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Case report

Primary pulmonary valve sarcoma involving pulmonary artery and right ventricular outflow tract

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ARTICLE INFO

Article history:
Received 12 February 2015
Accepted 16 February 2015
Available online 11 March 2015

Keywords:
Pulmonary valve
Leiomyosarcoma
Pulmonary embolism
Heart surgery

ABSTRACT

The group of pulmonary valve and pulmonary artery primary tumors is the most frequently represented by schwannomas, myxomas, papillary fibroelastomas, primary choriocarcinomas and sarcomas. These tumors are very rare. The most common clinical manifestation of pulmonary artery tumor is dyspnea, followed by chest pain, cough, and haemoptysis.

A case of 44-year-old male with history of progressive dyspnea, fever, cough, and weight loss is presented. Imaging methods showed large saddle embolus in the right ventricle outflow tract, pulmonary valve and pulmonary artery trunk. He was admitted to a hospital for anticoagulation therapy. Since there was no clinical improvement, the patient had to undergo surgery. Nearly full artery caliber filling tumor in pulmonary artery was found with its origin in pulmonary valve. Next exploration showed several little tumors in right ventricle outflow tract and also in pulmonary artery. The final outcome of histological examination showed the presence of leiomyosarcoma high grade 3.

Presented case highlights that pulmonary artery tumor should always be included in the differential diagnosis of pulmonary embolism, especially if the symptoms progress while on adequate anticoagulation, and if any risk factor for deep vein thrombosis is not present. Unfortunately, prognosis of pulmonary artery sarcomas is usually dismal, secondary to late diagnosis.

Introduction

Primary tumors of the pulmonary artery and pulmonary valve are rare and may pose a difficult clinical diagnosis. The group of these tumors includes schwannomas, myxomas, papillary fibroelastomas, primary choriocarcinomas and sarcomas [1–4]. Pulmonary artery sarcomas (PASs) are the most common primary tumors of the pulmonary artery, but the incidence of this diagnosis is extremely rare. PAS was first described in autopsy by Mandelstamm in 1923 [5]. There have been reported around 200 cases of PASs in the literature up to date [6].

PASs is predominantly presented among patients from their third to seventh decade, with an average age of 49 years [7,8]. PASs are always highly malignant, women are involved twice as often as men [9]. The most common clinical

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http://dx.doi.org/10.1016/j.crvasa.2015.02.006

Table 1 – Incidence of PASs subtypes described by several authors [8,16,17].

Author:	Cox et al.	Nonomura et al.	Gaumann et al.
Year	1997	1988	2008
Number of patients	138	110	18
Subtype of PASs and incidence in groups			
Undifferentiated sarcoma	31%	31%	
Leiomyosarcoma	16%	21%	
Spindle-cell sarcoma	14%		
Malignant fibrous histiocytoma	7%		22%
Fibrosarcoma	5%	6.4%	
Rhabdomyosarcoma		6.4%	
Myxofibrosarcoma			44%
Epitheloid			22%

manifestation of PASs is dyspnea (72%) followed by chest pain (45%), cough (42%), and haemoptysis (24%) [6,10]. The presence of these symptoms often causes misdiagnosis of pulmonary embolism [7]. Systematic symptoms of PASs are less rare and they include weight loss (21%), syncope (9%), and fever (8%) [6]. PASs more frequently affect segments of the pulmonary artery with large diameter, predominantly the trunk (85%), right (71%) and left (65%) arteries. The pulmonary valve (32%) and right ventricular outflow tract (10%) are involved less frequently [6,11–14].

There are two major categories of PASs known: intimal and mural. The intimal PASs show an intraluminal polypoid growth pattern and histological evidence of fibroblastic and myofibroblastic differentiation. These subtypes are typically positive for vimentin and osteopontin and negative for endothelial markers. The mural PASs show characteristics related to the soft tissue sarcomas [8,15]. There have been many histopathological subtypes of PASs reported, including undifferentiated sarcomas, leiomyosarcomas, rhabdomyosarcomas, fibrosarcomas, chondrosarcomas and osteosarcomas [6]. Table 1 shows the incidence of PASs subtypes described by several authors (Table 1) [8,16,17].

Case report

A 44-year-old male with medical history of appendectomy and hepatitis was accepted to the emergency room with several week history of progressive dyspnea, fever, cough, and weight lost of 4 kg. He denied chest pain. Initial transthoracic echocardiography showed right ventricle dilatation and pulmonary hypertension. Contrast CT scan of chest was performed and revealed what appeared to be a large saddle embolus in the right ventricle outflow tract, pulmonary valve and pulmonary artery trunk (Figs. 1–4). According to these findings the patient was admitted to a hospital for anticoagulation therapy using low molecular weight heparin and warfarin. Since there was no clinical improvement recorded, the patient was transported to the cardiology institute.

Repeated transthoracic echocardiography was performed with suspicion of pulmonary valve and right ventricle outflow tract tumor. Ventilation/perfusion scan recorded moderate asymmetry with declining of signal in right lung without evident segmental defects. Coronarography did not reveal any coronary artery stenosis. The patient was indicated for cardiac surgery.

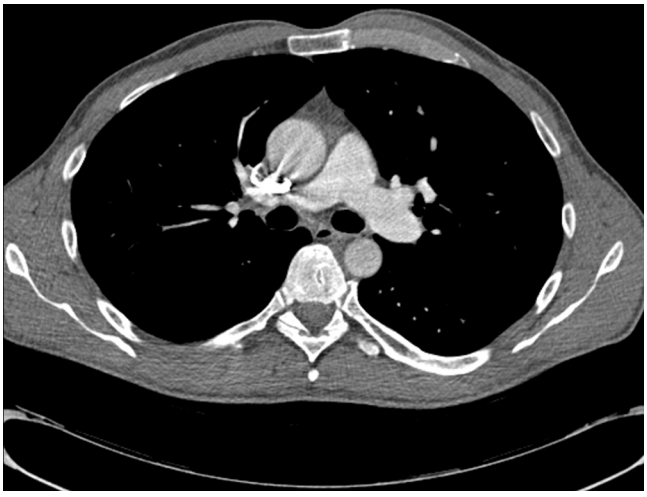

Fig. 1 – Computer tomography image (high of left pulmonary artery distance) – no sign of thrombus or tumor.

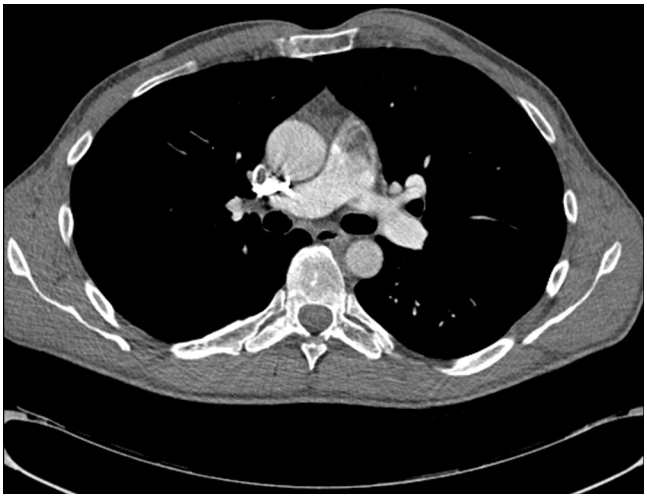

Fig. 2 – Image of computer tomography (high of right pulmonary artery distance) – no sign of thrombus or tumor in right pulmonary artery/defect of contrast filling in pulmonary artery trunk.