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Pulmonary Artery Intimal Sarcoma: Case Report and Revue of Literature

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Abstract **Case Report**

Pulmonary artery sarcoma is a rare tumor of the great vessels .It is aggressive and rapidly evolving disease, which is underdiagnosed and misunderstood. The diagnosis is usually made postoperatively or at autopsy a highly. Rapid recognition is needed in order to decrease the time between diagnosis and treatment. Radiological examinations are useful in the rapid diagnosis of pulmonary artery intimal sarcoma. Distinguishing this disease from thromboembolic events is imperative in order to hasten the initialization of surgical resection and chemotherapy.

Keywords: Pulmonary artery, sarcoma, intimal, diagnosis.

INTRODUCTION

Pulmonary artery intimal sarcoma is a rare disorder arising from the intimal wall of the pulmonary artery. As the tumor decreases the lumen of the pulmonary artery. Patients usually present with symptoms and signs of right ventricular failure, and it has been often misdiagnosed as pulmonary thromboembolism. It is important to differentiate between the two entities [10].

This tumor's evolvement is characterized by endoluminal growth with later vessel obstruction. This type of tumor can also originate from the heart valves and the right ventricular outflow tract. Peripheral thromboembolism is a common concomitant finding, often leading to the false diagnosis of chronic thromboembolism [7].

CASE REPORT

We present the case of a 35-year-old man and having as FDRCVx a chronic smoking and not having a notable background, presents for dyspnea and chest pain gradually evolving.

all evolving in a context of deterioration of the general condition. On physical examination, the patient exhibited tachypnea, with 24 breaths per minute, a regular pulse at 85 beats per minute and a blood pressure of 120/80 mmHg. Cardiac auscultation revealed a burst of B2 and a systolic murmur at the pulmonary focus with dorsal irradiation, there is also a systolic murmur of tricuspid insufficiency. Further physical examination was unremarkable.

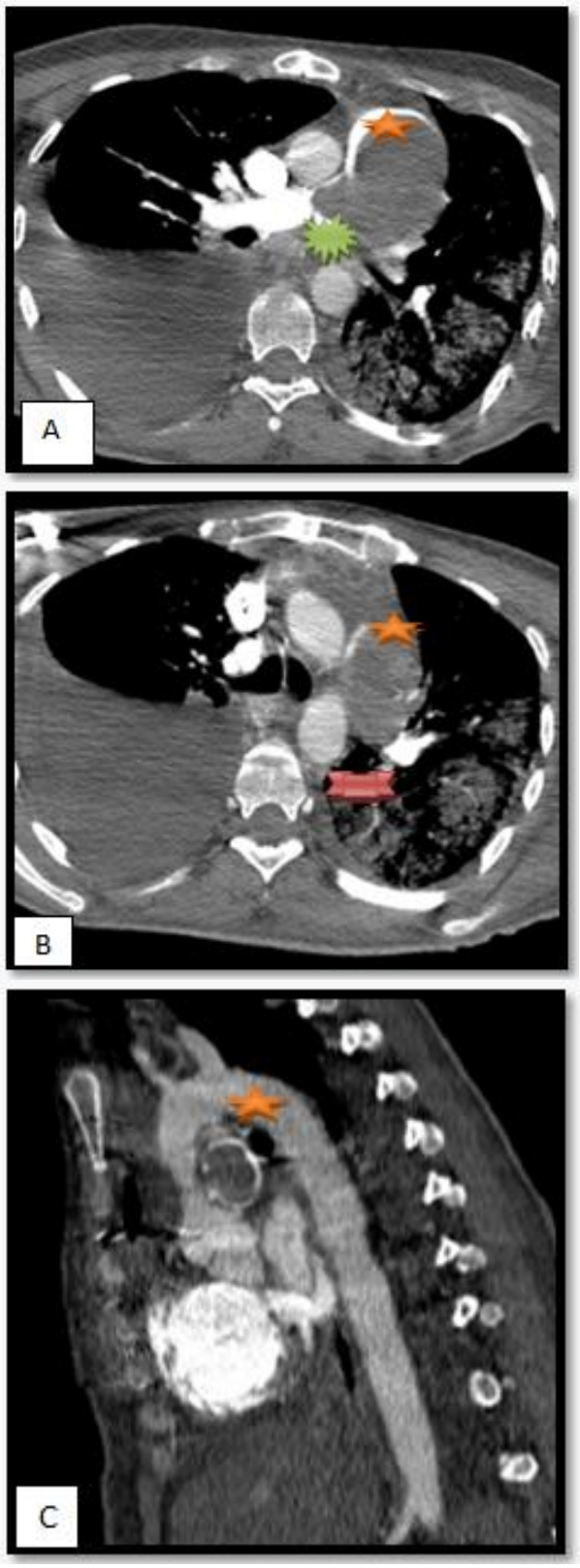
Resting ECG is registered in sinus rhythm with a PR space at 160ms and a QTc at 384ms, with no disturbance of repolarization in right precordial and absence of epsilon wave or IDM sequelae.

Blood count and chemistry profile were within normal range, including the level of brain natriuretic peptide. D-Dimers were at 0.5 (normal range\0.55 mg/l); CRP was at 1.1 (normal range\0.5 mg/dl).

Doppler echocardiography objective of a mass obstructive in the pulmonary artery with a gradient to 64mmhg with dilated and hypertrophied RV at systolic function preserved and a PAH supra systemic (PAPS at 140mmhg).Thoracic CT angiography shows dilation of the pulmonary artery measured to 48.3 mm in diameter, the site of a tissue like material hypodense with increased contrast, occupying almost all of the vascular lumen, extending to the origin of the left and right pulmonary artery (Fig.1) and budding at the level of the right ventricular. Dilation of the right atrium and the right ventricle with hepatic cavosus reflux. Cardiac MRI shows an intraluminal mass of the trunk of the pulmonary artery extending to the right bronchi and left. CT-TAP does not show any primary tumor or secondary lesions, and tumor markers (ACE, PSA, AFP, CA 19-9) are normal.

After consultation with the HEART TEAM, a pulmonary thromboendarterectomy for diagnostic purposes and therapy was carried out and whose study pathologic discovered intimal sarcoma of the pulmonary artery.

The patient was treated with chemotherapy



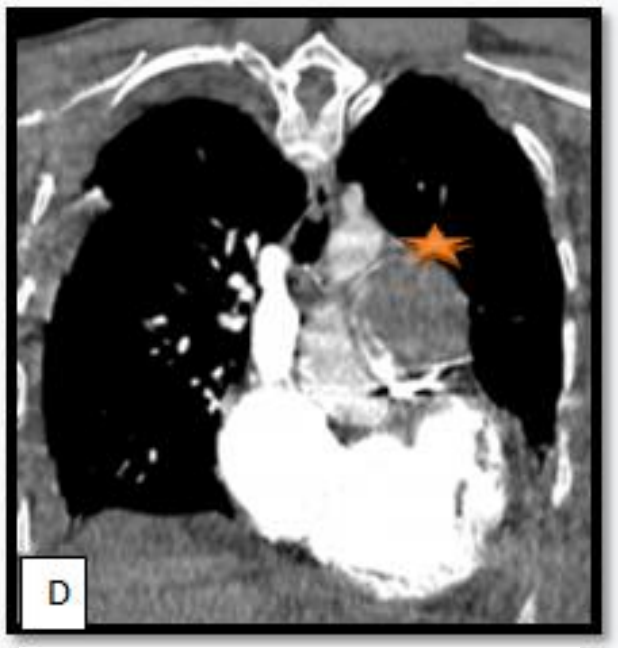


Fig-1: (A) and (B) axial, (C) sagital and (D) coronal Computed tomography images: pulmonary artery the site of a tissue like material hypodense, involving the pulmonary artery trunk , and extending to the origin of the left and right pulmonary artery.

DISCUSSION

Angiosarcomas (AS) are rare malignant tumors, representing less than 2% of soft tissue sarcomas [2]. Pulmonary artery sarcoma is an extremely rare pulmonary Malignancy [10], with an incidence of 0.001–0.03 % [3]. It was first described by Mandelstamm in 1923.

Before retaining their primary character, it is advisable to eliminate a primary extra-thoracic tumor which could have metastasized [2].

Sarcomas of the pulmonary artery are commonly divided into mural and intimal sarcomas. The first is usually the typical leiomyosarcoma that develops along the medial smooth muscle of the large veins involving mainly the vena cava. The second affects the intimal layer of the large blood vessels along the pulmonary artery and the aorta. Intimal sarcomas are usually twice more common in the pulmonary artery than in the aorta [3].

The risk factors for developing PAIS are poorly defined. There are case reports in the literature that have looked at the genetics of certain patients, and a hypothesis has been made that individuals who are homozygous for some sequence variants that are responsible for folate levels and DNA methylation can cause instability and maybe potentiate the development of PAIS [13].

The tumors arise in the pulmonary trunk, the left or right pulmonary artery and may rarely spread to the right ventricle and the pulmonary valve in a retrograde manner. The tumor may also arise from the right ventricular outflow tract and the heart valves [3].

The median age at diagnosis is 49 years. The sex-ratio is close to 1, as for the other locations of soft tissue sarcomas. Symptoms evolve insidiously for several months, on average ten. The most common symptoms of pulmonary artery sarcoma are insidious dyspnoea, which is a nonspecific symptom and might be associated with other aetiologies of right ventricular failure. Other symptoms in cases of pulmonary artery sarcoma include tightness in the chest, cough, haemoptysis, and constitutional symptoms such as fever, fatigue, and weight loss are the features of malignancy [11]. All this no specific symptoms at an early stage and has been easily misdiagnosed as pulmonary thromboembolism due to the resemblance in clinical features, such as dyspnea, pleuritic pain, cough and hemoptysis [8]. In 20% of cases, it is an autopsy discovery, in a completely asymptomatic subject [2].

The majority of patients were usually misdiagnosed with pulmonary vascular diseases such as chronic thromboembolic pulmonary hypertension (CTEPH) or acute pulmonary emboli [3]. From the viewpoint of imaging, the differential diagnosis between PAS and thrombo-embolic disease is difficult, even though the CT features may be specific in patients with advanced malignant disease [6]. The absence of predisposing factors for pulmonary embolism (no venous thrombosis of the lower limbs on color doppler ultrasound, no finding suggestive of hypercoagulable state) and the progressive worsening of dyspnoea in patients with an intraluminal filling defect despite anticoagulation therapy are factors suggestive of PAS. The lack of response to anticoagulation therapy correlated with the imaging findings may be decisive for the diagnosis [6].

Imaging features

The radiological features of pulmonary artery sarcoma are filling defect in the pulmonary artery lumen, extra luminal tumor invasion, and a nodular appearance. However, despite specific features, published studies have shown that more than half of the patients with pulmonary artery sarcoma are initially misdiagnosed as pulmonary thromboembolism.

Several studies have been conducted in an attempt to distinguish between the two entities [11].The diagnostic workup of ISPA relies mainly on radiological imaging with computed tomography - angiography of the chest and cardiac ultrasound. Magnetic resonance imaging and PET scan are complimentary in the management plan. Lung perfusion scintigraphy may be of help in ruling out an initial suspicion of lung thromboembolism [3].

Chest X-ray

The main role of chest X ray is to exclude other diseases that may present with similar symptoms and to provide elements to guide subsequent diagnostic investigations [6].

Computed Tomography Scan (CT Scan)

Contrast-enhanced CT scan of the chest is usually the first imaging technique performed in patients with respiratory symptoms. ISPA manifests radiologically in a heterogeneous low-density filling defect of the pulmonary vasculature with irregular distribution or a hyperdensity with a risk of vascular distention when the tumor occupies the entire lumen of the common trunk. The ISPA diagnosis is favored by an average increase of 25 housefield units (HU) in density after contrast enhancement. The contrast uptake is usually delayed during the venous phase after an initial iso-hypodense filling defects on enhanced images. This delay can be attributed to the presence necrosis, hemorrhage or foci of ossification, but these signs are not specific of ISPA [3]. However, foci of calcification can also be detected in cases of chronic embolism or thrombosis in situ, although they tend to occur on the artery wall and peripherally in these cases [6]. The dual-energy CT-based iodine (DECT) quantification was evaluated to discern ISPA from lung thromboembolism. The mean density did not differ between the two entities but the mean iodine-based HU and iodine concentrations were significantly different between both groups [3]. In addition to aiding in the diagnosis of ISPA, CT scan detects local spread of the tumors into the mediastinum and parenchyma. It may also evaluate the cardiac repercussions of ISPA through indirect radiologic signs of right ventricular pressure overload such as the dilation of the right chambers and the abnormal regurgitation of the contrast [4].

Cardiac ultrasound

Cardiac ultrasound is essential in the evaluation of patients diagnosed with ISPA to detect any dilation of the right ventricle or outflow tract obstruction. Transoesophageal echocardiography offer additional information concerning the ventricular and valvular function, the significance of the lesions and their impact on cardiac. Invasive evaluation with right heart catheterization and intravascular ultrasound may be helpful. The first can reveal the presence of intraluminal opacification defects, parenchymal perfusion defects as well as abrupt termination of arterial branches. The second can help in the evaluation of the pulmonic valve involvement and differentiation among mural thrombusor ISPA [3].

Magnetic resonance imaging (MRI)

The current role of MRI of the chest is not well established and is usually limited to evaluate the equivocal findings encountered with CT scans. Gadolinium-enhanced MRI helps in identifying intraluminal involvement of ISPA. A classical finding is

the presence of a grape-like appearance of the pulmonary artery that is the results of the intraluminal filling defects extending into peripheral arteries [1]. These tumors have higher levels of gadolinium enhancement compared to thrombi and this enhancement can correlate to the tumor differentiation and the myxoid matrix .The multiparametric MRI can also contribute to define ISPA, in fact, the presence of hyperintensity on fat-suppressed T-weighted imaging and diffusion-weighted imaging are all significantly in favor of ISPA [3]. MRI, despite a greater capacity for tissue characterisation, has some disadvantages compared to CT: these include a lower spatial resolution and a need for a longer breath-hold time, which is a problem if we consider that the majority of the patients examined for this condition are heavily dyspnoeic. The intensity of contrast enhancement after contrast injection is correlated with the degree of tumour differentiation and can be also used in post intervention follow-up and monitoring to evaluate residual or recurrent tumor [6].

PET-Scan

PET-CT was used to confirm the suspicion of PAS and to distinguish between PAS and pulmonary embolism on the basis of the increased radiopharmaceutical uptake of tumors. Combining CT and PET-CT therefore proved to be extremely useful in assessing patients with suspected PAS [6]. The role of PET-scan in the evaluation of treatment response is being Investigated with promising results [3].

Prognostic

Prognosis for intimal sarcomas is very low with the median survival time being 11 ± 3 months without resection and 36.5 ± 20.2 months with resection. Patients undergoing multimodal treatments showed a median survival of 24.7 ± 8.5 months [5]. The prognosis seems to depend on the location and vascular extension. Long-term survival is lower compared to other easier excisable high-grade sarcomas, such as in the extremities [14].

Treatment

Due to the presence of pulmonary arterial occlusion and the acute symptoms associated with this, surgical resection is usually the mainstay of therapy. Surgery for PAS can include pneumonectomy, lobectomy, PEA or tumor debulking with or without pulmonary artery reconstruction, and the choice of procedure is dependent on factors such as the tumor location and distal extension [9]. Surgery offers the best way to prolong survival and is successful only if complete resection of tumor is performed [12]. An analysis of combined series by Blackmon *et al*. suggested that patients who underwent an attempt at curative resection have longer OS compared to those who had incomplete resection (median OS of 36.5 vs11 months) [9]. Immediate post-operative mortality rate has been previously reported to be around 13-15%, and surgical resection was nearly never R0 [9]. Post-operative chemotherapy has been reported to be effective in some cases, but its role in the treatment of PA intimal sarcoma is still not clearly defined. The same is true for radiation therapy and postoperative anticoagulation therapy [12]. Due to the rarity of this disease, data for efficacy of various treatment methods are limited however; there are several options of chemotherapeutic agents that may be used in the treatment of intimal sarcomas. Targeting the angiogenic factors associated with the endothelial proliferation has been a point of great interest. The rapid progression of the tumor along with the multiple proteins associated with angiogenesis should dictate the types of chemotherapeutic agents used [5].

CONCLUSION

PAS is a highly aggressive and rapidly evolving disease, which is underdiagnosed and misunderstood. Early diagnosis with the help of integrated imaging remains today the main direction to be pursued in order to obtain improvements in prognosis. CT scan of the chest and cardiac ultrasound, magnetic resonance imaging and PET scan may help in limiting the differential diagnosis in equivocal cases. PET scan may have a role in evaluation response to treatment. Surgical resection remains the optimal treatment for patients with intimal sarcoma involving the pulmonary artery. Due to the extremely poor outcome, radiotherapy and chemotherapy appear to be methods to consider for inoperable patients.

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