

The so called pulmonary sclerosing hemangioma

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Abstract

Background: The so called pulmonary sclerosing hemangioma (PSH) is categorized as a miscellaneous tumour in the new 1999 World Health Organization. **Aims:** Our objectives are to report 2 cases of PSH with documented radiological and pathological features, to make a review of the literature about the origin, the means of diagnosis, the treatment and the prognosis of this rare and benign tumour. **Materials and methods:** We report the cases of 2 patients aged 60 and 38 years that presented non specific respiratory signs. The radiological findings were challenging in both cases and the diagnosis was based on histological findings. one case was difficult because of the presence of many granulomas dealing with the diagnosis of tuberculosis which is endemic in our country. **Conclusion:** PSH are benign tumours with a good behaviour. Although the progress made in radiological techniques to elucidate these lesions, the diagnosis remains based on histological findings.

INTRODUCTION

The so called pulmonary sclerosing hemangioma (PSH) is a relatively rare benign tumour which was described initially in 1956 by Liebow and Hubell (1). It has been also called benign alveolar cell tumour and hamartoma (2, 3, 4). It is categorized as a miscellaneous tumour in the new 1999 World Health Organization (5). We report 2 new cases of PSH diagnosed based on histological and immunohistochemical findings.

CASE REPORTS

A 60-year-old asymptomatic woman underwent in 1998 a chest roentogram for preoperative evaluation of a breast carcinoma. Chest-X-ray showed a peripheral, homogeneous and well-circumscribed nodule located in the right lung. The CT-scan revealed a lesion of the middle lobe. No lymphadenopathy was observed (Figure 1).

{image:1}

The diagnosis of a metastatic disease was initially suspected. A right middle lobectomy was undergone through a right postero-lateral thoracotomy. Per-operative findings consisted in a well circumscribed 2-centimeter nodule associated with multiple adenomegalies which were excised. Extemporaneous examination of the tumoral nodule didn't allow to rule out a malignant lesion. Histological examination showed a well circumscribed but non

encapsulated nodule measuring 2, 5 x 2 cm. Two patterns were predominantly observed: solid and papillary. Blood-filled spaces were also noted [Figure 2].

{image:2}

The solid pattern was composed of a sheet-like proliferation of round to polygonal cells with a pale cytoplasm. The papillary pattern was lined by cuboidal surface cells. Whereas, polygonal cells appeared in the stroma. Polygonal cells, which lined the stroma, expressed the epithelial membrane antigen (EMA) and the thyroid transcription factor-1 (TTF-1). Cuboidal cells expressed EMA, TTF1 and cytokeratin [Figure 3].

{image:3}

CD34 and Desmin were also used and were negative in both types of cells. According to the histological and immunohistochemical findings, the diagnosis of PSH was retained. The patient didn't present any recurrences after a ten-year follow up period.

The second patient is a 38-year old patient without a particular past medical history who presented with an unproductive cough, anorexia and asthenia. The onset of the symptoms was one month. A chest-x-ray showed 2 peripheral nodules. Pulmonary fibroscopy showed inflammatory lesions. A thoraco and abdominal CT-scan showed 2 peripheral nodules of the left lung. The 2 nodules

showed a marked contrast enhancement. There were no adenomegalies. Multiple hepatic nodules corresponding to biliary cysts were also noted. In order to rule out a malignant disease, a postero-lateral thoracotomy was performed. Two nodules measuring 2 cm and an adenomegaly were discovered and resected. The extemporaneous examination concluded to a benign disease evoking tuberculosis because of the presence of many granulomas. Histological and immunohistochemical examination concluded to a PSH with a granulomatous reaction. Sputum cultures were negative for mycobacterium tuberculosis. Seven years after the establishment of the first diagnosis, the patient didn't presented recurrences.

DISCUSSION

Although initially regarded as a variant of haemangioma, on the basis of the results of immunohistochemical and genetic studies PSH is now considered as an epithelial tumour. It is usually observed in women (80% of the cases) with a mean age of 16 years (range, 11 to 80 years) (8). Eighty percent of the patients are asymptomatic and the lesions are generally discovered incidentally (8). In symptomatic patients, that account for 10 to 20% of the cases, the cough, the dyspnea, the chest pain and hemoptysis are mainly observed. On chest-x-ray, these tumours appear as a well-defined round or oval masses like in our 2 cases (5). According to Im and coworkers, in a report of CT-findings in 8 cases, these tumours present as a well-defined juxta-pleural mass with marked contrast enhancement caused by its hemangiomatous component (9). This finding was observed in our second patient and these morphological features are characteristic enough to rule out a malignant nodule which is generally lobulated with speculated margins. However, these features can overlap with the CT-findings of other benign nodules such as hamartoma. Chung and colleagues stipulate that dynamic studies showing characteristic strong and early enhancement are indicators of PSH (5). Unusual manifestations include a mass tumour with a cystic appearance. Peckcolaklar and coworkers reported a case of PSH mimicking hydatid cyst (10). Metastatic lung tumours may be suspected in case of multiple nodules like in our second observation. Multiple slow-growing lesions have been reported by other authors (11). These cases suggest that multiplicity of the lesions does not necessarily imply biological aggressiveness. Metastases in mediastinal nodes have been also reported. Cho et al reported that this metastatic ability may be induced by an increased expression of a matrix metalloproteinase S (MMP-9) (12). This finding

doesn't worsen the prognosis which remains good. In our 2 cases, the lymph nodes were negative for tumour. The radiological findings may indicate the diagnosis but the final diagnosis remains based on histological examination. Macroscopic findings consist in a well circumscribed mass varying from 0, 3 to 8 cm. the cut-surface is generally hemorrhagic or cystic. Microscopic findings consist generally in a neoplasm with 4 possible histological components: papillary, sclerotic, solid and hemorrhagic. Most tumours have at least three of these components and a minority have only two (13). The presence of granulomas like those observed in our second case is unusual and hasn't been reported in the literature. Tumour cells are divided in 2 types: cuboidal cells and round to polygonal cells (5). Round and polygonal cells form generally solid sheets. They represent the true neoplastic cells. Cuboidal cells line generally the papillary structures. Some authors suppose that these cells have the same origin based on a uniform pattern of monoclonality and on immunohistochemical findings. They attribute the differences in their morphological phenotype to their different mature status (14). In the other hand, other authors advocate that these cells have different origins. Wang and coworkers demonstrated, based on immunohistochemical studies, that the cuboidal cells originate from type II pneumocytes and are the result of a responsive proliferation in the tumorigenesis. These cells express the TTF1, EMA, the cytokeratin and the surfactant proteins. The latter marker is expressed only in type II alveolar epithelium. The cuboidal cells express EMA but are negative with the cytokeratin, the TTF1 and the surfactant protein. These findings point their possible origin from respiratory primitive epithelium (15). Our immunohistochemical results are similar to those of Wang's study; in fact, the TTF-1 was expressed by the cuboidal cells. Histological differential diagnoses consist essentially in carcinoid tumours and papillary adenocarcinoma. In carcinoid tumours, in addition to the morphological characteristics, neuroendocrine markers are positive whereas they aren't expressed in PSH. Adenocarcinoma shows generally many nuclear atypia and many mitoses. Iyoda and coworkers compared 26 PSH to papillary adenocarcinoma and showed that the Ki-67 labeling index of PSH was significantly lower than that of adenocarcinoma.

Different pathways seem to play a key role in the tumorigenesis of PSH. Ami and colleagues proved that the genetic abnormalities that mediate its development may be in relation with an aberrant mammalian target of rapamycin signaling (6). Chiang and coworkers proved that the

Wnt/beta-catenin pathway is involved in this genesis (7). The PSH are benign and their treatment is based on anatomic resection (17).

PSH are benign tumours with a good behaviour. Although the progress made in radiological techniques to elucidate these lesions, the diagnosis remains based on histological findings. The treatment is surgical and no case of malignant transformation has been reported in the literature.

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