

Sclerosing hemangioma of the lung

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SUMMARY

Sclerosing hemangioma of the lung is a rare benign tumor that can metastasize to regional lymph nodes. It is a neoplasm that originates from type II pneumocytes and primitive respiratory epithelium. It is most common in middle-aged women in form of peripherally localized node. Tumor is composed of two types of cells, and there are papillary, solid, sclerotic, and hemorrhagic histological pattern. Preoperative and intraoperative diagnosis of the tumor is difficult. In a 53-year-old female patient, radiologically was discovered hyperdense nodule with smooth margins, which was followed for two years, and then underwent enucleation of the tumor. The tumor was 25 mm in diameter and consisting of round and cuboidal cells. Microscopically, it showed papillary and solid arrangement. Based on immunohistochemical analysis, we made a diagnosis of sclerosing hemangioma of the lung. Three months after surgery, the patient was in good health condition. Because of the numerous differential diagnostic dilemmas, diagnosis is almost always based on permanent paraffin sections, using a wide range of immunohistochemical analysis. Type of surgery depends on the tumor location, and in case of larger tumors, lymph node dissection needs to be done as well.

Key words: Lung Neoplasms; Pulmonary Sclerosing Hemangioma; Immunohistochemistry

INTRODUCTION

Sclerosing hemangioma of the lung (SHL) is a rare benign tumor that was first described by Liebow and Hubbel in 1956 (1). Its morphology is well described, but histogenesis is not yet fully elucidated (2). Although it is one of the benign tumors, rare cases with regional lymph node metastases were observed (3, 4).

SHL is predominant in women in fifth decade (5). It typically manifests as asymptomatic, well demarcated, and peripherally localized node (2). Microscopically, it is consisted of round stromal cells and cuboidal superficial cells, and there are four histological patterns (papillary, sclerotic, solid and hemorrhagic) (6, 7).

In most studies, SHL diagnosis was made after surgical extirpation of the tumor using immunohistochemical analysis (8, 9). Complete surgical resection of the SHL is the method of choice for treatment of these patients (9, 10). We presented a case of a female patient with SHL, which was discovered because of worsening of chronic obstructive pulmonary disease.

CASE REPORT

A 53-year-old woman was admitted to our hospital for resolving etiology of node, which was incidentally detected because of the worsening of chronic obstructive pulmonary disease. The patient complained of shortness of breath, choking, coughing, and expectoration of greenish sputum. In the left lower lobe, CT of the chest detected peripherally located, well circumscribed, round, hyperdense node with smooth margins, 25 mm in diameter, most likely to be hamartoma. There was a medium degree of obstructive lung ventilation disorder and mild degree of pulmonary hypertension, while bronchoscopic examination detected thick pus in trachea and bronchial tree bilaterally. Comorbidities included hypertension and hypothyroidism, both medically corrected. At that moment the etiology was not clear, and the patient was not motivated for surgical treatment. It was decided to follow the patient using imaging methods, every six months. Radiologic appearance of the lesion has remained unchanged over the next two years, when the patient accepted surgical treatment. Left anterolateral thoracotomy was

performed; node was fully enucleated and diagnosed as benign tumor on frozen sections. Tumor measured 25 mm, it was of medium firm consistency and yellow-whitish colored with brown areas on cut surface.

Histologically, the tumor consisted of round stromal cells and cuboidal superficial cells in papillary and solid arrangement (Figure 1). Round cells showed mild atypia, had finely dispersed chromatin, scant cytoplasm, no mitoses and no visible nucleoli. Cuboidal cells with scant eosinophilic cytoplasm were on the surface of largely sclerosed papillae. Tumor contained numerous trapped small airways, dystrophic calcification, clusters of lymphocytes and macrophages with foamy cytoplasm. The blood vessels were dilated and filled with blood. Immunohistochemically, round and cuboidal cells were EMA (Figure 2) and TTF1 positive, pan-CK was negative in round cells and positive in cuboidal cells (Figure 3), CK7, chromogranin, and synaptophysin were focally positive in round cells and negative in cuboidal cells. Estrogen receptors, progesterone receptors, HMB45, Melan-A, actin, desmin, CK5/6, calretinin, and CD34 were negative in both cell types. Based on histological features and immunohistochemical profile, diagnosis of SHL was made. The patient was discharged on the third day after surgery and three months after enucleation of the tumor she was feeling good.

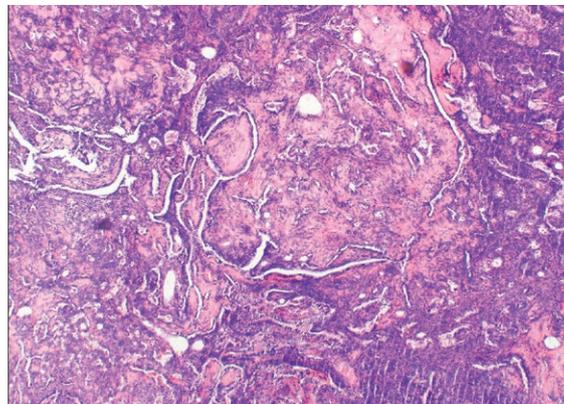


Figure 1. Sclerosing hemangioma of the lung: papillary and solid arrangement of tumor cells, H&E x 10

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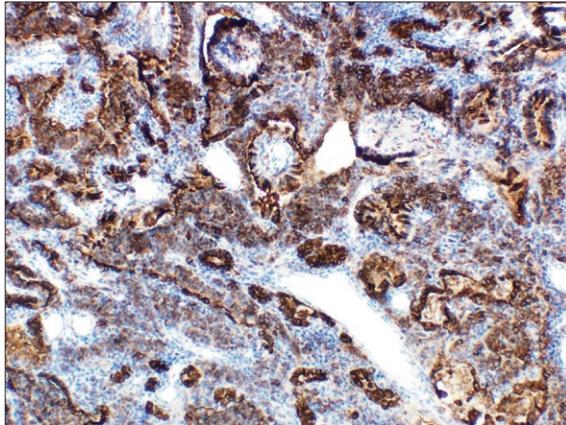


Figure 2. Positivity of round and cuboidal cells, EMA x 10

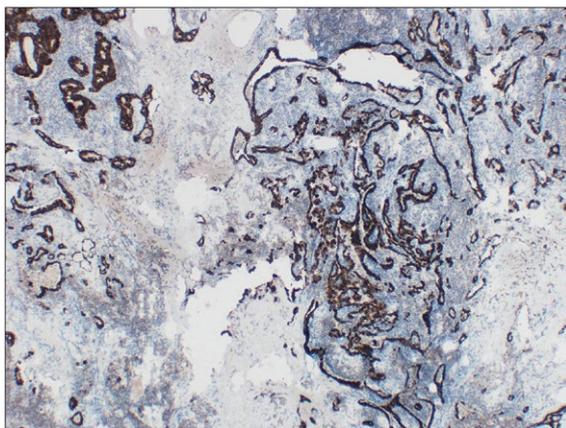


Figure 3. Positivity of cuboidal cells and negativity of round cells, PanCK x 4

DISCUSSION

Since the discovery of SHL numerous studies have shown that it is a rare benign and slow growing tumor of the lung (11, 12). The original concept, based on electron microscopy, indicated that it is a tumor originating from endothelial cells (1, 13). However, recent immunohistochemical analysis showed that it is a tumor originating from reactively altered type II pneumocytes and primitive respiratory cells (14, 15).

Although SHL is considered benign, several cases have been described in the literature (tumors larger than 30 mm) with metastases to regional lymph nodes (3, 15). Therefore, Motoki and colleagues pointed out that dissection of lymph nodes is necessary if SHL is larger than 30 mm (16). In our case, the tumor was less than 30 mm and lymph nodes less than one centimeter, so we performed enucleation of the tumor only. At the time of diagnosis of SHL our patient was 46 years old which is consistent with the data of other authors (2, 17). The symptomatic patients (40%) most often present with cough, hemoptysis, while chest pain and shortness of breath are rare (5). Analyzing the relationship between tumor size and the probability of clinical symptoms, Yang and colleagues came to conclusion that there is no correlation between symptoms and tumor size (9). Symptoms, which our patients had, were more likely a result of complications of primary pulmonary disease and not the tumor itself. The tumor in our case was 25 mm in diameter, which is consistent with the data from the literature (2). More than 95 % of the tumors were

peripherally localized, in the form of a solitary node or mass on CT images, with appearance of homogeneous soft-tissue lesions with clear margins and numerous calcifications, as it was in our case. However, some authors described the extremely rare endobronchial and endobronchiolar localization of SHL, radiologically resembling a hydatid cyst of the lung (2, 18, 19). The radiological differentiation of SHL can further be performed by bronchial arteriography and PET (20). SHL is well demarcated, non-capsulated, yellowish to brown colored, consisting of round stromal cells and cuboidal superficial cells resembling type II pneumocytes. Round cells show discrete atypia. Their cytoplasm is light eosinophilic, nuclei are hypochromatic without visible nucleoli and with no mitosis as in cuboidal cells (14, 15). Large vascular spaces are surrounded by epithelioid cells, numerous hemosiderophages, foamy macrophages, cholesterol crystals and psammoma bodies, and the surrounding lung parenchyma is compressed (21). Immunohistochemically, round and cuboidal cells are EMA and TTF1 positive. PanCK and surfactant proteins A and B were negative in round, but positive in cuboidal cells. The round cells showed focal CK7 positivity. Calretinin and CK5/6 were negative in both cell types. This immunophenotype suggests that round cells are real neoplastic cells that originate from multipotent respiratory epithelium, while cuboidal cells are reactive type II pneumocytes (14). SHL is histologically consisted of mixture of papillary, sclerotic, hemorrhagic and solid arrangements, with a predominant papillary arrangement, as in our case (11). Iyoda and associates immunohistochemically analyzed biological activity of SHL by determining the expression of Ki-67 and p53 and showed that SHL has no malignant phenotype and that proliferative activity is low. Based on the results of this study they suggested that potential recidives are most likely due to incomplete tumor resection (22).

Despite numerous radiological and pathological criteria, preoperative diagnosis of SHL is difficult (10, 11). Differential diagnosis includes primary and metastatic clear cell tumors of the lung, metastases of papillary thyroid cancer, carcinoid, hamartoma, hemangioma, malignant teratoma, arteriovenous malformations, inflammatory processes, mesenchymal and mesothelial tumors; therefore, a wide range of immunohistochemical markers is needed to confirm the diagnosis of SHL, as we have done in our case (6). Complete surgical resection is the treatment of choice in therapy of SHL and type of surgery depends on preoperative and intraoperative diagnosis and tumor localization (9, 10, 23). Because most SHL are peripherally localized, enucleation is most frequently used surgical method, as it was in our case (9). Bearing in mind that SHL is tumor of unknown histogenesis and unclear biological behavior, we need to emphasize the importance of monitoring such patients.

CONCLUSION

SHL is a rare benign lung tumor that originates from type II pneumocytes and primitive respiratory epithelial cells. The differential diagnosis includes primary and secondary lung tumors of different origin and use of broad panel of immunohistochemical markers is essential in diagnostic approach. Complete surgical resection is the treatment of choice for SHL, but in case of larger tumors and those associated with enlarged regional lymph nodes, lymphadenectomy is also needed with a mandatory patient's follow-up.

Conflict of Interest

We declare no conflicts of interest.

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