Title : chest pain in the setting of scimitar syndrome.

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Running title : scimitar syndrome , congenital heart disease

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Introduction :

Scimitar syndrome (SS) is rare heart malformation occurring in one to three 100,000 live births [1].

Patients can be divided into two groups: infantile and adult depending on the severity of symptoms

and associated malformations. The incidence may be higher in adult, due to the minor form of the

Syndrome, they remain asymptomatic or mildly symptomatic for many years and are able to lead a

normal life. Thus, the adult form is not frequently reported in the literature . We’ll describe an

unusual presentation with atypical chest pain in one of the two consecutives cases.

Case 1 :

A 22- year- old boy presented in New York Heart Association functional class 2 with history of

Transient palpitations. Physical examination revealed a constant splitting of the second heart sound

And a grade 2/6 systolic ejection murmur on the left sternal border. The ECG showed signs of right

Ventricular hypertrophy. A chest X-Ray revealed a mediastinal shift to the right with dextrocardia

and small right hemithorax. Echocardiography showed a partial anomalous pulmonary venous

connection of the right pulmonary veins to inferior vena cava without associated atrial septal

defect and marked dilatation of the right ventricle (RV): RV end-diastolic diameter of 50 mm with a ratio of RV to left ventricle (LV) > 1. A computed tomography allowed the visualization of the anomalous vessel draining the right lung in totality into the inferior vena cava . He underwent surgery using intracardiac baffle repair technique by baffling the orifice of the scimitar vein(SV) into the left atrium through a newly created atrial septal defect . The materiel used was autologous pericardium and the procedure was performed under cardiopulmonary bypass at 25° C without circulatory arrest (figure 1) . The patient was discharged from the hospital at day 7 postoperatively. Echocardiography showed no distorsion or stenosis of the tunnel , specially at the vena cava end. The patient doesn’t report any episode of palpitations at one year follow-up.

Case 2 :

A 57 year –old – women attended the cardiac department complaining of chest

discomfort and dyspnea on exertion . She was on CCS grade 4 and NYHA class 2 symptoms. Her

breathlessness was found to trace back on young adulthood and worsening over time and her angina

was present in the preceding 6 weeks . Coronary risk factors included menopause and

hypercholesterolemia. ECG showed an incomplete right bundle branch block with ST wave change

from V1 to V3. The chest X- ray revealed moderate right lung hypoplasia with dextroposition of the

heart , indistinct right heart border and the SV collector near the cardiophrenic angle.

Echocardiography demonstrated that right pulmonary veins are not joining the left atrium and the

SV collector to the inferior vena cava associated with mild pulmonary hypertension and marked

right ventricular dilatation: RV end-diastolic diameter of 53 mm and RV/LV > 1 . Computed tomography identified the SV collector and revealed right lung hypoplasia (figure 2).The coronary angiography didn’t reveal any stenoses .She underwent surgery using intracardiac baffle technique repair by autologous pericardium , as case 1.The postoperative recovery was uneventful and the patient was discharged home ten days later. Echocardiography doesn’t reveal any obstruction of the tunnel. Six months after surgery , the patient remained completely asymptomatic.

Discussion :

The first anatomical observation of this peculiar anomalous venous connection was described by

Cooper and Chassinat in 1836 but the term “scimitar” didn’t appear in their reports. This anomaly

Accounts for 3% - 5% of partial anomalous pulmonary venous connection[2] but the true incidence

may be higher in “adult” forms because many patients remain asymptomatic. Available small

retrospective series and review confirm that “infantile” forms of the syndrome have a higher

incidence of symptoms , aortopulmonary collaterals , associated congenital heart defect ,

extracardiac anomalies and pulmonary hypertension and have a worse prognosis compared with

patients diagnosed later[3]. On the other hand, adult patients, because of the mild form of the

syndrome, are often asymptomatic or may present with dyspnea or arrhythmias , as in case 1 .

Chest pain was the main symptom in our second patient, the only reported case of angina revealing

a scimitar syndrome was described in 66 -year-old women that had associated ischemic heart

Disease [4]. Our patient had some coronary risk factors but the coronary arteries were free from

Stenoses at the coronary angiography. The only way to explain this angina was the marked right

ventricular dilatation, related to the long lasting left to right shunt of the entire right lung , that

tether the pericardium causing chest pain which was independent from physical activity.

Common indications for surgical treatment in adult patients include : dyspnea (NYHA 2 or

more), marked right heart chambers dilatation , and lung sequestration with recurrent respiratory

infection. The intracardiac repair and reimplantation technique have been reported for many years

as the most frequently used reparative techniques. However, there is no consensus for which is the

best surgical option. The first method consists in the creation of a long intracardiac tunnel by baffling

the SV orifice to the left atrium . The second one, includes the disconnection of the SV from the IVC

and reimplanting it to the left atrium. The risk of postoperative SV stenosis is the same for both

procedure but is higher in infant than adult. Some authors, have reported balloon dilatation

to deal with postoperative SV obstruction[5]and the right pneumectomy is reserved when patients

continue to complain from severe respiratory symptoms.

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Figures – legends:

Figure 1 : operative view showing the intracardiac baffle.

Figure 2 :CT scan revealing a scimitar vein (a) and right heart chambers dilatation(b).