Ebstein's anomaly and left proximal pulmonary artery stenosis complicated by pulmonary embolism: a case report

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The clinical course of Ebstein's anomaly patients varies considerably. Commonly associated cardiac structural defects include: ASD, PFO, pulmonary stenosis and VSD.

We present a case of a 26-year-old patient admitted with dyspnea, exercise intolerance, epistaxis and tachycardia deteriorating for 4 months prior to admission. In a CT angiography performed prior to admission, a stenosis of the proximal left pulmonary artery was noted with a post-stenotic dilatation and a well developed bronchial circulation. A trans-thoracic echo upon admission showed a typical image of Ebstein's anomaly with the septal leaflet of the tricuspid valve displaced into the RV cavity, attached to a hypertrophied myocardium. Severe eccentric tricuspid regurgitation was present. Right heart catheterization measured a low pulmonary pressure of 30 mmHg. The catheter was blocked at the proximal left pulmonary artery. A CT angiography revealed a complete occlusion of the left pulmonary artery. In surgery, the septal leaflet of the tricuspid valve was dysplastic and non-functional with an abnormally high insertion of both the septal and posterior leaflets. The patient underwent pulmonary thrombo-endarterectomy, RVOT myotomy and tricuspid valve replacement with a mechanical valve ATS 31 mm. Histologically, the excised leaflets contained fibrinoid necrosis. The postoperative course was uneventful and the

patient was discharged on the 7th postoperative day and is doing well since (two months).

We conclude that the patient underwent an acute event of pulmonary embolism with a background congenital pulmonary artery stenosis and Ebstein's anomaly. This case is significant for the unusual association of two rare congenital entities not known to be related.

KEYWORDS: Ebstein's anomaly, pulmonary artery stenosis, pulmonary embolism.

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