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Coarctation of the aorta in adults with double superior vena cava: a case report

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ABSTRACT

Coarctation of the aorta is an isthmus stenosis of the aorta, which is rarely seen in adults. It affects 1/5000 newborns, with a male predominance. It is associated in most cases with other diseases such as bicuspid aortic valve, dystrophic aneurysm of the ascending aorta, mitral regurgitation or coronary insufficiency. The association in a double superior vena cava is rare. If conventional treatment is surgical resection suture proposed since 1945. It is considered today that after this treatment the patients are not cured and require special attention. Endovascular treatment has been used for years in children. It is indicated mainly in case of recoarctation. In adults, this technique has had a lot of progress in recent years. Indeed, in case of native coarctation with favorable anatomy, angioplasty with placement of a stent has become the treatment of choice in many centers.

INTRODUCTION

Coarctation of the aorta is an isthmus stenosis of the aorta. It can be observed in the neonatal period. Most often the diagnosis of coarctation in infants is referred to the combination of a breath and femoral pulses diminished or abolished. More rarely, it occurs in adulthood during a blood pressure assessment. It affects 1/5000 newborns, with a male predominance. It is associated in most cases with other diseases such as bicuspid aortic valve, dystrophic aneurysm of the ascending aorta, mitral regurgitation or coronary insufficiency.

MATERIALS AND METHOD

We report the case of a 45 year old man with no history of particular disease, with cardiovascular risk factors such as high blood pressure recently discovered. He was admitted to our clinic for hypertensive crisis complicated with moderate chronic renal failure. The review found a significant difference in blood pressure between the upper limbs and lower limbs. The ulnar and radial pulses were present, but the pulse of both lower limbs was abolished. Dipstick noted traces of protein. The resting electrocardiogram registered sinus and regular rhythm with disorders of repolarization in lateral territory. Chest radiography showed a normal heart size and rib notching caused by the dilated intercostals arteries (Figure 1). The cardiac ultrasound showed an important left ventricular

hypertrophy. Size and systolic left ventricular function were normal. The ascending aorta was of good size with a tricuspid aortic valve. The usual biological assessment was unremarkable apart renal failure with creatinine clearance 58.5 ml / min / 1.73 m² according to MDRD. Proteinuria 24 h was 0.2 g / 24h. The thoraco-abdominal computed tomography (CT) scan showed a coarctation of the proximal portion of the descending thoracic aorta, spanning 9 mm (Figure 2) and a permeable double superior vena cava (Figure 3). The patient underwent successful endovascular treatment (Figure 4) of the coarctation, with placement of a stent (Figure 5).The various checks after injections iodinated contrast agents noted stability of renal function.

The particularity of this observation is firstly discovered in adulthood aortic coarctation and also its association with a double superior vena cava.

RESULTS AND DISCUSSION

Coarctation of the aorta is a common malformation that accounts for 7% of congenital heart malformation. It is seen in about a newborn in 3000 so it relates 400-500 newborns a year in France. Over 90% of them will reach adulthood. It is two to three times more common in boys than in girls. It is found in nearly 25% of Turner syndrome. She usually carries a localized narrowing of the aortic arch usually located at the isthmus region bounded by the origin of the left subclavian artery, first, and the insertion of the arterial ligament other. The narrowing may sometimes extend over the entire length of the aortic isthmus or may be accompanied, upstream, more or less long hypoplasia of the transverse aorta. Morgagni was the first to describe this anomaly during the autopsy of a monkey in 1760 (1).

In 1903, Bonnet gave a more detailed description in humans and identified two forms depending on their position in relation to the arterial duct (ductus arteriosus). He distinguished between "child" forms (preductal) and "adults" forms (postductal) (2). Rarely coarctation of the aorta can also be located in the abdominal aorta. Associated injuries are common. A bicuspid aortic valve is found in 1-2% of the general population against 50-85% of patients with coarctation of the aorta. It associates itself with a fragility of the aortic wall, the texture is similar to that of aortic walls of patients with disease of the elastic tissue as Marfan and shares with these diseases the risk of aneurysm of the ascending aorta and aortic dissection (3).

The exact pathophysiology of the isthmic coarctation of the aorta remains uncertain but two hypotheses are frequently given. During fetal life, decreased blood flow in the ascending thoracic aorta secondary to left heart obstructive lesion or a ventricular septal defect would explain in some cases the presence of a coarctation associated with hypoplasia of the aortic arch. In other cases, the extension of the contractile ductal tissue to isthmic region would cause, when spontaneous closure of the ductus arteriosus after birth, a constriction of the aorta (4). In our case, no embryological explanation has been provided in the literature to the association between coarctation of the aorta and a double superior vena cava.

The double superior vena cava is a rare abnormality present in 0.3 to 0.5% of the general population (5). In nearly 40% of cases, it may be associated with other cardiac anomalies such as atrial septal defect (6,7), bicuspid aortic valve, coarctation of the aorta, pulmonary atresia, atresia of the ostium of the coronary sinus (8), or cor triatriatum. The association with anatomical or architectural abnormalities sinus node or conduction tissues has also been described (9).

The explanation of the anomalies of superior cava system venous return requires knowledge of its embryology. In 80-90% of cases, the venous return is made in the right atrium via the coronary sinus and is inconsequential hemodynamic, the persistent left superior vena cava is asymptomatic and is discovered incidentally during central venous catheterization. In other cases, venous return may flow into the left atrium leading to a right to left shunt, may be causing cyanosis and exposing to a risk of paradoxical embolism. Confirmation of this type of malformation required performing a

contrast echocardiography and cardiac catheterization. Currently chest CT angiography and MR angiography are noninvasive alternatives (10). Multiplane Transesophageal echocardiography can also be used to establish the diagnosis (11).
Therapeutically, surgical treatment of coarctation is performed for more than 60 years. It is most often practiced by left thoracotomy. Sternotomy is performed only when cardiopulmonary bypass is required to treat an intracardiac anomaly associated or when there is a severe hypoplasia of the aortic arch or a complex coarctation.Crafoord (12) described in 1945 the intervention of resection anastomosis which is addressed to localized coarctations of the aortic isthmus. The intervention of Waldhausen (13) sacrifices the left subclavian artery and uses its proximal portion by folding down on the aortic isthmus performing an enlargement plasty without the use of prosthetic material. In case of complex coarctation, the establishment of a bypass tube between the ascending aorta and the descending aorta overcomes safer vascular obstacle (14). The 1980s saw the birth of interventional catheterization techniques. The balloon angioplasty of coarctation was proposed in 1982 for localized stenosis (15,16). However, this technique involves the risk of aortic aneurysms at the dilated area. Few teams currently advocated in pediatric age. In adults, with the improvement of technology, the risks of aneurysms are lower in the order of 5%. Nowadays, most teams couple the balloon dilatation with the establishment of a stent which reduces both the risk of recoarctation and occurrence of aortic aneurysm (17,18).

CONCLUSION

Coarctation of the aorta is a disease of the arterial system that requires regular monitoring during life, by doctors trained to specific complications which may occur in these patients. Coarctation should be systematically suspected in hypertension of the child, in whom surgery remains the treatment of choice. In adults, balloon dilatation of coarctation and recoarctations, coupled with the establishment of a stent reduces the risk of recurrence and occurrence of aortic aneurysm. The significant risk of recoarctation imposes a regular cardiac monitoring. However, the association with a superior double vena cava has rarely been described and its embryonic explanation could be adopted due to the pathophysiology of these two malformations.

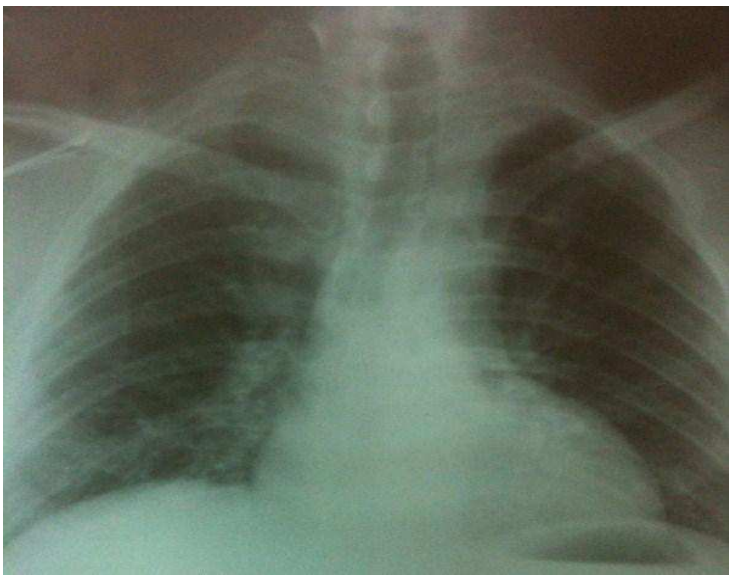

Figure 1: Chest radiography showing a normal heart size and rib notching caused by the dilated intercostals arteries.

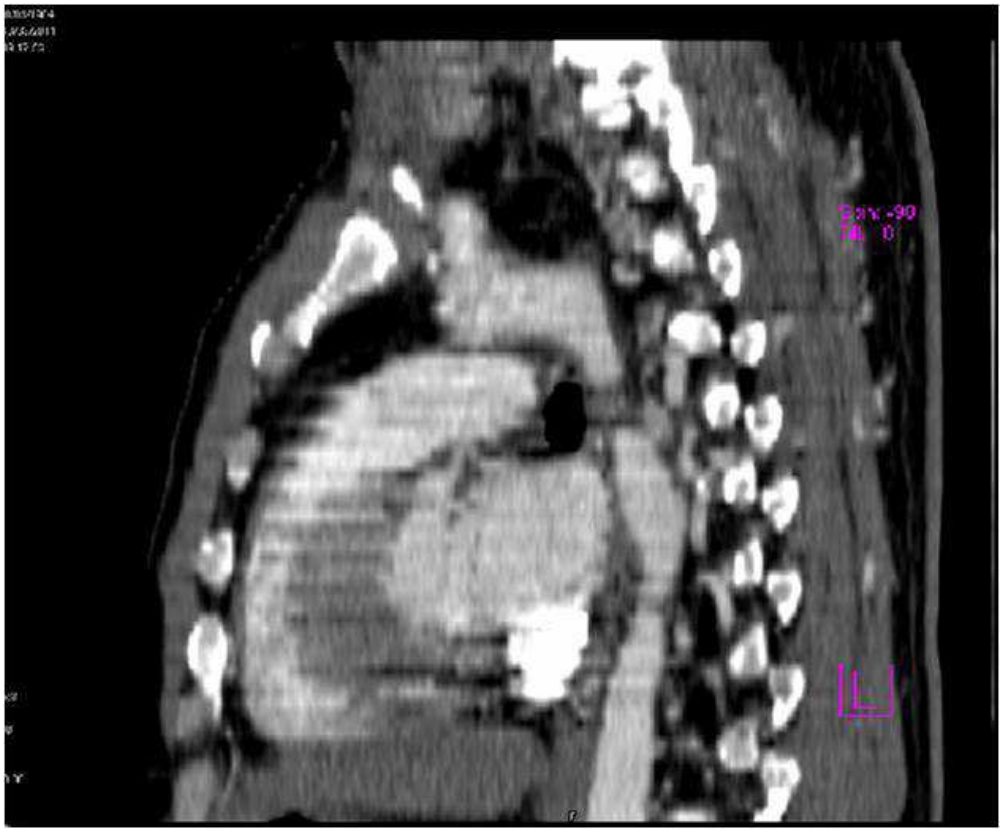

Figure 2: Coarctation of the proximal portion of the descending thoracic aorta.

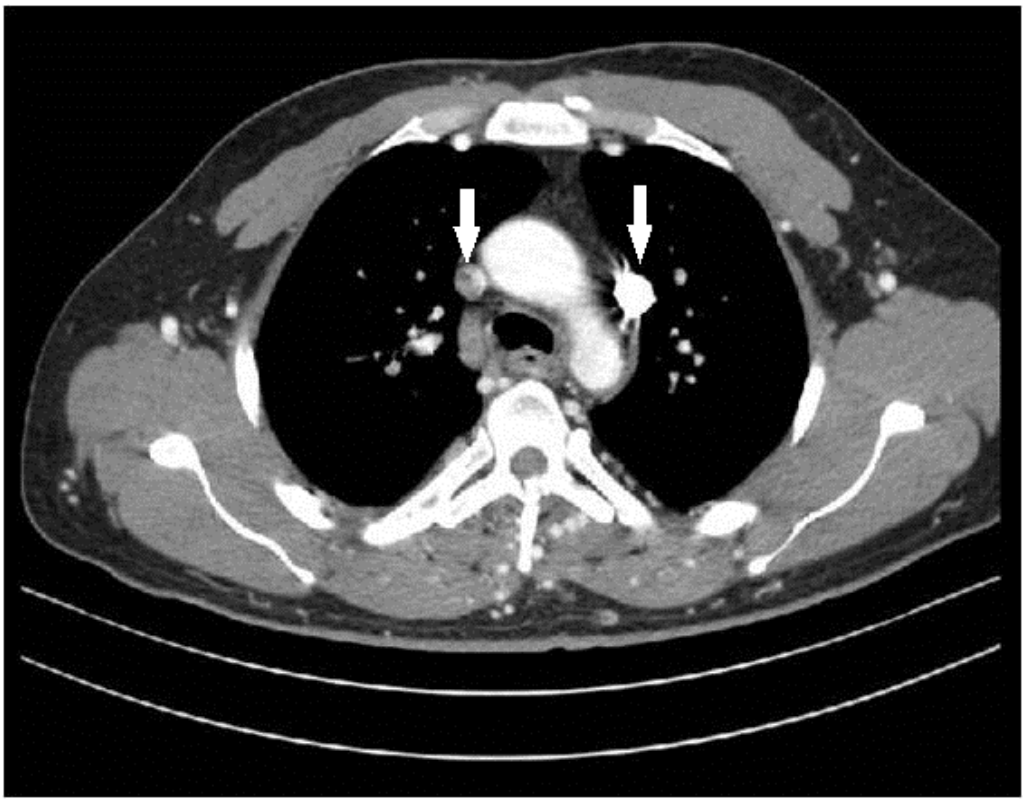

Figure 3: Chest CT Scan Angiography: axial section showing the presence of 2 superior vena cava (white arrows).

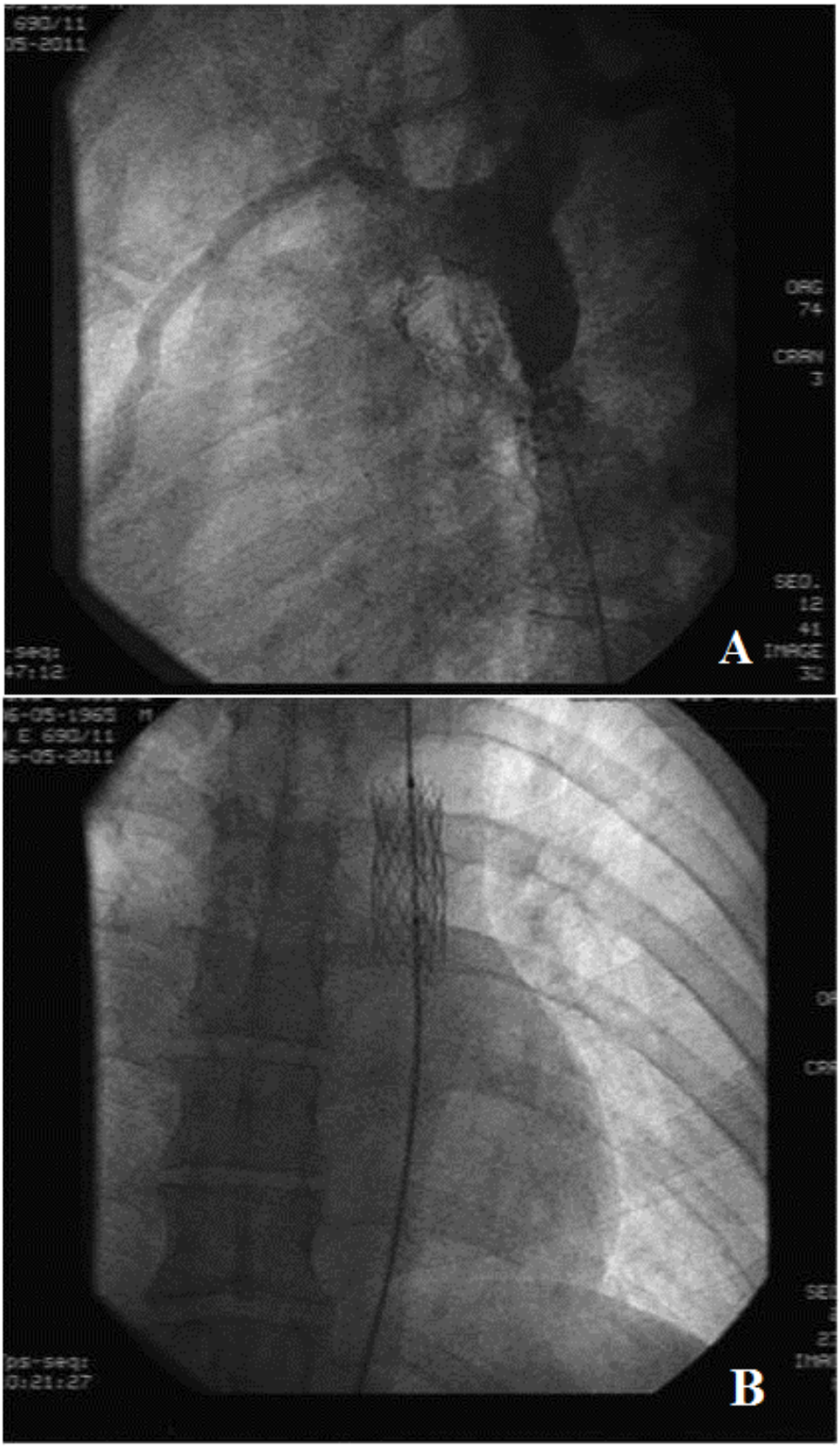

Figure 4: Balloon Dilatation and implementation of a stent.

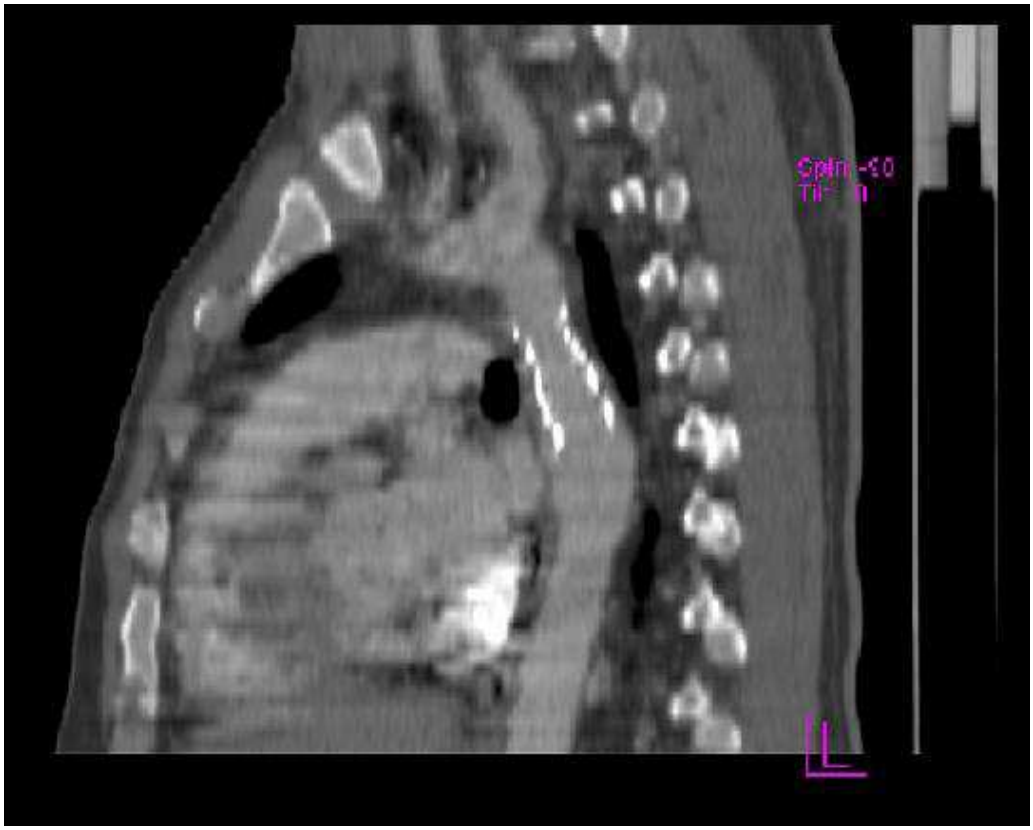

Figure 5: Chest CT Scan angiography: aortic endoprosthesis in place.

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