Pulmonary Sclerosing Pneumocytoma: Case Report and Review of the Literature

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Abstract

Aims: Pulmonary sclerosing pneumocytoma is a rare benign neoplastic tumour. It is often discovered incidentally in chest X-ray or chest CT scan performed for others reasons. We present here a case of pulmonary sclerosing pneumocytoma and discuss the clinical, histopathological features and treatment of this tumour in light of the data of the literature.

Methods and Results: We report the case of pulmonary sclerosing pneumocytoma found in 66-year-old woman on chest CT scan performed for staging of a breast cancer. The patient underwent a left lower lobectomy and ipsilateral mediastinal lymph node dissection because frozen section examination suggested papillary adenocarcinoma.

Conclusion: Most occurrences of pulmonary sclerosing pneumocytoma is often discovered incidentally. It is a rare benign tumour that frozen section examination is unable to diagnosis them in 25 to 56% of cases. The prognosis after surgical resection is excellent.

Keywords: Sclerosing pneumocytoma; Lung; Rare benign tumor; Incidentally

Case Report

A 66-year-old woman was referred to our institution in April 2012 for management of invasive lobular carcinoma of the right breast. Chest CT scan performed on 12.04.2012 for staging of the breast cancer demonstrated an isolated pulmonary nodule measuring about 1 cm in the left lower lobe (Figure 1). The patient reported no respiratory symptoms and was a non-smoker. Positron emission tomography performed on 10.05.2012 revealed the left lower lobe pulmonary nodule, which was very slightly hypermetabolic (SUV max: 1.4). Right total mastectomy and axillary lymph node dissection were performed on 30.05.2012. Histological examination concluded on a 30 mm invasive lobular carcinoma, histoprognostic grade EE II, associated with invasion of 18 out of 37 axillary lymph nodes. The patient was treated by adjuvant radiation therapy and hormonal therapy with Femara® (letrozole). CT-guided chest aspiration of the left lower lobe pulmonary nodule proved to be impossible. After discussion at the multidisciplinary consultation meeting, surgical resection was performed on 3.10.2012. Frozen section examination suggested papillary adenocarcinoma. Left lower lobectomy and ipsilateral mediastinal lymph node dissection were then performed. Macroscopic examination of the cut surface of the resection specimen revealed a circumscribed, yellowish nodule measuring 0.8 x 0.7 cm, with a solid appearance and haemorrhagic zones that did not appear to invade the pleura. On histological examination, the tumour was composed of a double cell contingent with a papillary architecture (Figure 2). One contingent lined the axis of the papillae, composed

Figure 1: Chest CT scan on 12.04.2012.

Figure 2: HES-stained histology slide clearly showing the double cell contingent with a papillary architecture: a surface contingent and a stromal contingent in the axis of the papillae (H and E, x 200). The inset in the bottom right corner shows a higher power view of cytokeratin AE1/AE3 immunohistochemistry demonstrating the existence of two cell populations: negative round stromal cells and positive surface cuboid cells (x 200).

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of cuboid cells with round nuclei and no visible nucleoli. The other
contingent was situated in the axis of the papillae and was composed
of small round cells with more or less clearly visible cytoplasmic
limits, with nuclei comprising fine chromatin and a small nucleolus.
No mitotic figures were observed. Haemorrhagic changes were noted.
Immunohistochemistry showed that stromal cells were EMA (+), TTF1
(+), pancytokeratin AE1/AE (-) and that surface cells were EMA (+),
TTF1 (+), and pancytokeratin AE1/AE (+) (Figure 2). The definitive
histological examination concluded on sclerosing pneumocytoma with
no hilar and/or mediastinal lymph node invasion.

Discussion

We report a case of pulmonary sclerosing pneumocytoma and
discuss the clinical and histopathological features and the treatment of
this very rare lung tumour in the light of the data of the literature.

Pulmonary sclerosing pneumocytoma is a benign tumour that was
described for the first time by Liebow and Hubbell in 1956 [1]. It is
a rare lung tumour, accounting for approximately 1% of all benign lung
tumours. It predominantly affects women in the fifth decade [2-4] and
has a higher incidence in Asia, similar to that of carcinoid tumours
[3,4].

The majority of patients are asymptomatic and the lung lesion is
usually discovered incidentally on chest x-ray or chest CT scan. Only a
few patients have reported clinical symptoms such as cough, breathing
difficulties, chest pain and/or haemoptysis [2-4].

In the great majority of cases, sclerosing pneumocytoma presents as
a solitary, homogeneous, well delineated nodule or peripheral mass [2-4]. The frequent presence of a haemorrhagic component is responsible
for marked contrast enhancement on computed tomography and a high-intensity signal on magnetic resonance imaging. An "air meniscus sign" is
frequently observed around the tumour [4]. Although benign, the
great majority of sclerosing pneumocytoma present increased uptake on
18FDG positron emission tomography with standardized uptake values
(SUV) that are significantly correlated with tumour size [5].

Macroscopic examination of sclerosing pneumocytoma reveals a
well circumscribed solid nodule sometimes associated with the presence
of haemorrhagic changes and more rarely cystic changes or
calcifications. In a series of 100 cases, the mean tumour diameter was
2.6 cm. In 96% of cases, the tumour presented as a solitary nodule, while
multiple nodules were observed in only 4% of cases [2]. On histological
examination, two cellular contingents are always present, central round
stromal cells and peripheral cuboid cells, with a variable architecture,
papillary, sclerotic, solid and haemorrhagic [2], and a low mitotic index
(usually less than 1 per 10 high power fields) [6]. Calcifications, or more
rarely adipose tissue, tumourlets, or exceptionally an associated carcinoid
tumour may be observed [2]. Immunohistochemistry reveals TTF1 (+)
and EMA (+) stromal and peripheral cells, but pancytokeratin negative
in stromal cells, in more than 90% of cases (Figure 2) [2]. Differential
diagnoses include a metastatic renal cell carcinoma, clear cell 'sugar'
tumour, carcinoid and papillary pulmonary epithelial neoplasms.
Sclerosing pneumocytoma can be usually be distinguished from these
by bland cytology, heterogeneous architecture and a characteristic
immunostaining pattern. Kim et al. reported that cytoplasmic and
membrane labelling with anti-Ki67 (MIB-1) antibody and a low Ki-67
proliferation index can be useful to distinguish pulmonary sclerosing
pneumocytoma from non-small cell lung carcinoma [6].

Surgical resection allows histological diagnosis in the majority of
cases. Frozen section examination is unable to diagnose sclerosing
pneumocytoma in 25 to 56% of cases. In our case, frozen section
examination suggested a diagnosis of malignant adenocarcinoma with
a papillary architecture.

The pathogenesis of this tumour has not been fully elucidated.
Multiple origins have been proposed since the first description of
this tumour in 1956: endothelial, mesothelial, mesenchymal, neuroendocrine and bronchial and alveolar pulmonary epithelial
cells. The morphology of surface cells is similar to type II pneumocytes,
while no equivalent of stromal cells is present in the normal lung. The
positive TTF1 and EMA staining of the two contingents combined with
the absence of expression of surfactant A and B proteins confirm the
epithelial origin of the tumour, derived from primary undifferentiated
respiratory epithelium. Molecular studies demonstrate the monoclonal
nature of the two cell types, indicating that pulmonary sclerosing
pneumocytoma constitutes a true tumour and not a hamartoma [2].

Pulmonary sclerosing pneumocytoma is now generally considered
to be benign lesion and surgical resection alone is considered to be
curative. However, some authors consider it as a potentially low-grade
malignant tumour, as several cases of lymph node metastases [7,8] and
postoperative local recurrences [9] have been reported. The need for
lymph node dissection, in view of the possibility of local lymph node
invasion in 2 to 4% of cases, remains controversial [10]. The prognosis
after surgical resection is excellent, even in the presence of lymph node
metastases, multiple lesions or recurrences [3]. Although it is generally
a benign disease, follow-up is necessary.

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