

Pulmonary artery sarcoma diagnosed using intravascular ultrasound images

Yoshiaki Okano, Toru Satoh, Teiji Tatewaki, Takeyoshi Kunieda, Satoru Fukuyama, Naoki Miyazaki, Yasuo Beppu

Division of Cardiology and Pulmonary Vascular Medicine, Department of Internal Medicine, National Cardiovascular Center, 5-7-1 Fujishirodai, Suita, Osaka, Japan
Y Okano
T Satoh
T Tatewaki
T Kunieda

Division of Respiratology, Nippon Steel Cooperation Yawata Works Hospital, 1-1-1 Haruno-cho, Yahata-Higashi-ku, Kitakyushu, Japan
S Fukuyama
N Miyazaki

Division of Orthopedics, National Cancer Institute, 5-1-1 Tsukiji, Chuo-ku, Tokyo, Japan
Y Beppu

Correspondence to:
Dr Y Okano, Director,
Division of Cardiology,
Shinshiro City Hospital, 32-1
Kitabata, Shinshiro, Aichi
441-13, Japan.

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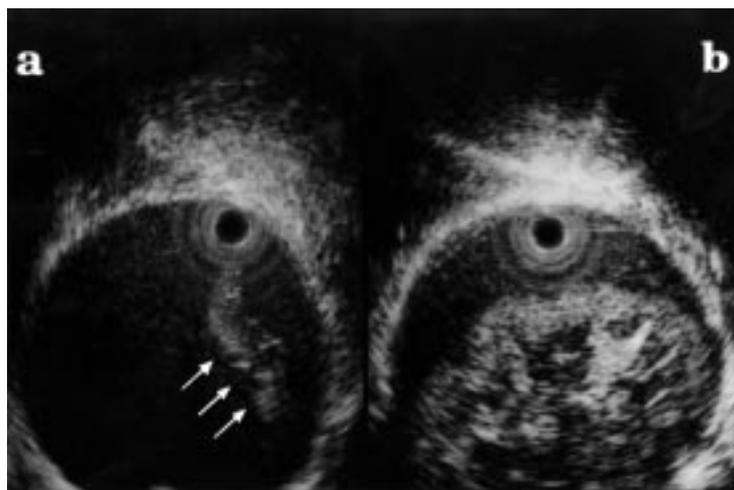


Figure 1 (a) A string shaped projection (arrows) was visualised in the proximal portion of the left main pulmonary artery by IVUS. (b) A unique echogenic mass was also visualised by IVUS which was continuous with the projection shown in (a) and had an irregular surface and a mosaic pattern containing flecked echolucent and increased echogenic areas. It was quite different from both echolucent fresh and echogenic organised thrombi.

Abstract

Primary and secondary malignant intravascular tumours of the pulmonary artery occur infrequently and the diagnosis is usually delayed as symptoms and findings from conventional examinations are non-specific. The case is presented of a patient with a pulmonary artery sarcoma, probably arising from ribs resected some years previously, in which intravascular ultrasound (IVUS) provided important diagnostic findings.

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Keywords: pulmonary artery; intravascular ultrasound; intravascular sarcoma

Although malignant tumours of the pulmonary arteries are rare,¹ they must be considered in the differential diagnosis of pulmonary embolism. Since the clinical presentation and conventional imaging features of such lesions are usually non-specific and mimic those of chronic thromboembolism,² a definite diagnosis is often delayed until surgical exploration or necropsy.³ The following unusual case of pulmonary artery sarcoma is presented because of its interesting and diagnostic images obtained by intravascular ultrasound (IVUS).

Case report

A 66 year old man was referred to our hospital with the presumptive diagnosis of chronic thromboembolic pulmonary hypertension for

evaluation and determination of surgical indication. He had had gradually progressive dyspnoea on exertion for one year prior to examination.

He had undergone a thoracotomy and resections of the right anterior fifth, sixth and seventh ribs for chondrosarcoma six years ago. On admission he was of normal stature with a slightly reduced overall general condition. Although pulmonary valve closure was slightly accentuated, no pathological murmur was detected on auscultation. Normal vesicular sounds were detected over both lungs. Routine blood values were within normal limits. Analysis of arterial blood gas tensions (breathing room air) showed pH 7.46, Pao₂ 7.6 kPa, Paco₂ 5.9 kPa, and Sao₂ of 85%. Chest radiography revealed mild cardiomegaly, dilated left main pulmonary artery, and a reduced vascular pattern of the left lung. Transthoracic echocardiography (TTE) demonstrated right atrial and ventricular dilatation suggesting the presence of pulmonary hypertension, but showed no abnormalities in the pulmonary artery. Lung perfusion/ventilation scintigraphy showed no perfusion in the entire left lung with almost normal ventilation. Computed tomographic (CT) scans and magnetic resonance images (MRI) showed a non-specific uniform mass consistent with organised thrombi in the proximal left main pulmonary artery. Pulmonary angiography confirmed complete occlusion of the left pulmonary artery and equivocal irregularity in the right descending portion with a normal peripheral vascular pattern throughout the right lung. Pulmonary artery pressure was 48/18 (30) mm Hg and cardiac output was 5.6 l/min at that time. IVUS was carried out for further clarification using a commercially available imaging system (SSD-550, Aloka Inc, Tokyo, Japan). A unique echogenic mass with an irregular surface, a string shaped projection (fig 1a), and a mosaic pattern containing flecked echolucent and increased echogenic areas (fig 1b) were clearly visualised by IVUS. These appearances differed from those seen in thromboembolic lesions⁴ and were diagnostic for a malignant pulmonary artery tumour. A second examination, including a biopsy specimen to confirm the histopathological diagnosis, was proposed but the patient declined. Attempts to locate the primary tumour and further metastases were unsuccessful. We scheduled surgical resection of the entire left lung. His respiratory condition, however, rapidly deteriorated and he died before the procedure. Postmortem necropsy revealed that the tumour of the left main pulmonary artery had

grown into the vascular lumen, distally over the bifurcation of the left pulmonary artery. Histopathological examination of the tumour resulted in a diagnosis of a myxomatous chondrosarcoma showing a close resemblance to those in the ribs resected previously. We made a final diagnosis of secondary pulmonary artery chondrosarcoma by intravascular growth or embolisation⁵ in this case, although we could not completely exclude the possibility of primary tumour.

Discussion

In this case IVUS allowed the diagnosis of malignant tumour in the pulmonary artery to be made prior to planned surgical intervention. Tumour involvements of the pulmonary vessels are not uncommon in necropsy cases of malignancy.⁶ However, most of these are microemboli and are infrequently detected clinically. Shepard *et al*⁷ reported only four cases of pulmonary artery intravascular tumour detected among 14 000 consecutive cases after CT scanning of the chest and concluded that the findings of dilated and beaded peripheral pulmonary arteries on the CT scan are highly suggestive of intravascular tumour emboli. However, we found no such characteristic features on the CT scan in this case.

Gadolinium-enhanced MRI is another potentially useful diagnostic tool to differentiate between intraluminal tumours and large thrombi. Although there is a case report in the literature documenting the value of gadolinium-enhanced MRI,⁸ our study did not show reliable differential diagnosis.

Ultrasound examinations seem to be more sensitive for qualitative evaluation of mass lesions. Only one case report,⁹ however, has been published, indicating the diagnostic significance of TTE in a patient with primary sarcoma of the pulmonary artery trunk. In

many patients it is difficult to obtain a clear image even at the level of the pulmonary valve and pulmonary artery trunk using TTE. In addition, the left main portion of the pulmonary artery is difficult to visualise even though a transoesophageal approach is applied. In these circumstances IVUS presents a potential method for evaluating the intravascular lesions because of its ability to visualise directly from inside the vessel wall.¹⁰ Ricou *et al*⁴ suggested that IVUS might be useful in assessing the location and extension of the pathological process involving the pulmonary artery.

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