

# Case report

## Pulmonary myopericytoma: a case report and review of the literatures

CAO Jian-hua, XU Jin-ping, LI Yong-cheng, LAI Jun and LI Qiang

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**M**yopericytoma belongs to a family of benign tumors that exhibit a myoid/pericytic line of differentiation.<sup>1</sup> The most common anatomical sites for myopericytoma are the skin and superficial soft tissues of the distal extremities.<sup>2</sup> With increased clinical recognition, a wider distribution has been described,<sup>3-5</sup> however, a pulmonary lesion has never been reported. We now report a case of primary pulmonary myopericytoma and review the literature on this rare entity.

### CASE REPORT

A 52-year-old woman with a 21-year history of rheumatic heart disease (mitral stenosis and tricuspid incompetence) and a 2-year history of nodule in the right lower lung had no respiratory symptoms. Physical examination revealed a rumbling, mid-diastolic murmur (grade 3/6) over the mitral valve area. The remaining physical examination was normal and no skin lesions or subcutaneous mass was detected. Routine blood tests, biochemical tests, urine analysis, myocardial enzymes, anti-streptolysin "O", and rheumatoid factor were within normal limits and the ESR was 15 mm/h. Cardiac ultrasound showed mild mitral stenosis and tricuspid incompetence, mild pulmonary hypertension, biatrial enlargement and normal left ventricular systolic function. There was a nodule in the right lower lung field upon chest radiography (Figure 1A). Routine chest CT scans demonstrated that a well-delineated round mass was located in the right lower lobe; no calcification or fat attenuation was seen (Figure 1B). After injection of contrast medium, the peripheral portion of the mass was markedly enhanced, but the central portion was poorly enhanced (Figure 1C). No other pulmonary lesion and no lymphadenopathy was detected. She underwent the right lower pulmonary lobectomy for a suspected lung tumor. The patient is presently alive and well with no evidence of recurrence or metastatic disease 3 years after surgery.

Pathological examination demonstrated macroscopically, a 3.5 cm × 3.0 cm × 2.0 cm, gray, circumscribed mass in the right lung lower lobe. Microscopically, a well-circumscribed with encapsulated, nodular neoplasm was present, and cut sections showed that it was homogeneous, without areas of hemorrhage within it. The neoplasm contained numerous thin-walled blood vessels and was composed of ovoid, plump-spindled, and/or

round myoid tumor cells with an eosinophilic cytoplasm and spindled or round nuclei. Characteristically, there was a distinct perivascular and concentric growth of neoplastic cells, which had Catherine wheel features or a typical spinning off from the vessel walls was seen (Figure 2). No heteromorphism, mitotic figures, lymphovascular invasion or necrosis was observed in the tumor cells.

Under immunohistochemical examination, the tumor cells showed diffuse staining for smooth muscle actin (SMA; Figure 3A) and vimetin (Figure 3B) but were negative for desmin, cytokeratins, HMB45, epithelial membrane antigen, chromogranin, P63, synaptophysin, carcinoembryonic antigen and S-100 protein. Only endothelial cells showed staining for CD34 (Figure 3C). As a result of these pathological and immunohistochemical findings, pulmonary myopericytoma was diagnosed.

### DISCUSSION

Perivascular neoplasms traditionally include glomus tumors and hemangiopericytoma.<sup>6</sup> Hemangiopericytoma was described in 1942 by Stout and Murray as a distinctive soft tissue neoplasm, presumably of pericytic origin, which shows a characteristic well-differentiated staghorn branching vascular pattern.<sup>1,7</sup> Currently, it appears that this growth pattern is non-specific and appears in numerous, unrelated benign and malignant lesions. Myopericytoma is now classified within the heterogeneous group of hemangiopericytoma-like neoplasms.<sup>8</sup>

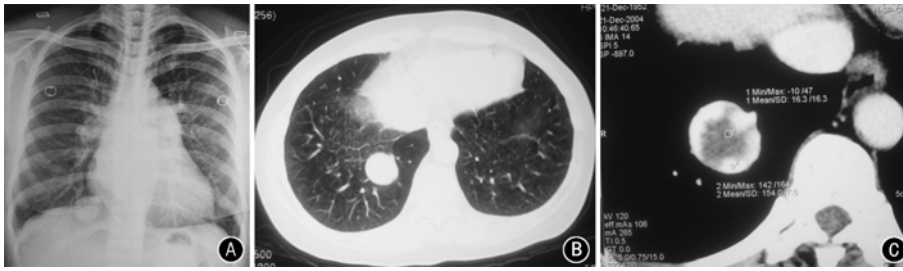
A number of lesions diagnosed as hemangiopericytoma were previously referred to as myopericytoma. In 1996 Requena et al<sup>9</sup> suggested that some cutaneous adult

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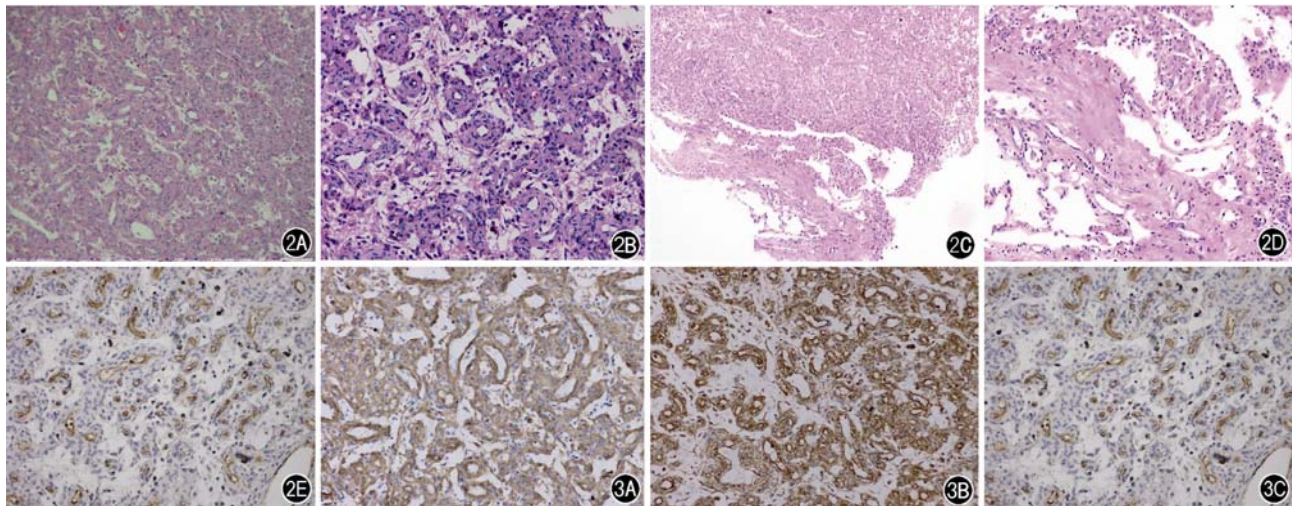
Department of Respiratory Medicine (Cao JH, Li YC and Lai J), Department of Pathology (Xu JP), Xiaoshan No.1 People's Hospital, Medical College of Hangzhou Normal University, Hangzhou, Zhejiang 311201, China

Department of Respiratory Medicine, Changhai Hospital, Second Military Medical University, Shanghai 200433, China (Li Q)

Correspondence to: Dr. LI Yong-cheng, Department of Respiratory Medicine, Xiaoshan No.1 People's Hospital, Medical College of Hangzhou Normal University, Hangzhou, Zhejiang 311201, China (Tel: 86-571-82621086 ext 2516. Fax: 86-571-82623324. Email: Lyc192002@yahoo.com.cn)



**Figure 1.** A round mass appears in the right lower lung field. A well-defined mass is visible in the right lung lower lobe. The peripheral zone of the mass is enhanced strongly, but the central zone is enhanced poorly (**A**: chest radiograph. **B**: unenhanced CT. **C**: enhanced CT).



**Figure 2.** The tumor was composed of ovoid-spindled, myoid tumor cells with a characteristic multilayered concentric perivascular growth (HE, original magnification, **A**  $\times 40$ ; **B**  $\times 200$ ; **C**  $\times 400$ ); Pulmonary myopericytoma with encapsulated (HE, original magnification, **D**  $\times 40$ ; **E**  $\times 100$ ).

**Figure 3.** Immunohistochemical staining of pathological sections (original magnification  $\times 200$ ). The tumor cells in this pulmonary myopericytoma are diffusely positive for SMA (**A**) and vimentin (**B**). Endothelial cells stained only for CD34 (**C**).

myofibromas were actually benign perivascular neoplasms, composed of cells with the morphological and immunohistochemical features of immature pericytes or myopericytes. However, it was not until 1998 that the concept of neoplasms that showed perivascular myoid differentiation was established by Granter et al<sup>10</sup> and the terms myopericytoma or perivascular myoma were proposed for these neoplasms. Histologically, myopericytoma is recognized by the presence of a distinctive concentric (onion skin) perivascular proliferation of round-to-spindle cells with a myoid appearance. As a tumor derived from perivascular myoid cells, myopericytoma is characteristically reactive for muscle-specific actin and SMA.<sup>10,11</sup> Desmin reactivity may occasionally be observed. In the present case, positive vimentin was also observed. Myopericytoma generally shows a benign clinical course and follow-up of the current case supports their benign nature of myopericytomas in that there was no evidence of disease recurrence after 3 years.

The differential diagnosis includes myofibroma, angioleiomyoma and glomus tumor. Myopericytoma represents a perivascular myoid neoplasm that shares morphological features with myofibroma, angioleiomyoma and glomus tumor. Despite overlapping morphologic features, there are enough distinctive

features to characterize this lesion as a separate entity. The myofibroma of the adult type consists of a distinct biphasic pattern of varying proportions.

One of the two patterns is fascicular and is composed of mature spindle cells that resemble smooth muscle cells. The other pattern is cellular with immature-appearing cells and a haemangiopericytoma-like vascularity with thin-walled branching vessels. The more immature areas are the dominant pattern in most tumors that have been described to date.<sup>6,12</sup> Angioleiomyoma contains thick-walled vessels, especially in the periphery of the lesions, and is composed predominantly of elongated spindle cells that stain positively for desmin (besides actin and SMA), which is usually negative in myopericytoma, or may be expressed only focally.<sup>13,14</sup> Finally, glomus tumor most closely resembles myopericytoma and overlapping morphological features are present. Immunohistochemical staining is of little help in distinguishing the two entities. However, histologically, glomus tumor usually lacks the characteristic concentric perivascular growth of neoplastic cells typically seen in myopericytoma.<sup>15,16</sup>

Although myopericytoma appears to arise most commonly in middle adulthood, there is a wide age range at presentation. The vast majority of lesions arise in the

dermis and subcutaneous tissue, with an apparent predilection for the extremities. Duration of the lesion before excision is not known for most patients, and none of the lesions is painful. Most myopericytomas behave in a benign fashion; however, rare cases are malignant. One of the 54 cases reported by Mentzel et al<sup>13</sup> was malignant. The treatment of choice is surgical excision. Although myopericytomas have been reported recently, we presented a rare case of pulmonary myopericytoma, which has not been previously reported. Differential diagnosis is very difficult; while routine chest CT scans demonstrated a well-delineated round mass, no calcification or fat attenuation was seen. After injection of contrast medium, the peripheral portion of the mass was markedly enhanced, but enhancement of the central portion was poor. In these cases, myopericytoma should be considered in the differential diagnosis. Complete surgical removal would be expected to be associated with an excellent clinical outcome.

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