CASE REPORT

A surgically treated pulmonary sclerosing haemangioma that clinically and radiologically mimicked a thrombosed pulmonary artery aneurysm

Keiji Yamanashi, Tsuyoshi Shoji, Yuichiro Ueda, Cheng-long Huang

Respiratory Disease Center,
Tazuke Kofukai Foundation,
Medical Research Institute,
Kitano Hospital, Osaka, Japan

Correspondence to

Dr Keiji Yamanashi,
jjiike6180@yahoo.co.jp

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SUMMARY

We report the case of a 70-year-old woman with a surgically treated pulmonary sclerosing haemangioma that clinically and radiologically mimicked a thrombosed pulmonary artery aneurysm. She underwent intravenous thrombolytic therapy because of acute pulmonary embolism in the bilaterally segmental arteries. The pulmonary embolism had almost disappeared except for a nodular lesion in the middle lobe branch of the pulmonary artery, and a thrombosed pulmonary artery aneurysm with thrombus was suspected in the CT image. The patient underwent a video-assisted thoracoscopic middle lobectomy. The final pathological diagnosis was sclerosing haemangioma. She had experienced no recurrence for 1 year after surgery, and we continue to take great care during her follow-up.

BACKGROUND

Sclerosing haemangioma (SH) of the lung is a rare tumour comprising a mixture of four histological patterns, namely, papillary, sclerotic, solid and haemorrhagic. It is defined as a very low-grade malignancy, with a few cases involving regional lymph node metastasis.^{1 2} We report a case of a surgically treated SH that clinically and radiologically mimicked a thrombosed pulmonary artery aneurysm.

CASE PRESENTATION

Case report

A 70-year-old woman presented with dyspnoea and syncope. Oxygen saturation was 89% in room air, pH 7.49, PaO₂ 62.2 mm Hg and PaCO₂ 34.4 mm Hg. D dimer was elevated to 20.0 µg/mL (reference range: <1.0 µg/mL). Acute pulmonary embolism (PE) was suspected and the patient underwent enhanced CT, which showed major PE in the bilaterally segmental arteries (figure 1A), and deep venous thrombus in the left calf vein (figure 1B). She underwent intravenous thrombolytic therapy with 240 000 urokinase units a day; after 5 days the repeated enhanced CT showed that the PE had almost disappeared except for a nodular lesion in the middle lobe branch of the pulmonary artery (figure 1C), and a thrombosed pulmonary artery aneurysm exceeding double the size of the peripheral pulmonary artery was suspected in the three-dimensional CT image (figure 1D). The intravenous thrombolytic therapy was considered ineffective and a thrombosed pulmonary artery aneurysm was suspected; the patient underwent a video-assisted thoracoscopic middle lobe lobectomy. Macroscopically,

the nodule in the middle lobe was 8×8 mm in size, blackish-brown and circumscribed without invasion of the surrounding tissue, similar to a thrombus (figure 2A). Microscopically, the tumour cells were arranged in various patterns including papillary, sclerotic, solid and haemorrhagic (figure 2B). Immunohistochemically, the tumour cells were immunopositive for epithelial membrane antigen, thyroid transcription factor-1 (figure 2C). The final diagnosis was SH. The patient remains alive after 1 year with no sign of recurrence of the tumours.

OUTCOME AND FOLLOW-UP

The patient underwent a video-assisted thoracoscopic middle lobe lobectomy. Macroscopically, the nodule in the middle lobe was 8×8 mm in size, blackish-brown and circumscribed without invasion of the surrounding tissue, similar to a thrombus (figure 2A). Microscopically, the tumour cells were arranged in various patterns including papillary, sclerotic, solid and haemorrhagic (figure 2B). Immunohistochemically, the tumour cells were immunopositive for epithelial membrane antigen, thyroid transcription factor-1 (figure 2C). The final diagnosis was SH. The patient remains alive after 1 year with no sign of recurrence of the tumours.

DISCUSSION

There were two reasons for the indication of surgical treatment in this case. First, Meyer *et al*³ showed three contributing factors indicating surgical intervention of PE; (1) patients have haemodynamic disturbances, (2) medical treatment is thought to be unacceptable and (3) thrombolytic treatment is contraindicated. In our case, unacceptable medical treatment was a factor for surgical intervention. Second, Veldtman *et al*⁴ showed that conservative treatment for pulmonary artery aneurysm may be advocated when the patient has no symptoms and there are no associated congenital lesions, no right ventricular dysfunction and no significant pulmonary arterial hypertension or left to right shunt; however, Fukai *et al*⁵ showed that once pulmonary artery aneurysm has been diagnosed, surgical resection should be considered because of a possible fatal outcome such as rupture of the aneurysm. Moreover, Steireif *et al*⁶ showed a cut-off diameter of the aneurysm, indicating an increasing risk for complications, such as dissection or rupture, has not been defined yet, and surgical intervention is inevitable for aneurysms exceeding double the size of the peripheral pulmonary artery. Thus, the patient underwent surgical

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Figure 1 Enhanced CT showing pulmonary embolism in the bilaterally segmented arteries (A) and deep venous thrombus in the left calf vein (B). (C) Enhanced chest CT after intravenous thrombolytic therapy showing that the pulmonary embolism has almost disappeared except for a nodular lesion in the middle lobe branch of the pulmonary artery. (D) Three-dimensional CT showing a nodule near the middle lobe branch of the pulmonary artery, similar to a thrombosed pulmonary artery aneurysm exceeding double the size of the peripheral pulmonary artery.

treatment in our case. SH is a rare primary pulmonary tumour and thought to be an epithelial tumour derived from primitive epithelial cells. Histologically, the hallmark of SH is heterogeneity, with various architectural patterns, namely papillary, solid,

sclerotic and haemorrhagic. Preoperative and intraoperative diagnosis of the tumour is difficult because its pathological features are similar to those of atypical adenomatous hyperplasia and bronchioloalveolar carcinoma.² Although SH is considered a

Figure 2 The specimen showing an 8x8 mm nodule-like thrombus macroscopically (A) and papillary, sclerotic, solid and haemorrhagic patterns in most portions of the tumour microscopically (B). (C) The tumour cells were immunopositive for thyroid transcription factor-1.

Learning points

- ▶ A surgically treated pulmonary sclerosing haemangioma (SH) that clinically and radiologically mimics a thrombosed pulmonary artery aneurysm is rare.
- ▶ SH of the lung is rare and defined as a very low-grade malignancy, with a few cases involving regional lymph node metastasis.
- ▶ Great care should be taken during follow-up of patients with SH because of the tumour's unclear potential malignancy.

very low-grade malignancy, rare cases of lymph node metastasis have been reported in the literature. Miyagawa-Hayashino *et al* reviewed 100–200 cases of SH, and their rough estimate of the incidence of lymph node metastasis was between 2% and 4%. The clinical significance of metastasis to the regional lymph nodes in SH is unclear, and it is controversial whether nodal dissection should be performed in SH.¹ In our case, after pathological diagnosis we explained these data to the patient, and she hoped not to require a secondary operation with nodal dissection. Recurrence of SH after surgery is very rare. Wei *et al*⁷ reported a case with local recurrence 10 years after surgery, thus we must take great care during follow-up for our patient.

In conclusion, SH is a rare lung neoplasm that has been defined as a very low-grade malignancy. Great care should be

taken during resection and patient follow-up because of the tumour's unclear potential malignancy. Further investigations with a larger number of cases are needed to establish effective treatment strategies.

Contributors YU participated in the clinical diagnosis. C-IH gave information about lung surgery. TS supervised the concept and design of the manuscript and gave final approval of the version to be published. All authors read and approved the final manuscript.

Competing interests None.

Patient consent Obtained.

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