerosing Pneumocytoma of lung (sclerosing hemangioma), Thorax 37 (1982) 404–412.

[9] M. Devoussaoux, T. Hayashi, R. Linnoila, M. Koss, et al., 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 primitive respiratory epithelium, Am. J. Surg. Pathol. 24 (7) (2000) 906–916.

[10] A.S. Leong, K.W. Chan, H.S. Seneviratne, A morphological and immunohistochemical study of 25 cases of so-called sclerosing haemangioma of the lung, Histopathology 27 (1995) 121–128.

[11] N. Nagata, M. Dairaku, K. Sueishi, K. Tanaka, Sclerosing hemangioma of the lung. An epithelial tumor composed of immunohistochemically heterogenous cells, Am. J. Clin. Pathol. 88 (1987) 552–559.

[12] J. Rodriguez-Soto, T.V. Colby, R.V. Rouse, A critical examination of the immunophenotype of pulmonary sclerosing haemangioma, Am. J. Surg. Pathol. 24 (2004) 906–916.

[13] J. Neuman, A. Rosioreanu, A. Schuss, et al., Radiology-pathology conference: sclerosing hemangioma of the lung, Clin. Imaging 30 (2006) 409–412.

[14] Gy Kim, J. Kun, Y.S. Choi, et al., Sixteen cases of sclerosing haemangioma of the lung including unusual presentations, J. Korean Med. Sci. 19 (2004) 352–358.

[15] M.L. Rosado de Christenson, G.F. Abbott, W.M. Kirejczyk, et al., Thoracic carcinoids: radiologic-pathologic correlation, Radiographics 19 (1999) 707–736.

[16] A. Yoda, K. Hiroshima, M. Shiba, Y. Haga, Y. Moriya, Y. Sekine, et al., Clinocopathological analysis of pulmonary sclerosing hemangioma, Ann. Thorac. Surg. 78 (6) (2004) 1928–1931.

[17] S.H. Yoo, K.C. Jung, J.H. Kim, S.W. Sung, J.H. Chung, Y.S. Shym, et al., Expression patterns of markers for type II pneumocytes in pulmonary sclerosing hemangiomas and fetal lung tissues, Arch. Pathol. Lab. Med. 129 (7) (2005 Jul) 915–919.

[18] A.G. Nicholson, C. Magkou, D. Snead, H.A. Vohra, M.N. Sheppard, P. Goldstraw, et al., Unusual sclerosing haemangiomas and sclerosing haemangioma-like lesions, and the value of TTF-1 in making the diagnosis, Histopathology 41 (5) (2002 Nov) 404–413.

[19] P.B. Illei, J. Rosai, D.S. Klimstra, Expression of thyroid transcription factor-1 and other markers in sclerosing haemangioma of the lung, Arch. Pathol. Lab. Med. 125 (10) (2001 Oct) 1335–1339.

[20] G.S. Hill, J.C. Eggleston, Electron Microscopic study of so-called “pulmonary sclerosing haemangioma”: a report of a case suggesting epithelial origin, Cancer 30 (1972) 1092–1106.

[21] A. Kennedy, “Sclerosing haemangioma” of the lung: an alternative view of its development, J. Clin. Pathol. 26 (5) (1973) 390.

[22] A.L. Katzenstein, D.L. Weise, K. Fulling, H. Battifora, So-called sclerosing hemangioma of the lung. Evidence for mesothelial origin, Am. J. Surg. Pathol. 7 (1) (1983) 3–14.

[23] M. Huszar, S. Suster, E. Herczeg, B. Geiger, Sclerosing Hemangioma of the lung. Immunohistochemical demonstration of mesenchymal origin using antibodies to tissue specific intermediate filaments, Cancer 58 (11) (1986) 2422–2427.

[24] H.M. Xu, W.H. Li, N. Hou, S.G. Zhang, H.F. Li, S.Q. Wang, et al., Neuroendocrine differentiation in 32 cases of so-called sclerosing hemangioma of the lung: identified by immunohistochemical and ultrastructural study, Am. J. Surg. Path 21 (9) (1997) 1013–1022.

[25] I. Tanaka, M. Inoue, et al., A case of pneumocytoma with lymph node metastasis, Jpn. J. Clin. Oncol. 16 (1) (1986) 77–86.

[26] M. Fukayama, M. Koike, So-called sclerosing haemangioma of the lung. An immunohistochemical, histochemical and ultrastructural study, Acta Pathol. Jpn. 38 (5) (1988) 627–642.