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Pulmonary Involvement in TAKAYASU Disease: From Medical Treatment to Surgery? A Case Report and Review of the Literature

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Abstract **Case Report**

Background: Takayasu disease is a nonspecific inflammatory arteritis occurring in young people and affecting large arteries. Pulmonary involvement is well documented during the course of the disease but may pose a problem of diagnostic delay. ***Case Description:*** We report a case of pulmonary artery involvement following exertional dyspnea in a context of fever with arthralgia revealing Takayasu disease and we will detail the different cardiovascular and pulmonary manifestations of this disease. ***Conclusion*:** Takayasu arteritis (TA) is a rare disease that commonly affects women in their second or third decade. The manifestations are highly polymorphic, ranging from asymptomatic presentations to catastrophic neurological presentations. Pulmonary involvement may be a challenge for diagnosis.

Keywords: Takayasu arteritis, pulmonary involment, management.

INTRODUCTION

Takayasu's disease is a chronic inflammatory arteritis of unknown etiology, preferentially affecting young women, with segmental involvement of the aorta and its main branches, the pulmonary arteries [1]. Thickening of the vascular wall is the most characteristic early sign of the disease, progressively leading to stenosis, thrombosis and sometimes aneurysm development [1, 2]. We report a case of pulmonary artery involvement following exertional dyspnea in a context of fever with arthralgia revealing Takayasu disease and we will detail the different cardiovascular and pulmonary manifestations of this disease.

CASE DESCRIPTION

Clinical and para clinical findings:

A 55-year-old man, without modifiable cardiovascular risk factors, without any particular history, presented 5 years ago with a vesperal fever associated with joint pain, prompting an internal medicine consultation where explorations were performed suggesting a systemic disease and put on corticosteroid therapy with a good evolution. Two years later, the evolution was marked by the appearance of an effort dyspnea of stage NYHA II without thoracic pain.

A thoracic CT angiogram showed parietal thickening of the pulmonary artery trunk (PAT) as well as of the right and left pulmonary arteries (R/L PA) with a predominantly right-sided tight stenosis at the origin of the right pulmonary artery trunk without affecting the right cavities but associated with dilatation of the ascending aorta: a dilatation of the right pulmonary artery (RPA) was performed but without success

As the symptoms persisted under medical treatment, a right catheterization was performed, finding severe stenosis of the proximal segment of the right pulmonary artery with a RPA-PAP gradient of 55mmHg and moderate stenosis of the left pulmonary artery (LPA) with a LPA-PAP gradient of 28mmHg and an elevation of the right ventricular systolic pressure (RVSP) of 75mmHg. A surgical indication was retained but postponed because of the inflammatory context. The patient was put on corticosteroids and immunosuppressive treatment.

Two (2) years later, we admitted the patient for a worsening of his dyspnea from stage II to stage III NYHA, associated with a dry cough and atypical precordialgia on effort, so we performed a right

catheterization finding a worsening of the pulmonary artery stenoses and a worsening of the pressures.

The clinical examination on admission notes a conscious patient, who supports the supine position, his conjunctiva are normally colored, a tachycardia at 104bpm with a BP of 113/70 mmHg, The cardiac examination finds regular heart sounds, with a 3/6th systolic murmur at the pulmonary focus and a diastolic murmur at the aortic focus without peripheral signs of right heart failure. Peripheral pulses were present bilaterally and symmetrically with no murmur on auscultation of the major accessible vascular axes. The pleuropulmonary examination and the rest of the somatic examination are without abnormality. The resting electrocardiogram showed an RRS at 104 c/min, a right axial deviation with micro voltage in the standard leads (Figure 1). The biological workup shows a biological inflammatory syndrome (WBC: 10180, CRP: 26 mg). Resting transthoracic echocardiography shows stenosis of both pulmonary arteries at their proximal segments with a Gmoy of 65mmHg, high intra-VD systolic pressure, non-dilated right cavities, moderate functional tricuspid insufficiency (TI), mild IAo with dilatation of the sinuses of Valsalva, stricture of the aortic isthmus without stenosis. A non-dilated, non-hypertrophied wall LV with good left ventricular systolic function (EF estimated at 61% by Simpson biplane).

Thoracic angio-CT reveals ostial stenosis of the bilateral pulmonary arteries with significant calcifications of the ascending and descending aorta (Figure 2). Right catheterization reveals worsening pulmonary artery stenosis (RV-RAP gradient to 100mmHg, RV-LPA gradient to 80mmHg)) and deteriorating pressures (PSRV to 124mmHg) (Figure 3). Coronary angiography revealed angiographically healthy arteries, and ultrasonography of the supra-aortic trunks and lower extremities was unremarkable except for moderate calcification of the walls of the superficial femoral arteries and the right and left tibio-peroneal trunks.

MANAGEMENT AND RESULTS

In view of the worsening of the symptomatology, the worsening of the pulmonary artery stenoses and the deterioration of the pressures on right catheterization with repercussions on the right cavities, the patient was proposed for surgical treatment. He underwent pericardial patch plasty of the pulmonary artery trunk (TAP) and tube anastomosis of the right and left pulmonary artery to the pulmonary artery trunk with good postoperative results. Pathological examination of the surgical excision biopsy revealed an appearance of subacute granulomatous arteritis consistent with Takayasu's arteritis disease.

The patient was discharged with the following medical treatment: VKA (Sintrom® 4 mg/day) for 3 months then aspirin (Kardégic® 75 mg/day), and prednisone 7.5mg, Calcifix D3 1cp / day, Omeprazole 20 mg/day then referred to the internal medicine department for follow-up.

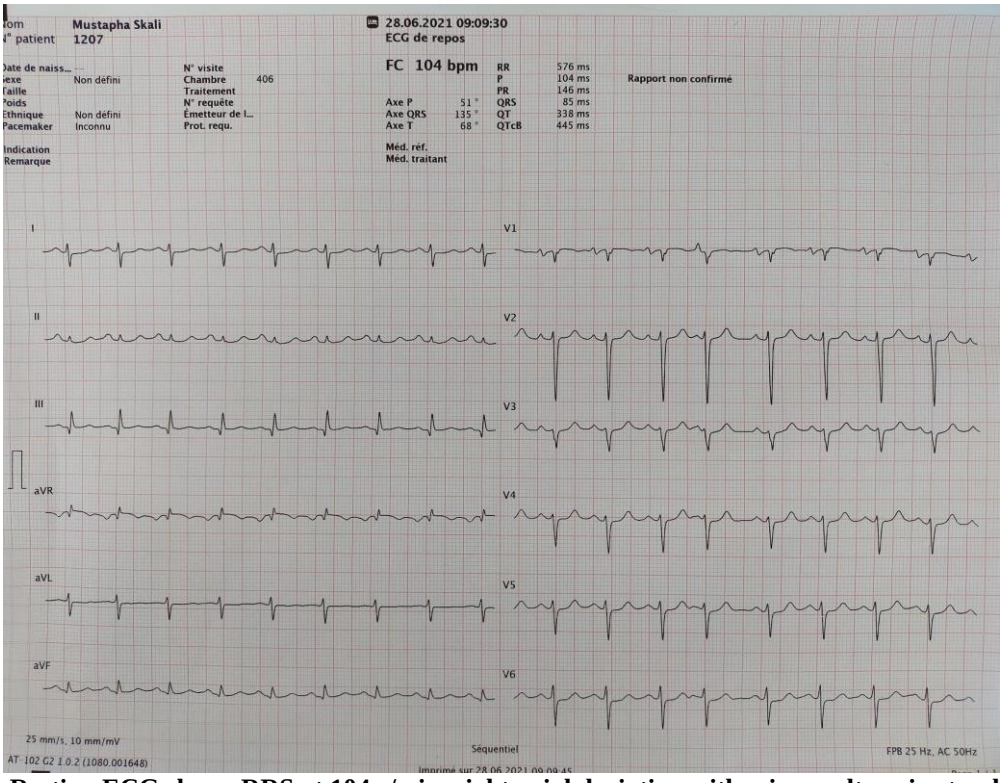

Figure 1: Resting ECG shows RRS at 104 c/min, right axial deviation with micro voltage in standard leads

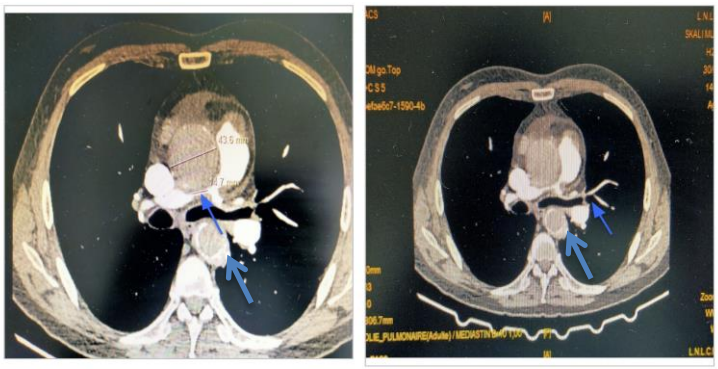

Figure 2: Image on left and Right: Thoracic angio scan reveals ostial stenosis of the bilateral pulmonary arteries with significant calcifications of the ascending and descending aorta

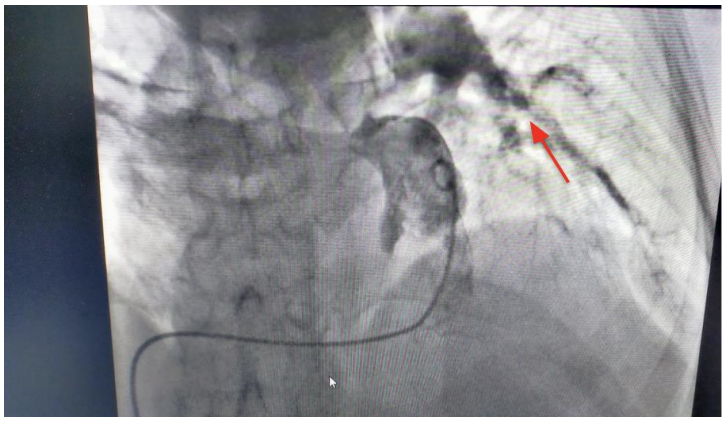

Figure 3: Right catheterization reveals worsening pulmonary artery stenoses

DISCUSSION

Pulmonary artery involvement in Takayasu disease (TD) was first described in 1940 and was included in the Ishikawa minor diagnostic criteria for the disease. Its early discovery relies on imaging because it is often asymptomatic or marked by frustrating and nonspecific clinical signs: cough, hemoptysis, pulmonary embolism picture [3].

The appearance of these symptoms in a patient with Takayasu disease should raise suspicion of underlying pulmonary arteritis and make it necessary to systematically search for it by thoracic angioscan or ventilation-perfusion scintigraphy. This condition was initially considered a rare manifestation of the disease [4]. Its frequency varies in published series depending on the diagnostic means used, from 20 to 56% in post-mortem series [5, 6] and from 14 to 70% in angiographic series [1]. This large variability can be explained by the difficulty of detecting an early parietal attack with this technique.

In contrast, ventilation-perfusion scintigraphy detects this involvement in 60 to 76% of cases [1, 5].

Isolated pulmonary arteritis at the time of diagnosis of Takayasu disease (TD) is rare. It is more often associated with involvement of the aorta and its collaterals [7, 8]. However, Toledano et al. reported in their review of the literature that its frequency is 31.8% [6].

The diagnosis of pulmonary artery involvement in Takayasu disease (TD) relies on several imaging techniques. Conventional angiography has long been considered the gold standard. However, this diagnostic modality often faces practical difficulties in the face of stenoses of the aorta or other peripheral vessels that are common in patients with Takayasu disease. It is also an invasive technique that has the disadvantage of not recognizing parietal thickening, which may be the only sign of the disease at an initial stage [9, 10].

Cross-sectional imaging, notably angioscanner or cervico-thoraco-abdomino-pelvic MRI, is an efficient and non-invasive diagnostic tool. Angioscanner, given its availability and speed of acquisition compared to MRI, is an interesting alternative to pulmonary angiography. It allows the study of both thoracic and abdominal vessels. Involvement can range from the trunk of the pulmonary arteries to the peripheral branches with a predilection for segmental and subsegmental arteries and a more frequent localization in the upper lobes [11]. The lesions are more frequently straight [6, 11] as was the case in our patient at the beginning of the disease. The quality of life is severely impaired in Takayasu disease, while the prognosis is

generally good. The main causes of death are heart failure, stroke, renal failure and persistent inflammatory syndrome. Corticosteroid therapy is the first-line treatment, in case of failure, the addition of methotrexate would allow to control the disease.

Revascularization procedures are sometimes necessary, but the progressive evolution of the stenosis means that an effective collateral circulation is often established. There is no need to intervene on radiological images of stenosis, but it may be necessary if there is downstream repercussions, a severe clinical expression. For both angioplasty and surgery, revascularization procedures should not be performed in the acute inflammatory phase because of the very high risk of restenosis. This was the case in our patient.

CONCLUSION

Takayasu disease (TD) is a rare disease frequently affecting women in their second or third decade. The manifestations are very polymorphic, leading to asymptomatic presentations as well as catastrophic neurological pictures. Pulmonary involvement is rarely revealing but should be systematically investigated by a ventilation-perfusion pulmonary scan and an angioscanner. The prognosis is essentially related to the existence of complications (retinopathy, hypertension, aneurysm, aortic insufficiency, PAH) and to the initial evolution of the disease. The majority of patients respond to corticosteroid treatment. In case of failure, methotrexate would be the second-line treatment of choice.

Revascularization procedures are sometimes necessary if there is a downstream impact and a severe clinical expression.

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Abbreviations:
TD: Takayasu's Disease
TA: Takayasu Arteritis
APT: Artery Pulmonary Trunk
RPA: Right Pulmonary Artery
LPA: Left Pulmonary Artery
PAP: Proximal Artery Pulmonary
PSRV: Systolic Pressure of Right Ventricular
RV: Right Ventricular
LV: Left Ventricular
TI: Tricuspid Insufficiency
EF: Ejection Fraction

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