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Pulmonary Artery Intimal Sarcoma with Prolonged Survival: A Case Report and Review

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Abstract **Case Report**

Pulmonary artery intimal sarcoma (PAIS) is an extremly rare and agressive tumor. The prognosis is very poor, with median overall survival between 11 and 18 months. We report a case of a 38 year old man who presented to our oncology department with diagnosis of PAIS confirmed by histological examination after surgical resection and bioprosthetic pulmonary valve replacement. Six months after adjuvant chemotherapy, the patient presented a recurrence of his disease that requires other lines of chemotherapy with prolonged survival.

Keywords: Pulmonary artery, intimal sarcoma, thromboembolic, chemotherapy.

INTRODUCTION

Pulmonary artery intimal sarcoma (PAIS) is an extremely rare malignant tumor that develops in the intima of the pulmonary artery, first described by Moritz Mandelstamm [1] in 1923. Since then, approximately 400 cases have been reported in the literature [2, 3].

The symptomatology usually includes chronic dyspnea and other features of a right ventricular failure. A definitive diagnosis can rarely be made on the basis of clinical signs alone. Further investigations, including echocardiography, computed tomography (CT), magnetic resonance imaging (MRI), and positron emission tomography (PET), are often required [4].Clinical and radiological findings usually mimic chronic pulmonary thromboembolic disease, resulting in diagnostic delays [5].

Surgical intervention of the primary tumor is the best therapeutic option to prolong survival (2, 3) which can be combined with adjuvant chemotherapy and radiotherapy.

And according to recent studies, multimodal treatment offers better survival outcomes than monotherapy such as surgery alone [4].

The prognosis is generally poor [2], and despite new treatment strategies in the future, patients with PAIS continue to have a poor prognosis [4].

We report the case of a patient followed for PAIS in the medical oncology department of Military Hospital MOHAMED V in Morocco, treated by surgery associated with adjuvant chemotherapy, while trying to clarify the diagnostic and therapeutic difficulties posed by this rare pathology.

CASE REPORT

A 38 years old man, with no cardiovascular risk factors , no past medical or surgical history and any history of tobacco use or other recreational drugs. He was initially admitted to the cardiovascular surgery department for two episodes of syncope during exercise with spontaneous and complete recovery of consciousness, preceded by prodromal symptoms such as dyspnea, chest pain and hemoptysis. Moreover, the patient stated a weight loss and fatigue.

On physical examination, heart auscultation revealed a B2 burst and a systolic murmur with dorsal radiation. A systolic murmur of tricuspid insufficiency was also noted. His lung was clear to auscultation with no stridor, wheezes, rhonchi, or rales. The patient also

did not exhibit any palpable supraclavicular, cervical, axillary, or inguinal adenopathy. Laboratory testing, including complete blood count and basic metabolic panel, did not reveal any abnormalities.

Transthoracic cardiac ultrasound showed two cardiac masses obstructing the pulmonary arteries with signs of pulmonary arterial hypertension, as well as right ventricular hypertrophy (Figures 1, 2, 3).

Cardiac MRI demonstrates an intraluminal pulmonary artery mass that extends to the right and left pulmonary artery branches without transparietal infiltration**.**

The patient underwent surgical resection of the pulmonary artery tumor consisting of thromboendarterectomy, associated with replacement of the pulmonary valve by biological prosthesis. The resection was incomplete.

Anatomopathological examination with immunohistochemical analysis of the surgical specimen revealed a strong positivity of Mouse Double Minute 2 (MDM2) and focal positivity of CD34 (Figure 4). Pulmonary artery intimal sarcoma was diagnosed after team consultation within the tumour board.

The evaluation 1 month after surgery found a conscious, apyretic, and eupneic patient, heart rate was 81 bpm and blood pressure was 125/75 mmHg. The heart auscultation was normal. The rest of physical examination was unremarkable. Electrocardiogram revealed regular sinus rhythm at 74 bpm, with incomplete right bundle branch block. Chest X-ray demonstrate no cardiomegaly with a cardiothoracic index of 0.48 and good pleuroparenchymal transparency.

Based on these findings, the patient was referred to medical oncology department to discuss adjuvant treatment. As part of the extended assesment, a thoracic-abdominal-pelvic CT scan was performed (figure 5), and showed no signs of recurrence with the presence of a left subpleural nodule measuring 7x10 mm surrounded by ground glass, as well as a thoracic angiography scan, which showed a thrombosis at the level of the pulmonary valve, with a trunk of the pulmonary artery and a proximal stenosis of the origin of the right branch of the pulmonary artery with respect of its permeability, for which he was put under anti-coagulant treatment at curative dose.

The multidisciplinary staff decided adjuvant chemotherapy for our patient. A port-a-cath was inserted in preparation for chemotherapy; then he received six cycles of adjuvant combination chemotherapy based on Adriamycin 60mg/m2/day and Ifosfamide with mesna at 5g/m2/day (AI protocol). The regimen repeated every 21 days. The inter cure was characterized by the absence of clinical side effects, and the absence of biological abnormalities or any lesions of progressive appearance on the thoracoabdomino-pelvic CT scan 1 month after the last cycle (figure 6).

The first control after 3 months was normal. The second evaluation after six months finds a patient who has shortness of breath upon exertion. Computed tomography (CT) was performed, which revealed tumor recurrence (figure 7).

PET scan showed the rather intense pathological hypermetabolism described at the level of the right pulmonary artery prosthesis and hyerfixation of the pulmonary parenchymal nodule in the dorsal segment of the right upper lobe and visualization of intense hypermetabolism in the right and left ventricular walls (SUVmax = 11.2), which should be compared with the data from the cardiological investigations (Figure 8).

Paclitaxel was then chosen for the first palliative chemotherapy (80 mg/m2 on days 1, 8, and 15 of a 28 day cycle). Following three cycles of treatment, a CT scan was performed, which showed a stable aspect of the pulmonary artery lesion process, compared to the last CT scan. After six cycles totally, the patient underwent a thoraco-abdomino-pelvic CT scan, which showed a 40% increase in the size of the intra-cardiac process (figure 9) and increasing of pulmonary nodules, in favor of progression. The patient was put on Endoxan orally, and he is still following this treatment after 3 cycles.

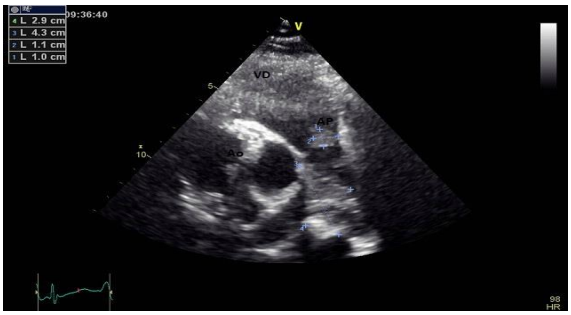


Fig-1: Para-sternal trans-aortic short-axis slice showing in two dimensions two cardiac masses (angiosarcoma) measuring respectively 10 x 11 mm and 43 x 29 mm *RV: right ventricle, Ao: aorta, PA: pulmonary artery*

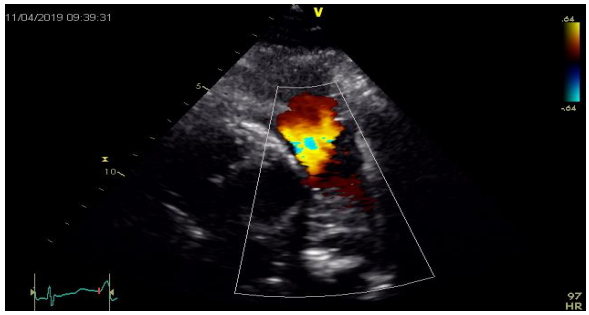


Fig-2: Para-sternal trans-aortic short-axis slice showing in color doppler an acceleration of the Pulmonary flow showing arterial obstruction

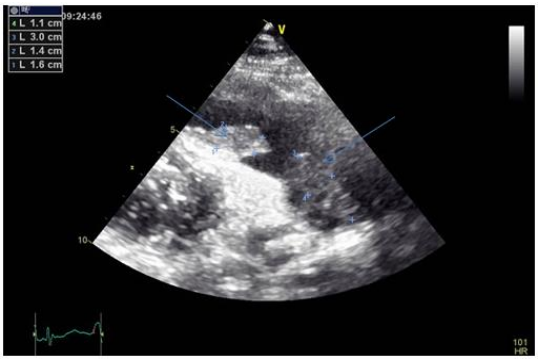
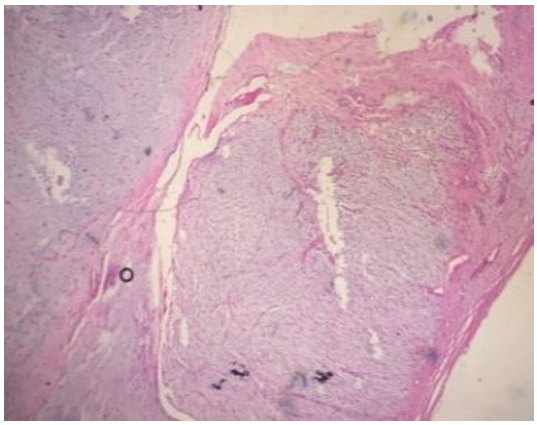
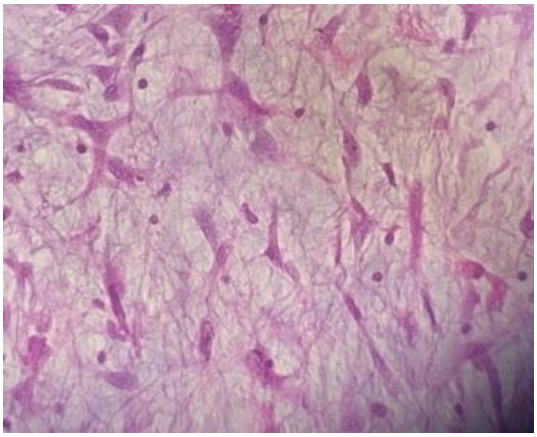


Fig-3: Section focused on the pulmonary infundibulum to better identify the 2 tumor masses (arrows)



A



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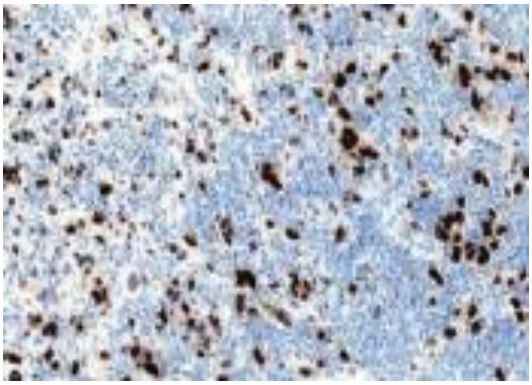

C

Fig-4: A: Histological image staining at low magnification Gx10 showing intraluminal tumor proliferation with a double cellular and hypocellular component

B: histological image with high magnification G x 40 showing spindle cells with marked cytonuclear atypia on a myxoid background. C: Immunohistochemical staining of the neoplastic cells shows focal reactivity to MDM2.

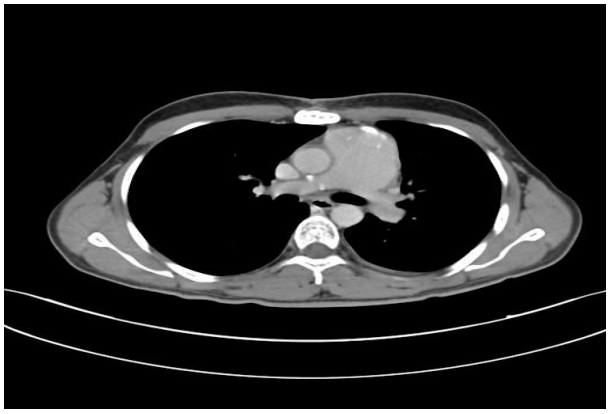

Fig-5: Immediate postoperative image of the pulmonary artery trunk dilated to 41 mm with absence of parietal or intra-luminal tumor syndrome

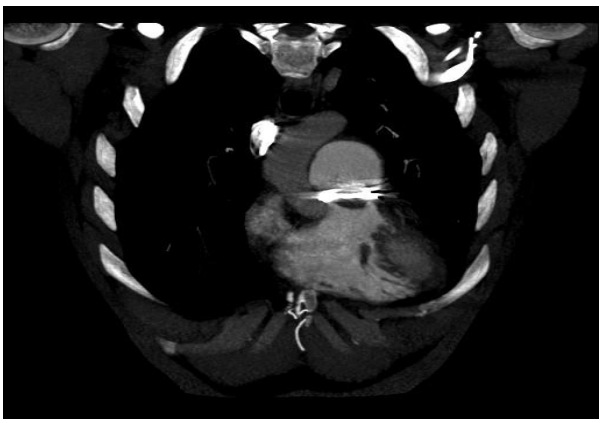

Fig-6: Coronal section showing the prosthesis in projection of the pulmonary valve with no signs of recurrence

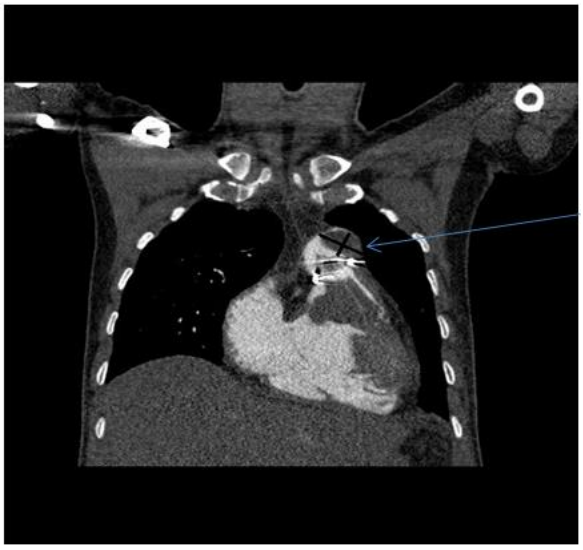

Fig-7: Coronal section showing a tumor recurrence in the lateral wall of the trunk of the pulmonary artery protruding into its lumen measuring 30x20 mm

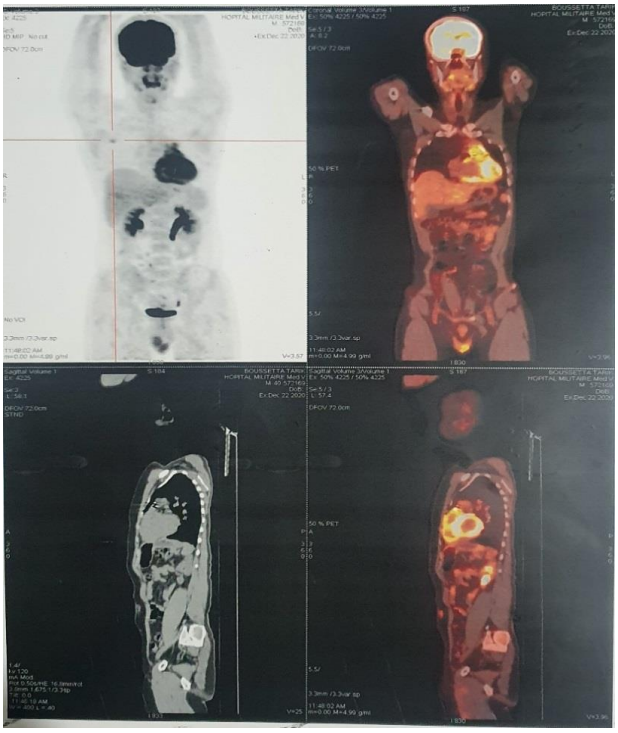

Fig-8: PET scan showing hyperfixation at the right pulmonary artery prosthesis with tumor progression

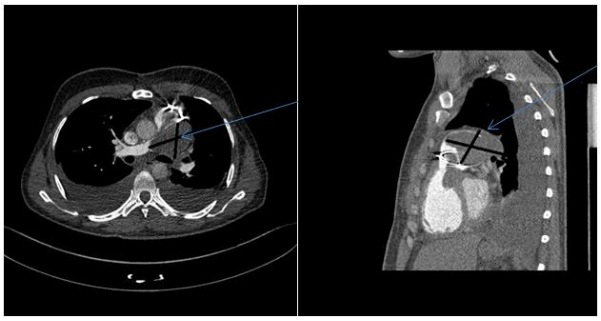

Fig-9: CT scan showing increase of cardiac mass (see arrow) A: transveral section B: sagittal section

DISCUSSION

Pulmonary artery intimal sarcoma is an extremely rare type of malignant tumor, which mimics pulmonary thromboembolism [6]. The incidence of this tumor is 0.001–0.03% [3]. The reported mean age is 50 years [4]. Pulmonary artery sarcoma arises from either the intimal wall or intramural wall of the pulmonary artery and can be accordingly classified into intimal and intramural sarcomas [4]. Due to its low incidence, the pathology and treatment strategies of pulmonary artery intimal sarcoma remain to be elucidated [7, 8].

PIAS are occupying the lumen of pulmonary artery and caused right ventricular dysfunction. The most common clinical sign of pulmonary artery sarcoma is dyspnoea, which is a nonspecific symptom of this pathology. Other symptoms in cases of PAIS include cough, tightness in the chest, haemoptysis, fatigue and weight loss in advanced cases [4]. From published studies, other modalities have been used for diagnosis: transthoracic echocardiography, CT, magnetic resonance imaging (MRI), and positron emission tomography (PET) [4].

Multimodal treatment including complete surgical resection with pulmonary endartrectomy followed by chemotherapy and/or radiotherapy remains the mainstay of treatment [9-11] and it provides the best survival benefit [4, 12].

There are efficacious chemotherapeutic regimens in patients with unresectable or metastatic pulmonary artery intimal sarcoma. Generally, doxorubicin-based regimens are used for the treatment of soft tissue sarcomas [13].

Protocols of chemotherapy included in literature were: adriamycin plus isosfamide, gemcitabine, or dacarbazine [2]. Paclitaxel has demonstrated to be effective for soft tissue sarcomas [6]. In addition, there have been some case reports showing good response of PAIS to vinorelbine based regimens [14, 15].

For the targeted therapy, a multi-targeted receptor tyrosine kinase inhibitor, pazopanib, which was approved by the Food and Drug Administration (FDA), has been reported to be useful in treating pulmonary artery sarcoma. Kollar *et al.* [16] and Funatsu *et al.* [17] described a partial response of pulmonary artery sarcoma with pazopanib. Further studies with the use of other chemotherapy drugs and targeted therapies are needed to try to improve the prognosis of unresectable or metastatic PAIS.

Despite surgical resection and chemotherapy, the prognosis for pulmonary artery sarcoma remains very poor with the median survival time being 11 ± 3 months without resection and 36.5 ± 20.2 months with resection [2, 18].

In Wong study [10], we analyzed 20 patients diagnosed with PAIS and he's obtained a median overall survival of 17 months: patients who received multimodal treatment including surgery chemotherapy and radiotherapy showed better survival compared to those who had surgery alone (24 months vs 8 months).

Our patient is still in good control, 22 months after diagnosis which concording with prolonged survival case reports and series in literature.

CONCLUSION

Pulmonary artery intimal sarcoma is aggressive tumor and rapid diagnosis is essential to improve prognosis. Physicians must distingue this disease from thromboembolic events. Chemotherapy is one of the treatment options for patients with advanced and unresectable pulmonary artery intimal sarcoma and prolongs survival.

Disclosure Statement

The author of this case report declares no conflicts of interest.

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