Case report

Tetralogy of Fallot with restrictive ventricular septal defect by accessory tricuspid leaflet tissue

Mahipat Raj Soni a,*, Deepak A. Bohara a, Ajay U. Mahajan b, Pratap J. Nathani c

a Resident, Department of Cardiology, Lokmanya Tilak Municipal Medical College, Sion, Mumbai 400 022, India
b Professor and Unit Head, Department of Cardiology, Lokmanya Tilak Municipal Medical College, Sion, Mumbai 400 022, India
c Professor and Head, Department of Cardiology, Lokmanya Tilak Municipal Medical College, Sion, Mumbai 400 022, India

Abstract

In tetralogy of Fallot septal defect is usually large because of malalignment of outlet septum, restrictive defect has been reported rarely. We present a case of tetralogy of Fallot with accessory tricuspid leaflet tissue restricting ventricular septal defect. The report includes echocardiographic and catheter images of this rare presentation of tetralogy of Fallot.

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01. Key message

Tetralogy of Fallot with a restrictive ventricular septal defect and suprasystolic right ventricular pressure is a rare anomaly. Most common cause of restriction is accessory tricuspid leaflet tissue. Early surgical correction should be done to prevent right ventricular failure. The precise morphology of the accessory tricuspid valve tissue is of considerable surgical significance.

1. Introduction

In Tetralogy of Fallot VSD may be restricted by the accessory tricuspid valve tissue, tricuspid valve vegetation, prolapsed aortic valve and membranous flap, most common being accessory tricuspid valve tissue. Patients with accessory tricuspid valve tissue without signs of a restrictive ventricular septal defect might be overlooked during operation, so preoperative diagnosis is important. In cases of TOF with restrictive VSD, the RV systolic pressure shows post ectopic potentiation and ventricular pressure trace shows a characteristic 'isosceles triangle' pattern. These patients are prone to develop right heart failure.

2. Case history

12-year-old child was admitted with complaints of effort intolerance and cyanosis since early childhood, squatting for 5
years, and recurrent syncope for 1 year. He had no history of cyanotic spells. He had history of left sided hemiparesis at 10 years of age, from which he recovered completely. On examination, he had uniform central cyanosis, grade 3 clubbing, prominent ‘a’ wave in Jugular venous pulse, RV apex, grade 3 parasternal heave, single second heart sound, 3/6 ejection systolic murmur at second left intercostals space and 3/6 pansystolic murmur at left lower sternal border. His systemic oxygen saturation was 85% with PCV of 60%. Other routine blood investigation were normal. Electrocardiogram showed sinus rhythm, right axis deviation, clockwise loop and right ventricular hypertrophy with strain pattern. Echocardiography revealed evidence of overriding of aorta, right ventricular hypertrophy, evidence of accessory tricuspid leaflet tissue restricting VSD (Figs. 1 and 2), subaortic restrictive VSD with right-to-left shunt with Doppler gradient of 111 mmHg (Fig. 3) and severe infundibular and valvar pulmonary stenosis with peak Right ventricular outflow tract gradient of 179 mmHg (Fig. 4). The branch pulmonary arteries were confluent. Cardiac catheterization revealed non equalization of pressures in both ventricles with pressure difference of 83 mmHg between the right ventricle and the left ventricle but did not showed classical isosceles right ventricular tracing (Fig. 5). Right ventricular angiogram revealed thickened and doming pulmonary valve, infundibular hypertrophy and subaortic ventricular septal defect with accessory tricuspid leaflet restricting VSD (Fig. 6). LV angiogram showed overriding of aorta and right sided aortic arch, normal origin and course of Coronaries.

Intraoperatively, no major coronary artery crossing RVOT seen. Patient was put on cardiopulmonary bypass under moderate hypothermia (22–25 °C). Right atrial incision given paraseptally, accessory tricuspid leaflet tissue found to be anchored to posterior interventricular septum by short chordae and was immobile. VSD was closed with Dacron patch with interrupted pledgeted sutures with accessory tricuspid leaflet tissue was used as anchorage over posterior margin of ventricular septal defect. On Right ventriculotomy, pulmonary valve was bicuspid with fused commissures. Pulmonary valvotomy was done with easy passage of Hegar 16 with subsequent RVOT coring. After which, subannular patch-plasty with autologous fresh pericardial patch was performed. Right atrium closed with 5-0 prolene in continuous double layer. Patient rewarmed and weaned off CPB in sinus rhythm. Post operative RV/LV pressure was 0.4. Post operative course was uneventful.
3. Discussion

Tetralogy of Fallot associated with a restrictive ventricular septal defect due to accessory tricuspid leaflet tissue rare anomaly. Exact incidence of this rare presentation is not known. MF Flanagan describes the echocardiographic and anatomic features in 4 patients with a restrictive ventricular septal defect among 269 patients with tetralogy of Fallot undergoing surgical repair with incidence of 1.48%. Accessory or excessive tricuspid valve tissue obstructs the defect in a manner similar to spontaneous closure of isolated membranous ventricular septal defects. In patients with tetralogy of Fallot, recognition of an obstructed ventricular septal defect is important because it has a poor prognosis secondary to right ventricular pressure overload and subsequent right ventricular failure.

Faggian and associates described two types of accessory valve tissue, the mobile type and the fixed type. The mobile type is a parachute-like leaflet floating in the ventricular outflow tract, and may cause ventricular outflow obstruction. This type of accessory valve tissue is sometimes dysplastic.

The fixed type is firmly anchored to the interventricular septum by short chordae and lacks mobility. This type of lesion sometimes decreases the size of the VSD. Gunter Kerst reported a new born with restrictive VSD and critical subaortic stenosis due to prolapse of redundant accessory tricuspid leaflet tissue. Recently McBrien has reported case of tetralogy of Fallot with progressive restriction of the VSD by tricuspid valve tissue.

In TOF, right ventricular pressure does not exceed the systemic level because of the nonrestrictive VSD. However, rarely, flow across the VSD may be restricted by the accessory tricuspid valve tissue, tricuspid valve vegetation or a prolapsed aortic valve leaflet or membranous flap. These children have suprasystolic right ventricular pressure. The right atrial pressure trace shows a prominent ‘a’ wave which is a hallmark of restrictive VSD. The right ventricular pressure trace shows a characteristic ‘isosceles triangle’ pattern against a rectangular shape seen in those with nonrestrictive VSD. Following a ventricular premature beat, the left ventricular pressure shows mild or no augmentation in the systolic pressure because of the reflex systemic vasodilatation. In cases of TOF with restrictive VSD, the RV systolic pressure shows post ectopic potentiation. This phenomenon is typically absent in TOF as the right ventricle is influenced by the left ventricular pressure changes due the presence of nonrestrictive VSD.

The precise morphology of the accessory tricuspid valve tissue is of considerable surgical significance. It is possible that the accessory tricuspid valve tissue without signs of a restrictive VSD might be overlooked during operation. Surgeon should be familiar with the clinical and pathological features of accessory valve tissue so as not to overlook it during surgery. When mobile, the tissue must be resected at the time of surgical repair since it may result in left ventricular outflow tract obstruction if permitted to remain on the left ventricular aspect of the patch at the end of the procedure. In contrast, the fixed variety need not necessarily be excised. It may be left in situ and it can be used as a suture anchorage during closure of the VSD as done in our case.

In conclusion, Tetralogy of Fallot is rarely associated with restrictive VSD and this is most commonly caused by accessory tricuspid leaflet tissue. Right ventricle will show suprasystolic pressure. Early surgical correction should be done to prevent right ventricular failure.

Conflicts of interest

All authors have none to declare.

References

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**Obituary**

The Cardiological Society of India expresses deep condolences to the family members of Dr. D.S. Ursekar, Pune who expired recently. He has been a long standing member of Cardiological Society of India.