Case report - Congenital

Hybrid approach in a case of arterial tortuosity syndrome

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Abstract

Arterial tortuosity syndrome is a rare connective tissue disorder characterised by elongation, tortuosity, stenosis and aneurysms of the large and middle-sized arteries. The symptomatology is correlated to the artery affected by the pathology with correlated stenosis. We describe our hybrid surgical procedure in the treatment of a case of kinking of the pulmonary branches with significant gradient and hypertension. Aortic arch and supraaortic vessels presented various deviousness without hemodynamic alterations.

Keywords: Pulmonary arteries; Anatomy; Stents

1. Clinical summary

A four-year-old patient came to our Department because of the rise of dyspnoea and asthenia during a usual game activity.

During hospital stay, echocardiography investigation excluded heart diseases and showed a pulmonary hypertension. The angiographic evaluation revealed a severe stenosis of pulmonary branches associated to an excess in length (Fig. 1a,b). The angiographic image in Fig. 1c shows the characteristic morphology of aorta and supraaortic vessels. Fig. 2a,b shows the operative images of the deviousness and excessive length of pulmonary arteries. Angiographic images were characteristic for arterial tortuosity syndrome, a rare autosomal recessive connective disease. A genetic study has mapped the arterial tortuosity syndrome gene to chromosome 20q13 that determines abnormalities of the elastin network in the major arteries [1].

Our therapeutical strategy was based on a hybrid surgical and interventional cardiologic approach. The surgeon performed a median sternotomy, opening of the pericardium, and isolation of the pulmonary branches. An introducer was placed at the pulmonary bifurcation and the cardiologists positioned a stent in the superior left pulmonary branch and one in the lobar right superior branch (Fig. 1d).

Next, the surgical team established a cardiopulmonary bypass through an aortic and bicaval incannulation. An arteriotomy was performed through the pulmonary artery and its branches, and another stent was positioned in the lobar right inferior branches for a direct surgical view (Fig. 2c,d). We performed a plasty of the pulmonary artery and its branches. A quadrangular portion of arterial wall was resected by pulmonary bifurcation, and a direct suture of branches and pulmonary artery restored a normal length to these. The procedure required 32 min of cardiopulmonary bypass. The patient presented an uncomplicated postoperative period, and was discharged seven days after the intervention. We have followed the patient in the postoperative period with echocardiographic and angiographic examinations that revealed severe decreasing of pulmonary pressure to a value of 40 mmHg and a good right ventricular function at six months after intervention.

2. Comment

The collaborative interaction between surgeons and interventional cardiologists, supported by the advancement of new technologies, has enabled the development of new hybrid treatment strategies for patients with various heart diseases.

Hybrid strategy for the management of hypoplastic left heart syndrome, described by Gibbs et al. [2], has initial palliation. This new approach was based on stenting of the Ductus Arteriosus combined with banding of the pulmonary arteries and atrial septectomy or septostomy.

Bacha and colleagues reported a single centre experience, in 25 paediatric patients, on a hybrid technique for repairing a ventricular septal defect through Amplatzer followed by surgical intervention for associated cardiac defects [3].

Also, in adult patients affected by multiple vascular diseases, hybrid techniques have been reported. Surgical treatment of simultaneous coronary and carotid disease is still controversial, because of the high risk of morbidity and mortality after combined or staged carotid artery endarterectomy and the coronary artery bypass grafting approach.
Naylor and coworkers reported a hybrid strategy in simultaneous revascularisation by carotid stenting associated to myocardial revascularisation through coronary artery bypass grafting [4].

Recent advances in endovascular techniques have enabled non-surgical treatment of aortic coarctation by balloon dilatation with or without stenting. The treatment of coarctation of aorta associated to an additional cardiac lesion in adults remains a difficult surgical challenge. The mortality and morbidity of a staged surgical approach is significant, irrespective of the sequence of repair. A surgical combined procedure requires establishing cardiopulmonary bypass with right atrial cannulation and double cannulation of the ascending aorta and femoral artery, repair of the cardiac lesion, and concurrent transpericardial ascending aorta to descending thoracic aortic bypass graft. The complexity of the procedure determines an increased operative risk. Yiu et al. described a successful hybrid management of coronary artery disease and coarctation of aorta [5].

Also, in our experience, hybrid strategy has allowed an easier solution to a complex disease. A surgical approach in our patient would have required an extensive reconstruction of the pulmonary and lobar branches, with a long time on cardiopulmonary bypass. Moreover, our Institution did not have a previous experience in the surgical management of a so rare vascular malformation.

Our successful report led us to sustain hybrid approaches in the management of selected patients with complex pathologies.

References


