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Case Report

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PULMONARY HYPERTENSION SECONDARY TO PULMONARY ARTERY COMPRESSION BY A PSEUDO ANERYSM OF THE AORTIC ARCH REVEALING BEHCET DISEASE

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ABSTRACT

Pulmonary artery compression is a rare complication of the ascending aorta aneurysm which results in pulmonary hypertension and later on to right ventricle failure. All causes and all sites of the ascending aorte aneurysms can cause compression. The general outcome is poor. It depends on the underlying cause but also the timing of the repair. We report the case of a young patient who presents a compression of the pulmonary artery by an aneurysm of the aortic arch whose etiological investigation concluded to a Behcet's disease.

KEYWORDS: Pulmonary artery compression, Behcet disease, ascending aorta aneurysm.

INTRODUCTION

Extrinsic compression of the pulmonary artery by adjacent structures is a well known complication in oncology practice but much less so in cardiology as it is rarely due to an abnormality of the cardiovascular structures. Compression of the pulmonary artery by an aneurysmal formation of the aorta implies a significant dilatation of the ascending aorta which origin was dominated in the early 20th century by syphilitic aortic but nowadays by inflammatory and genetic diseases.

We report the case of a young patient who presents a compression of the pulmonary artery by an aneurysm of the aortic arch whose etiological investigation concluded to a Behcet's disease.

CASE REPORT

A young 40 years old male was referred to our cardiology department for an exertion dyspnea evolving for 4 months with a heart murmur at the physical examination. The patient's medical history was unremarkable and the patient took no medication. The cardiac examination found a regular pulse at 78 beat per minute, symmetric blood pressure in both arms at 110/80mmHg. The point of maximal impulse was in the fifth intercostal space at the midclavicular line. Auscultation found an ejection systolic murmur loudest in the pulmonary region radiating towards the upper parts of the chest and the carotid arteries with a preserved A2 and loud pulmonary P2. No signs of right or left volume overload were noted.).

ECG showed a regular sinus rhythm, unremarkable for rhythm, conduction and repolarisation abnormalities.

Tran thoracic ultrasound concluded to a high probability of pulmonary hypertension with Systolic pulmonary pression of 53mmHg measured on a tricuspid regurgitation (Fig. 1). The ventricles were morphologically and functionally normal, No significant valvular abnormalities was noted, left and right atrias were of normal size and no pericardial effusion was noted.

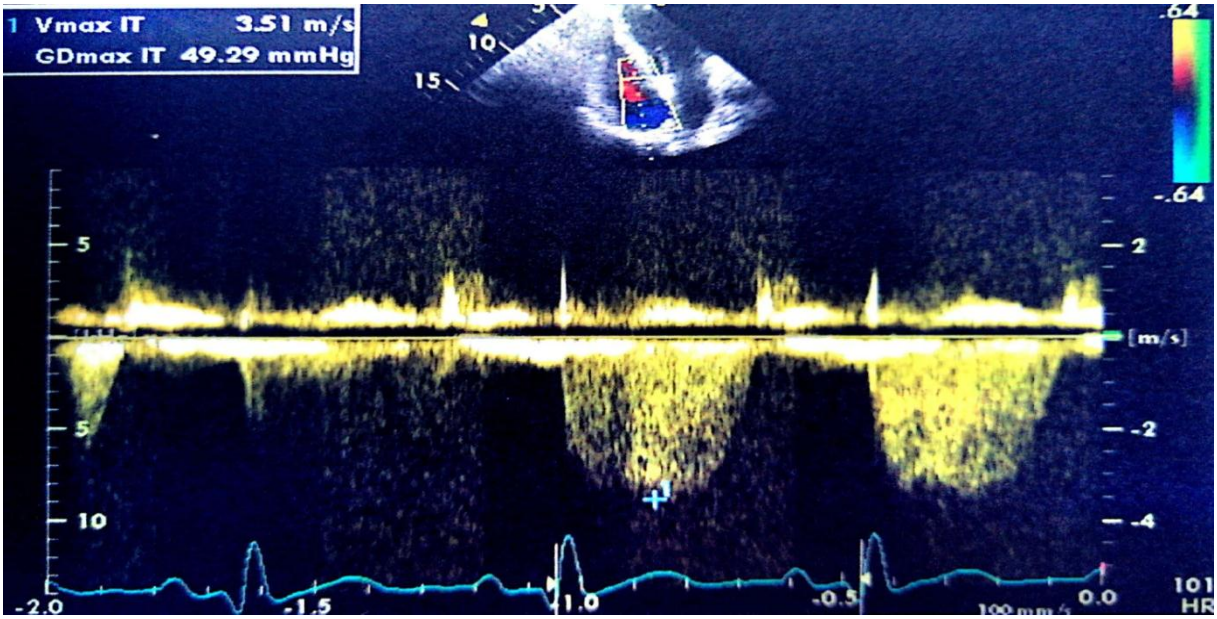

Fig. 1: Trans thoracic echocardiography showing CW Doppler envelope of the tricuspid regurgitation demonstrating increased pulmonary artery systolic pression. Tricuspid regurgitation maximal velocity at 3.51m/s which correspond to a trans-valvular gradient of 49mmHg.

CT chest scan completed by CT angiography showed a 10 cm thrombosed but still circulating pseudoanerysm thrombosed, fed by the aortic arch through a 2 mm orifice which caused the compression of the left pulmonary artery which remained permeable (Fig. 2).

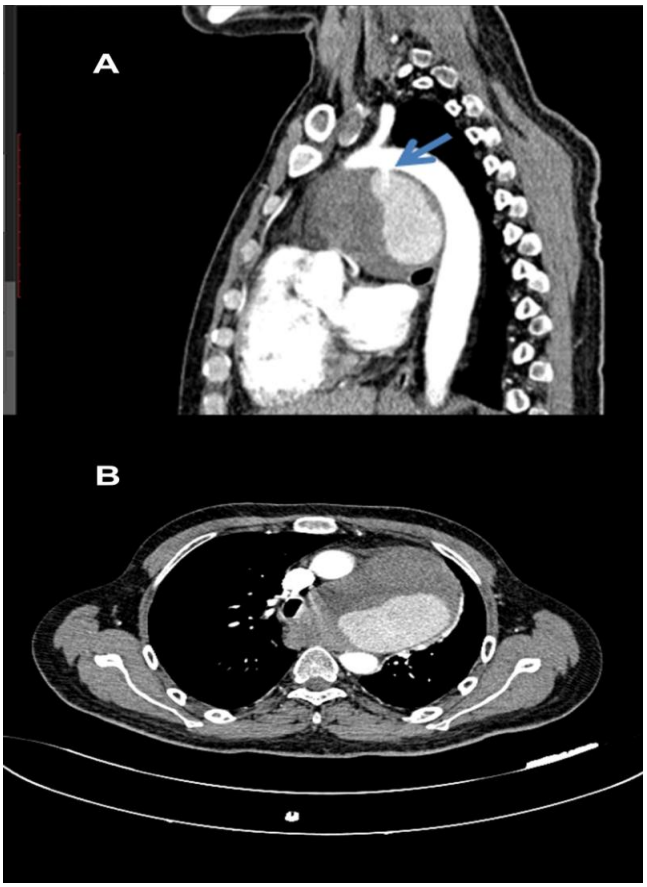

Fig. 2: A: CT section in injected sagital reconstruction (angio-thoracic protocol) showing a large circulating pseudoaneurysm thrombosed on more than 50%, fed by the aortic arch through a 2 mm orifice (Arrow). B: Injected axial section (angio-thoracic protocol) showing the mass effect of the pseudoaneurysm which pushes back the left bronchus stem and the left pulmonary artery which remains permeable.

Abdominal and lower limb CT angiography showed 2 small pseudoanerysms of the abdominal aorta and a femoropopliteal aneurysm. Cranial CT angiography was normal.

Abnormal aboratory test found a normochromic normocytic anemia with hb:11.5g/dl, Leucocytes:11.24 10^3/µl Neutrophile : 10.36 10^3/µl, Lymphocytes : 0.67 10^3/µl. CRP = 35mg/l. Coagulation workup, Electrolytes, renal, hepatic and thyroid functions and urine tests were all within normal.

After those findings, the medical history and examination were resumed which revealed a history of recurrent oral and genital sores and the presence of scrotal sores scars. Ophthalmic examination was normal. The diagnosis of Behcet disease was retained.

Faced with this active form of the disease, the patient was put first on a medical treatment combining corticsteroids and cyclophosphamide. Vascular surgery was scheduled at the end of the medical treatment.

DISCUSSION

Pulmonary artery compression by an aneurysm of the ascending aortic has been reported in the medical literature.[1-3] The cause of the aneurysm may be genetic, atherosclerotic or acquired. Most of the Compressing aneurysm we found in medical literature were acquired and of syphilic origin. Most often, as in our case, the compression affects the right pulmonary artery. It is chronic related to an increasing aneurysm. Some cases of acute compression in the setting of an aortic rupture or dissection have also been reported.[1] The compression may results from aneurysms originating from all parts of the ascending aorta from the sinuses of valsalva to the descending portion of the aortic arch. They may be saccular or diffuse in various sizes. An aneurysm rising from the concave portion of the aortic arch tends to be large and develop anteriorly and to the left to cause compression. Aneurysms rising from the initial parts of the aorta tend to be saccular and may compress adjacent Cardio-vascular structures (right or left atrium, right ventricle, coronary arteries, superior vena cava).[4,5]

The compression occurs progressively and results in no or little symptoms till the obstruction reaches a critical point causing pulmonary hypertension and a pressure overload for the right ventricle leading on the long term to right heart failure.

Dyspnea and chest pain are the most common revealing signs. Dyspnea reflects the beginning of a right heart failure while the chest pain is the result of the stretching of the aortic wall nerve fibers and may precede a rupture. The presence of a systolic murmur at the pulmonic area is common and must alert the physician towards an intrinsic or extrinsic compression of the pulmonary artery. Cyanosis, high venous pression, edema, hepatomegaly and Harzer's sign may appear in advanced forms.

Electric signs of right ventricle overload (Right axis, right bundle branch block and right ventricular hypertrophy) may be present at an early stage and chest x ray may show an enlargement of the mediastinum. Transthoracic echocardiography is not very effective in the diagnosis of aneurysms of the ascending aorta outside of its initial portion, but it may show indirect signs suggesting a pulmonary arterial origin. it finds an increase in pulmonary pressure with its repercussion on the right heart and in some cases it may show an acceleration of pulmonary flow due to the reduction in pulmonary caliber. CT angiography is necessary to the confirmation of the aneurysm and the compression.

In Behcet's disease, arterial involvement is observed in 5 to 10% of cases. This frequency is likely to be underestimated if we consider autopsy data in which arterial damage was observed in 1 in 3 patients.[6]

The prognosis for arterial disease remains severe since it is the main cause of death (approximately 30-40%) and is often recurrent.[7]

These can be arterial occlusions or aneurysms which are true arterial sores with a risk of rupture. Aneurysms are often multiple, preferentially involving the pulmonary arteries and the abdominal aorta, but all arteries can be affected.

It is now clearly established that immunosuppressive therapy is the cornerstone of the therapeutic strategy in these severe forms. It is based on the fact that inflammation of the vascular wall most likely plays a major role in the occurrence of vascular damage.

This treatment is based on high-dose glucsteroids combined with immunosuppression by intravenous cyclophosphamide or by anti-TNFα antibodies such as infliximab. Oral azathioprine or antibodies to TNFα are recommended in the management of recurrent vascular disease.

However, surgical or endovascular management should not be delayed if the patient is symptomatic. Corticosteroid therapy and immunosuppressive therapy should be instituted prior to surgical management to reduce the risk of post-operative complications. Postoperative complications are significantly less frequent in patients receiving immunosuppressive drugs. Multidisciplinary care in an expert centre is essential to decide on the optimal time for the operation. Long-term follow-up is essential to detect complications and recurrences.

CONCLUSION

Compression of the pulmonary artery remains an infrequent but very serious complication. It is important

for the cardiologist not to fatil to recognize an aneurysm but also the tumoral pathologies of the mediastinum which are much more frequently responsible for the compression of the pulmonary artery.

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