Primary extraskeletal myxoid chondrosarcoma of pulmonary arteries: a rare mimic of acute pulmonary thromboembolism

Karthik Gadabanahalli, Vinay V. Belaval, Venkatraman Bhat* and Imran M. Gorur

Department of Radiology and Pathology, Narayana Health, Narayana Multispecialty Hospital and Mazumdar Shaw Cancer Center, Bangalore, India

* Corresponding author. 309, Greenwoods Apt, Royal Gardenia, Bommasandra, Bangalore 560099, India. Tel: +91-9481027387; fax: +91-80-27832648; e-mail: bvenkatraman@gmail.com (V. Bhat).

Received 17 September 2014; received in revised form 9 December 2014; accepted 11 December 2014

Abstract

Primary extraskeletal myxoid chondrosarcoma of the pulmonary arteries is a very rare entity. Multimodality imaging reports on this entity are few. Myxoid chondrosarcoma is characterized by chondroid and neurogenic differentiation in extraskeletal locations. These tumours represent fewer than 2.5% of all soft-tissue sarcomas, and are most commonly found in the lower extremities, limb girdles, distal extremities and trunk. We report an unusual case of a 31-year old man with histopathologically proven extraskeletal myxoid chondrosarcoma of the pulmonary arteries mimicking acute pulmonary thromboembolism.

Keywords: Chondrosarcoma • Pulmonary arteries • Pulmonary thromboembolism • Multi-detector computed tomography • Positron emission tomography

CASE DESCRIPTION

A 31-year old male presented to the emergency care unit with a 3-month history of gradually progressive exertional dyspnoea, with symptoms worsening for the last 10 days. The patient had an episode of syncope and two episodes of haemoptysis. He was initially admitted to an outside hospital with breathlessness, and was diagnosed to have acute pulmonary thromboembolism (PE) by computed tomography pulmonary angiography (CTPA). He was thrombolysed with IV tenecteplase, but showed no improvement. Hence, he was referred to our hospital for further management. In view of the persistent breathlessness on admission, CTPA was repeated at our hospital and revealed ‘acute PE’ with hypodense filling defects with expansion of the main pulmonary artery (MPA) extending into both pulmonary arteries, i.e. segmental and sub-segmental branches.

A saddle-shaped, irregular filling defect was seen at the MPA bifurcation (Fig. 1). There were few enlarged bronchial collaterals. No significant adenopathy was seen. There were no features of extrapulmonary extension of the lesion. The patient was put on anticoagulants and antiplatelet agents with supportive oxygen therapy. He remained symptomatic. In view of the enlarged pulmonary artery branches and absence of webs, a decision was taken to investigate with positron emission tomography (PET)–CT to rule out a neoplasm. Whole-body 18 F-FDG-PET–CT performed 2 days later revealed a mildly hypermetabolic saddle thrombus in the MPA with extension into the segmental arteries, suggestive of a neoplastic aetiology. No other hypermetabolic lesion was found in the rest of the body. The possibility of pulmonary angiosarcoma was raised. The patient was maintained on supportive oxygen therapy and medications. He underwent preoperative catheter angiography a week after admission, which did not reveal neovascularity or any additional features. The patient then underwent surgical exploration with total circulatory arrest and cardiopulmonary bypass. Hypothermia was

Figure 1: Axial images of contrast-enhanced CT at initial presentation, at the level of MPA bifurcation, show a saddle-shaped, hypodense filling defect at the MPA bifurcation extending into both RPA and LPA (open arrow). CT: computed tomography; MPA: main pulmonary artery; RPA: right pulmonary artery; LPA: left pulmonary artery.
induced with systemic cooling to 20°C, with infusion of a cold blood cardioplegia solution at 4°C through the aortic root. On surgical exposure, the MPA was found to be distended and tense. Tumour excision was performed through the MPA removing the tumour thrombi from the MPA and pulmonary artery branches up to the hilum. Tumour excision was uneventful, and near-total removal was achieved. After tumour removal, left and right pulmonary arteries were closed directly during warming. The postoperative period was uneventful. A 2-week postoperative echo showed no residual mass in the MPA and its branches. The patient was discharged on anticoagulants.

The histopathological and immunohistochemical analyses of the surgical specimen showed features of extraskeletal myxoid chondrosarcoma (EMC; Fig. 2A and B). Repeat CTPA performed after 4 weeks showed a reduction in vascularized intraluminal filling defects. The patient underwent three cycles of chemotherapy with gemcitabine and docetaxel under the supervision of an oncologist. Patient had deterioration of symptoms over the next 2 months. Repeat CTPA showed a significant increase in tumour thrombus and pericardial fluid. He expired about 9 months after presentation of the initial symptoms.

DISCUSSION

Our case describes CT and PET appearances of a rare [1] case of primary EMC of the pulmonary arteries. Histological varieties of primary pulmonary artery sarcomas (PASs) described include undifferentiated sarcoma, angiosarcoma, chondrosarcoma, rhabdomyosarcoma, leiomyosarcoma, liposarcoma, myxosarcoma, fibrosarcoma, osteosarcoma, malignant fibrous histiocytoma and malignant mesenchyma. Undifferentiated sarcoma accounts for the most common type. There are three different histological subtypes of extraskeletal chondrosarcoma: classic, mesenchymal and myxoid. Primary PASs can be intimal (predominantly intraluminal growth pattern) or mural (predominantly exophytic growth pattern). EMC makes up only 1% of all chondrosarcomas [2] and only 2% of all soft-tissue sarcomas. EMC typically occurs in the extremities, the thigh being the most common site. The average age of occurrence is around 50 years. The importance of this unique lesion lies in the fact that it mimics PE in presentation. Yi et al. [3] reported that CT findings favouring the diagnosis of PAS include a low attenuation filling defect occupying the entire luminal diameter of the proximal or MPA, with expansion of the involved arteries and extra-luminal tumour extension.

PAS also has a more heterogeneous appearance with areas of necrosis, haemorrhage and ossification. It is frequently unilateral, in contrast with the often bilateral involvement of pulmonary arteries in thromboembolic disease. In the presence of saddle thrombus at MPA bifurcation without extravascular tumour extension, the differentiation between PAS and PE becomes difficult (as in our case). PET–CT can help differentiate PAS from PE [4]. On the cardiovascular application of FDG-PET, blood thrombi show negative FDG uptake, whereas a malignant tumour, such as a cardiac sarcoma, shows positive FDG uptake. FDG-PET may show intense tracer activity within a PAS. While some tracer activity might be seen in the context of thromboembolic disease, intense tracer localization within a central pulmonary arterial filling defect strongly suggests primary pulmonary arterial malignancy [5]. In our case, positive FDG uptake in the saddle-shaped filling defect pointed towards a diagnosis of PAS. Although the management of pulmonary artery sarcoma is mainly surgical, adjuvant chemotherapy and radiation have been tried in isolated cases. A combination of gemcitabine and docetaxel has shown good response in soft-tissue sarcoma, though response was not evident in our case.

In conclusion, despite its rarity, pulmonary artery sarcoma should be considered in the differential diagnosis of PE with atypical clinical and imaging features. Subtle radiological features favouring pulmonary angiosarcoma should be looked for. In doubtful cases, FDG-PET–CT features can help arrive at the diagnosis.

Conflict of interest: none declared.

REFERENCES