

CASE REPORT

Incidental finding of a sclerosing hemangioma in a Caucasian woman

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Abstract

Sclerosing hemangioma of the lung is a rare and mostly benign lung tumor, which affects especially Asian middle-age women (median age of 48 years). We report the case of a 27-year-old woman in which, a premarital routine chest X-ray investigation revealed a 2 cm well-defined opaque nodule in the lower left pulmonary lobe, confirmed by CT scan. Microscopically, the surgically enucleoresected nodule was represented by a heterogenic tumor (papillary, solid, sclerotic, hemorrhagic patterns), containing two cell populations: cuboidal surface epithelial cells lining the papillary structures and round stromal cells in solid areas, with distinct immunoprofile and low mitotic activity, consistent with sclerosing hemangioma. This case is particular because, being rare in Caucasian persons, intraoperative diagnosis on frozen sections is extremely difficult, and routine histopathological diagnosis needs immunohistochemical tests to set the correct diagnosis, hence the correct therapeutic attitude.

Keywords: sclerosing hemangioma, pneumocytoma, rare tumor, lung, immunohistochemistry, Caucasian woman.

Introduction

Sclerosing hemangioma (SH) of the lung (also known as pneumocytoma, sclerosing pneumocytoma or papillary pneumocytoma) [1] is a rare and mostly benign lung tumor, which was first described by Liebow & Hubbell in 1956 [2].

Initially thought to be of vascular origin, hence its name, nowadays it is widely accepted the origin in primitive (immature) respiratory epithelium [1, 3–5]. It affects the middle-age adults (11–80-year-old, median 48 years), with a predominance in women (female/male ratio of 4:1) [1].

Its incidence is higher in Asian population than in Western population [1, 6]. Usually, it is a solitary peripheral lesion (only in about 4% being multiple [3]), asymptomatic, rarely involving the pleura or the bronchial tract, therefore it is diagnosed incidentally, during a chest CT or routine X-ray investigation. The pathological examination on surgical specimens brings the definite diagnosis. Prognosis is excellent after complete excision.

Here, we report a case of a young Caucasian woman in which a routine chest X-ray revealed a tumor mass in the lower left pulmonary lobe, with histological and immunohistochemical pattern of sclerosing hemangioma.

Case report

The patient is a 27-year-old woman who performed a premarital routine chest X-ray investigation, which revealed an opaque mass in the lower left pulmonary lobe, therefore she was referred to Department of Thoracic Surgery.

She is a non-smoker and with no significant personal pathological history.

Physical examination could not identify any symptomatology or signs of disease.

Laboratory tests were normal, except a mild increase of PDW (platelet distribution width) (17.4; normal: 9.0–15.0), and P-LCR (platelet larger cell ratio) (46.4%; normal: 13.0–43.0); erythrocyte sedimentation rate (ESR) – 20 mm/h; total cholesterol 200.2 mg/dL (normal: 100.0–200.0); creatinin 0.56 mg/dL (normal: 0.60–1.30); ionic calcium 4.14 mg/dL (normal: 4.25–5.25).

No malignancy was diagnosed in sputum cytology.

Chest X-ray examination showed an opaque, round, well delineated mass, measuring 2 cm, in the left lower lobe field. The description of CT examination confirmed the presence of a solid well-defined round macronodule, measuring 1.95/1.69/2.0 cm, non-calcified with intense enhancement after intravenous administration of contrast iodine, antero-basal located (eighth segment) in the lower left lobe, imprinting the segmentary vein which remains permeable, but with no relation with pleura or left fissure (Figure 1, A and B).

After provided informed consent, the patient underwent a surgical enucleoresection with favorable post-operative evolution after drainage tube elimination.

On gross examination, the surgical specimen measured 2/1.6/1.2 cm, with a central nodular, well-circumscribed pink-grey lesion.

The specimen was fixed in neutral buffered formalin, paraffin-embedded and sections were routinely stained with Hematoxylin–Eosin (HE), Alcian blue and van Gieson trichromatic.

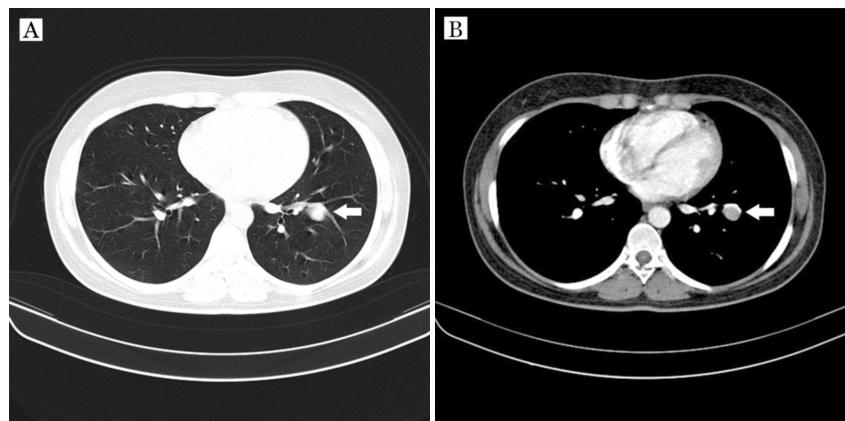


Figure 1 – Chest CT scan. (A) Native examination, pulmonary window: a solid, well-defined round nodule, located in the eighth segment of the left lobe. (B) Arterial examination, mediastinal window: intense enhancement of the nodule which abuts the segmentary vein.

Microscopically, the pulmonary tissue harbored a tumor nodule with a very heterogenic aspect, which included four architectural patterns: papillary, solid, sclerotic and hemorrhagic. The complex papillary structures were covered by cuboidal monomorphic surface epithelial cells, round to oval hyperchromatic or vacuolated nuclei, without nucleoli, and scant eosinophilic cytoplasm. The solid areas and the cores of the papillary projections contain round or polygonal medium-sized stromal cells, with central round vacuolated nucleus, no nucleolus, eosinophilic or clear cytoplasm (Figure 2A).

On Alcian blue staining, no mucin secretion was detected (Figure 2B). Mitosis figures were extremely rare (one mitotic figure/20 HPF – high power field), and typical. The stroma exhibited sclerohyaline component (hyalinized collagen) with abundant vascularization, hemorrhagic areas and rare psammoma-like microcalcifications.

Other microscopic findings included chronic inflammation with lymphocytes, histiocytes, hemosiderin and cholesterol clefts deposits. The tumor grows in wide front, compressing the surrounding pulmonary tissue, without invading it or the adjacent pleura. No necrosis was observed.

Immunohistochemical (IHC) tests showed that epithelial surface cells are immunopositive for multi-cytokeratins (CK) AE1/AE3 (clones AE1 and AE3, Bond™ RTU – ready-to-use, Novocastra, Germany) (Figure 2C), epithelial membrane antigen (EMA) (clone GP1.4, Bond™ RTU, Novocastra, Germany), and thyroid transcription factor 1 (TTF1) (clone SPT24, Bond™ RTU, Novocastra, Germany) (Figure 2D) and negative for progesterone receptor (PR) (clone 16, Bond™ RTU, Novocastra, Germany) (Figure 2E), while the round stromal cells are immunopositive for EMA, TTF1, cytokeratins AE1/AE3 (mild, focally), and PR. Ki-67 (clone MMI, 1:50, Novocastra, Germany) was positive in about 1% of tumor cells (Figure 2F).

For detection, we used NovoLink™ Polymer Detection System (Leica Biosystems, Germany).

Morphological aspects and immunohistochemical features were consistent with sclerosing hemangioma.

The patient was discharged four days after, without any further adjuvant therapy.

Two months later, the CT scan revealed good expansion of the lung.

Discussion

SH is a rare pulmonary tumor. Our SH case perfectly fits the typical profile of a SH. As most of SH, it was a benign, solitary, slow growing tumor, hence the asymptomatic evolution, until the imagistic investigation performed for alternative purposes (premarital medical check-up) identified it. Only a few cases were reported to present lymph node metastasis [3, 7]. Indeed, no lymph node lesions were diagnosed in our patient. It affects mainly Asian middle-aged women, hence the rarity of our case, a Caucasian woman. At diagnosis, the tumor may have 0.3–11 cm in diameter, with an average of 3 cm. In our case, the tumor had only 2 cm in greatest diameter due to the early detection, our patient being only 27-year-old (below average age for this tumor, which is 48 years [1]) at diagnosis. Being well-defined on X-ray and CT examination, the first diagnosis supposition was either an inflammatory lesion (tuberculosis) or a hamartoma.

The intraoperative diagnosis of SH on frozen sections is difficult for inexperienced pathologists considering the artifacts which modifies the architecture. The papillary architecture may falsely lead to a pulmonary papillary adenocarcinoma, hence the risk of false-positive diagnoses of malignancy vary between 25% and 56% [8, 9]. Therefore, in our case, the surgical attitude was the removal only of the nodule. If the pathological examination would have detect a malignant lesion, the local resection would have been followed by surgical removal of the entire lower left lobe and systematic lymph node dissection in a second step, which otherwise is not recommended.

Due to the papillary structures, on paraffin-embedded sections, the differential diagnosis included the papillary adenocarcinoma of the lung and a metastatic nodule from a papillary thyroid carcinoma. We excluded the malignancy taking into account the lack of cytological atypia of the epithelial lining cells on the surface of the papillary structures (although atypia may be rarely present in SH [10]), low mitotic rate, and the heterogeneity of the tumor (all four histological patterns being present in our case). Also, the presence of two types of cells (surface cells and round stromal cells), the sclerohyaline stroma with hemorrhagic areas and microcalcifications were major diagnostic criteria for SH. Immunohistochemistry came to confirm the initial supposition of SH. As also stated in most publications [3–5, 8, 10–14], the immunoprofile of our SH showed positivity in both cell types for

cytokeratins cocktail (AE1/AE3), EMA and TTF1. Of real help was the progesterone receptor positivity only for the polygonal round stromal cells, but not for the surface cells [1, 11, 15]. The correlation between SH and female sex

hormones might explain the predominance of SH in women.

Consistent with the final pathological report, there was no need for further surgical interventions, and the patient was discharged with follow-up recommendations.

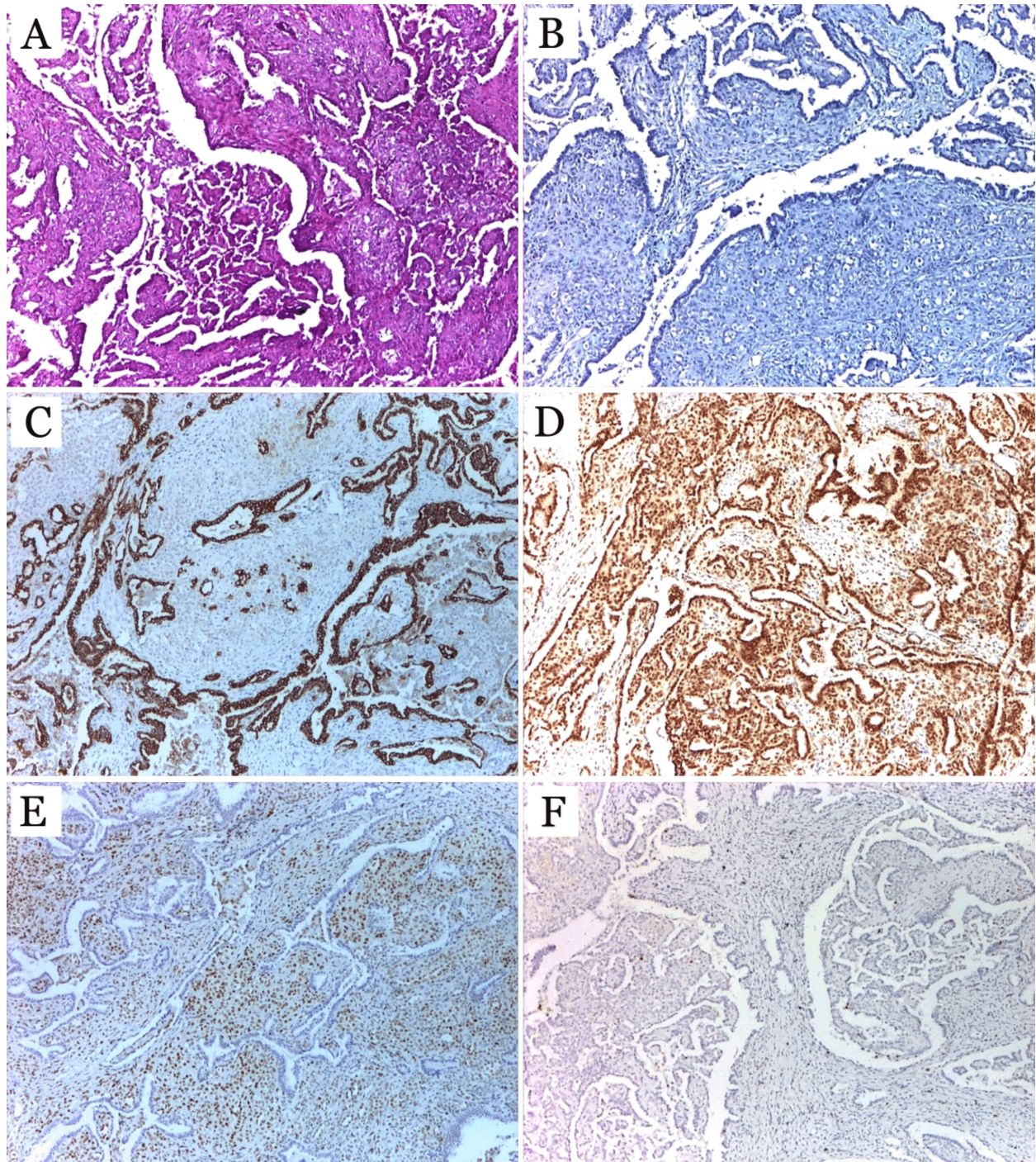


Figure 2 – Sclerosing hemangioma. (A) Papillary and solid areas with two cell populations: surface epithelial cells lining the papillary structures and round stromal cells in solid areas and papillary cores (HE staining, 40×). (B) No mucin secretion was present in tumor cells (Alcian blue staining, 40×). (C) Cytokeratin AE1/AE3 showed a diffuse and strong positive reaction in surface epithelial cells and mild, focally positive in round stromal cells (IHC, Ab – antibodies anti-CK AE1/AE3, DAB – 3,3'-diaminobenzidine, 40×). (D) TTF1 – strong, positive reaction in both epithelial surface cells and round stromal cells (IHC, Ab anti-TTF1, DAB, 40×). (E) Progesterone receptor – negative in surface epithelial cells, strong and diffuse positive reaction in round stromal cells (IHC, Ab anti-PR, DAB, 40×). (F) Low mitotic activity (one mitotic figure/20 HPF); Ki-67 positive in 1% of tumor cells (IHC, Ab anti-Ki-67, DAB, 40×).

Conclusions

We presented this rare case of pulmonary sclerosing hemangioma in a Caucasian woman stressing on its

favorable evolution tributary to the early diagnosis. Considering the differential diagnosis pitfalls, especially with the pulmonary papillary adenocarcinoma, careful histopathological examination and use of immunohisto-

chemistry are mandatory to avoid excessive surgical attitudes.

Conflict of interests

The authors declare that they have no conflict of interests.

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