CASE REPORT

Myopericytoma Presenting As Multiple Pulmonary Nodules

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Abstract

Myopericytoma is a benign tumor composed of cells that show apparent differentiation towards putative perivascular myoid cells called myopericytes. To date, only a small number of series describing myopericytomas have been reported. Here, we report a case of pulmonary myopericytoma presenting as multiple nodules in a 26-year-old man. Clinical presentation, radiological features and histopathologic findings of the patient are also discussed. The result of the histology combined with the immunohistochemical analysis led to a diagnosis of myopericytomas. To our knowledge, this is the first report of myopericytoma showing pulmonary involvement.

Key words: myopericytoma, histopathology, pulmonary


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Introduction

Myopericytoma is a recently described mesenchymal neoplasm of pericytic cells demonstrating myoid differentiation (1). Histologically, it is characterized by a concentric proliferation of oval to spindle-shaped cells around vascular channels. Myopericytoma appears to arise in a wide range at presentation and the most common anatomic setting for myopericytoma is the skin and superficial soft tissues of distal extremities (2). In the past few years, a wider distribution is described, including the oral or nasal region, the trunk, the intravascular location and even the intracalvarium (2, 3). This unusual tumor had hitherto not been reported in the lung. Here, we present a case who was diagnosed as myopericytoma of the lung, aiming to create awareness regarding this particular entity and the need to differentiate these tumors from usual metastatic carcinoma of the lung.

Case Report

A 25-year-old Chinese man presented with dry cough and slight expectoration, but no dyspnea, no hemoptysis, and no fever. The symptoms began five months prior to hospitalization without manifest motivations. The patient denied any positive family history or history of past illness. The physical examination findings showed no apparent abnormality. All of the laboratory data were within normal limits. Computed tomography (CT) and magnetic resonance imaging (MRI) of the chest was notable for the bilateral, heterogeneously, multiple nodules (see Fig. 1, Supplemental Digital Content 1, which showed the image features of CT and MRI). Bronchoscopy showed just light thickening of the mucosa of anterior segmental bronchus in the left upper lobe. Open lung biopsy was performed and two masses were resected from the right lower and middle lobe respectively. The partially resected middle lobe of right lung measured 8.5×5×2 cm. A mass was found on its cut section, its size was 1.7×1.5×1 cm (not shown). All tumors appeared to be well-circumscribed, but uncapsulated with tan-brown color tissues. The microscopic examination of the mass showed that the lesions were composed of oval to spindle-shaped cells with a striking concentric arrangement of cells around numerous variably sized lesional blood vessels. Tumor cells were ill-defined cytoplasmic membranes with eosinophilic cytoplasm. The nuclei appeared bland. In some areas, well-formed and dilated vessels, imparting the feature of a hemangiopericytoma, were observed. Mitotic activity and necrosis were absent (see Fig. 2, Supplemental Digital Content 2, which showed the pathology of the specimens from the

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right lower lobe lung).

Immunostaining demonstrated an accentuated vascular proliferative pattern with CD31 and CD34 antibody which stained endothelial cells. The cells surrounding vascular lumina are strongly positive for vimentin and smooth muscle actin, but negative for desmin or S-100 (see Fig. 3). By correlating the histopathological features with the immunohistochemical findings, a diagnosis of myopericytoma was established.

Since there were no symptoms after conventional postsurgical treatment and anti-infective therapy, we decided to carefully follow the patient without interventions. The patient has been followed up and no recurrence or metastasis has been found after 3 years.

Discussion

The concept of myopericytes was originally proposed by Dictor et al in 1992 to describe an unusual tumor in a boy. Lesional cells that the authors termed ‘myopericytes’ displayed features intermediate between pericytes and vascular smooth muscle cells (4). Subsequently, the term was used by Requena et al in 1996 as an alternative name for myofibroma (5). In 1998, the term myopericytoma was adopted by Granter et al to describe benign tumors characterized histologically by striking concentric perivascular proliferation of spindle cells and showing apparent differentiation towards perivascular myoid cells (1). WHO soft-tissue tumor pathologic and genetic categorization in 2002 classified myopericytoma as an independent type into angiopericytoma. Myopericytoma can arise at any age. The lower extremities are most commonly affected, followed by the upper extremities, the head and neck region and the trunk (2). Myopericy-
Figure 3. CD34 stained the endothelium of vessels, but the perivascular concentric tumor cells were not immunoreactive (A: magnification ×200). The perivascular cells expressed reactivity to smooth muscle actin (B: magnification ×200).

toma occurring in cavity mucosa, intracalvarium and thoracic spine have been recently reported (3, 6, 7). Unusual presentations of myopericytoma including myopericytoma associated with a chronic scar and with trauma have been described in the literature (8, 9). Some cases were multicentric involving a single or multiple anatomic regions also. To the best of our knowledge, the present case is the first reported pulmonary myopericytoma, further expanding the list of locations.

Myopericytoma showed a wide range of growth patterns (10). The histological features of the tumors are the presence of numerous blood vessels with a concentric perivascular arrangement of ovoid, plump, spindled and/or round myoid cells with eosinophilic cytoplasm. The neoplastic cells were diffuse and immunoreactively positive for vimentin, smooth muscle actin, and often for h-caldesmon, whereas desmin is usually negative. Ultramicrostructure as viewed under electronmicroscope helps to demonstrate its derivation of pericytes. The diagnosis of myopericytoma was made on the basis of the prominent morphological and immunoreactive findings.

Myopericytoma generally presents as a painless, slow-growing nodule, rarely measuring >2 cm, except for a few of infiltrative growth. In those cases, though seldom, if the tumor displays cellular pleomorphism, highly mitotic activity, necrosis should be diagnosed as malignant myopericytoma, which may lead to local recurrences and rarely metastases (11).

The differential diagnoses include a series tumor, such as angioleiomyoma, hemangioperithelioma, myofibroma and glomus tumor. Despite the overlapping of morphologic features with angioleiomyoma and myofibroma, myopericytoma represents a broad morphologic spectrum characterized by distinct concentrically, perivascularly growing myoid tumor cells that stain positively for smooth muscle actin and often for h-caldesmon, whereas they remain unlabelled for desmin, suggesting a less-differentiated smooth muscle phenotype. Angioleiomyoma is usually a painful nodule occurring commonly on the lower limb of females. Angioleiomyomas are tumors composed of mature smooth muscle cell bundles with abundant vascular channels which frequently show desmin immunoreactivity in the smooth muscle bundles. As for glomus tumors, pain and/or exquisite tenderness are the characteristic complaints. The ovate to round neoplastic cells of clear boundary are abundant of understained eosinophilic cytoplasm, grouping non-concentrically around blood vessels. Notably, spindle cells are rare in glomus tumors. Hemangioperithelioma, now categorized as under-pleural solitary fibrous tumour, shows staghorn-shaped spaces with an intervening proliferation of cells. In addition, vimentin and smooth muscle actin immunoreactivity is very rarely seen in cases of hemangiopericytoma and, even when present, it is only focal and weak. Myofibroma is a tumor occurring in superficial/deep part of soft-tissues, occasionally in an internal organ. Multi-myofibroma is called myofibromatosis. The presence of a zonation/biphasic appearance is the most characteristic of myofibroma. Myopericytoma can exhibit a spectrum of growth patterns that overlap with myofibroma (12). Tumors can be designated as myopericytoma or myofibroma depending on the predominant growth pattern. In spite of the usual lobus intermedius tumors, the differential diagnosis of the present case should also include other etiologies of multiple lung nodules, such as inflammatory pseudotumor, tuberculosis, sarcoidosis, lymphoma, primarily lung cancer and metastatic tumor.

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References

2. Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical

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