CASE REPORT

ACCESSORY MITRAL VALVE TISSUE CAUSING FEATURES OF LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION-A CASE REPORT AND UPDATED LITERATURE REVIEW

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Accessory mitral valve tissue is a rare congenital anomaly that is commonly incidentally diagnosed. When symptomatic, it tends to present with features of left ventricular outflow (LVOT) obstruction in about two thirds of cases. It is also commonly associated with other congenital anomalies, notably ventricular septal defects. Transthoracic echocardiography is a very useful diagnostic test to make a diagnosis and its widespread use will increase detection of this condition. We are presenting a case of a 29-year-old lady who presented with breathlessness in the third trimester of pregnancy and was subsequently found to have evidence of accessory mitral valve tissue on echocardiography.

Keywords: Accessory mitral valve; Outflow obstruction

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INTRODUCTION

Accessory mitral valve tissue (AMVT) is a rare congenital anomaly with an incidence of 1% in the general population[1] and there are around 124 cases reported in the literature. It is an incidental finding in most cases and is rarely symptomatic. It is commonly associated with other congenital malformations of the heart notably ventricular septal defect (VSD). It is mostly diagnosed in childhood; however, it might be diagnosed later in life, and the age of onset of diagnosis is currently debatable. Symptoms, when they occur, include breathlessness, palpitations and chest pain. Transthoracic echocardiography (TTE) is a very useful test to make the diagnosis. Here we present a case of accessory mitral valve tissue becoming symptomatic during pregnancy.

CASE PRESENTATION

A 29-year-old lady with a history of semi-membranous VSD repair as a child, and no other comorbidity, was referred to cardiology services for assessment prior to her fourth delivery as she was found to have a murmur. She has three daughters, all born vaginally with no complications. Her third pregnancy had to be induced due to gestational diabetes requiring insulin therapy. She was well in her first two pregnancies from cardiovascular point of view; however, she became quite breathless in the third trimester of her third pregnancy. She didn't have any history of chest pain or syncope. Cardiovascular examination revealed an ejection systolic murmur but no other abnormalities. Her ECG showed sinus rhythm and right bundle branch block with a QRS duration of 144 ms. Her TTE showed an accessory structure in the left ventricular outflow tract (LVOT) and that was thought to be an accessory mitral valve tissue (Figure-1), which was subsequently confirmed on a trans-oesophageal echocardiogram (TOE). The critical point was to assess the hemodynamic effects of this finding, and to exclude any significant LVOT obstruction that might develop in the third trimester of pregnancy and at the time of labour. TOE was performed under general anaesthesia, and it confirmed an accessory mitral valve tissue at the LVOT with no evidence of obstruction at rest with a gradient of just 11 mmHg, however, this increased to 74 mmHg at a maximum heart rate of 120 beats/min with administration of isoprenaline. We concluded that this would have been the explanation for her breathlessness in the last pregnancy. We recommended beta blocker treatment in the third trimester if she develops symptoms of breathlessness to decrease heart rate and outflow tract obstruction. She had to be started on beta blockers during the third trimester, which resulted in symptomatic improvement and she underwent normal spontaneous vaginal delivery without complications.

Literature review:

Accessory mitral valve tissue is an uncommon congenital abnormality[1,2] with an estimated incidence of 1/26000 on TTE[1], however the exact prevalence of this condition is probably underestimated as this condition is frequently first diagnosed intraoperatively[1]. The first case was reported in 1842 and McLean has introduced the first surgical management of this condition in 1963.[1,2] It is often detected in children, and rarely in adults.[2]

Accessory mitral valve tissue is a congenital abnormality that occurs during the delamination stage of endocardial cushion development. Though exact embryogenesis of this condition is unclear, it is thought to be due to incomplete separation of the

mitral valve from the endocardial cushion[1,3,4] and presence of associated congenital and vascular cardiac abnormalities supports this theory[1].

A literature review by Manganaro R *et al* in 2014, identified 104 cases[1] and we identified 12 more case in our more recent review up to July 2017.

Prifti *et al* classified AMVT into two types each with 2 subtypes. Fixed (Type I; type Ia= nodular, and type Ib = membranous) and mobile (Type II; Type IIb = pedunculated, Type IIb = leaflet like).[5]

AMVT is one of the three known congenital abnormalities of mitral valve, the other two including parachute mitral valve, and mitral valve with abnormal attachment of anterior leaflet to ventricular septum.[6,7]

Accessory mitral valve tissue is commonly associated with congenital abnormalities, commonly VSD, like in our case.[1] Indeed, two thirds of AMVT are associated with other congenital abnormalities.[8]

Accessory mitral valve tissue is a recognized cause of LVOT obstruction, and also of sub-valvular aortic stenosis. Left ventricular outflow tract obstruction tends to be progressive which explains the late development of symptoms in patients with AMVT.

In asymptomatic subjects AMVT is incidentally diagnosed.[1] Presence of a murmur in an otherwise asymptomatic subject can trigger the diagnosis.[1,3,6] As this abnormality is frequently associated with other congenital heart disease, presentation could be linked to the presence of associated anomalies.

When symptoms occur, they usually do in the first two decades of life. Symptoms are mostly related to LVOT obstruction which is seen in about two thirds of patients. Left ventricular outflow tract obstruction is either due to progressive deposition of fibrous tissue due to turbulence created by AMVT or due to mass effect of the accessory tissue itself if it is initially sizable. Symptoms include exertional shortness of breath, chest pain, syncope, and rarely cardio-embolic phenomena, and infective endocarditis.[1–3,9] Severity of symptoms depends on severity of LVOT obstruction[3], and symptoms develop only when LVOT outflow gradient increases to 50mmHg or above.

Another thing to note is that the severity of LVOT obstruction might be underestimated in presence of interventricular shunt[5], which could happen in those with an associated VSD like in our patient, though this is not applicable to her as she had a VSD closure procedure as a child.

In asymptomatic patients, indications for surgery would be significant LVOT obstruction or prophylactic excision of AMVT during surgery performed for other indications.[4,5,7]

When symptoms develop, surgical resection is warranted. Beta blockers are an option in patients who develop increased LVOT gradients on exertion like in our patient, who was actually treated with beta blockers during her last trimester of pregnancy. She progressed successfully through normal vaginal delivery without issues.

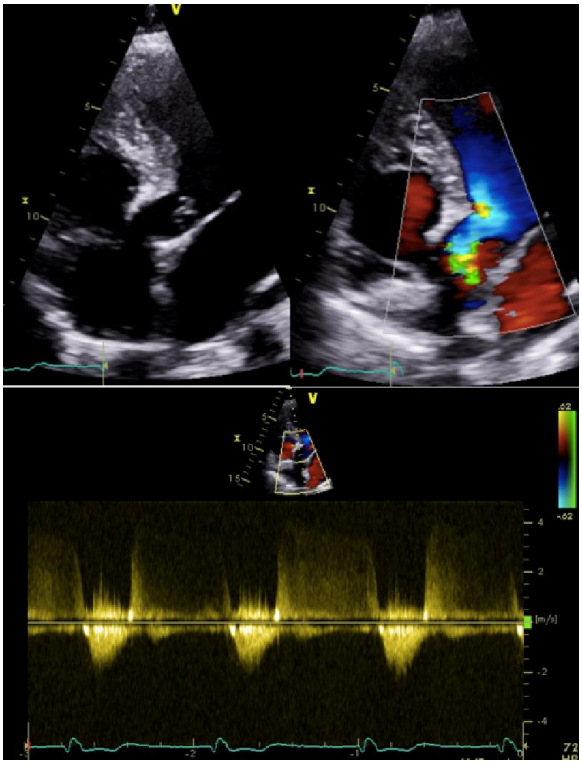

Figure-1: Accessory mitral valve tissue in LVOT causing turbulence at rest – 5 chamber view TTE.

CONCLUSIONS

The widespread use of echocardiography, which is currently the preferred imaging modality for AMVT, has made detection of this anomaly much easier. Echocardiography has a vital role in follow up and management of this condition. Other imaging modalities like cardiac computed tomography and cardiac magnetic resonance imaging can be used, and there is a growing interest in these modalities particularly when it comes to differential diagnosis of cardiac masses.[1]

In the differential diagnosis of LVOT obstruction, AMVT should be considered, especially in pre- and post-operative congenital heart disease patients[3], as this anomaly is consistently detected with different other malformations. While AMVT produces symptoms related to LVOT obstruction in

the majority of cases, it can also be an incidental finding.

Surgery is the mainstay of treatment in symptomatic patients. Beta blockers can be used in patients who become symptomatic on exertion in selected patients.

AUTHORS' CONTRIBUTION

AA: Writing of the initial draft of the case report and conduct of initial literature search. HK: Concept of manuscript, co-participation in literature search and editing of the report text. MS: Consultant in cardiovascular imaging proof read the document before publication.

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