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Type A Aortic Dissection following a Tetralogy of Fallot Repair

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Central Picture Legend:

Dilated aortic root measuring 7cm with dissection flaps

CENTRAL MESSAGE

An underlying TOF aortopathy may predispose to the rare occurrence of a type A aortic dissection following a TOF repair.
Summary

Progressive aortic root dilatation has been noted following repair of complex congenital heart diseases such as Tetralogy of Fallot (TOF)\(^1\). Of the dilated aortic pathologies post-TOF repair, aortic aneurysms are more frequently noted than aortic dissections. A 55-year-old gentleman presented with a subacute Type A aortic dissection (TAD) following a childhood TOF repair. The diagnosis of TAD was made incidentally following a routine echocardiogram, and characterisation of the dissection was completed via a computed tomography (CT) scan. Although complicated by a prolonged post-operative period, the TAD repair was successful with a good clinical outcome.

Introduction

Whilst aortic root dilatation has been recognised as a complication of conotruncal anomalies, TAD is an exceptionally rare complication with only seven individual case reports in the literature from 2005 to 2022\(^2\). Eleven further cases have been identified in the National Inpatient Sample Database of TAD cases amongst all hospital admissions in adults with repaired TOF from 2000 to 2014\(^3\). While the true incidence for TAD in post-TOF repair remains unknown, an estimated figure sits at approximately 6 in 10,000.

Case Report

A 55-year-old man, with a background of essential hypertension, depression, smoking, and stress, underwent a TOF repair (at the age of 3 years). He had a secondary operation (at the age of 54 years) to repair a residual ventricular septal defect (VSD) alongside an aortic valve replacement (Edwards Lifesciences INSPIRIS RESILIA valve) for regurgitation and relief of
a right ventricular outflow tract obstruction, from which he recovered well. Although a
dilated root of 52mm was noted pre-operatively, dissection flaps were initially absent and
intervention was deemed unnecessary as per the Adult Congenital Heart Disease MDT.

Investigations

An incidental finding of a large aortic root was found on a routine yearly follow-up
transthoracic echocardiogram (TTE). The ejection fraction was noted at 41%. He had mild
left ventricular hypertrophy with mildly reduced left ventricular systolic function and
regional wall motion abnormality. The aortic root was dilated at 7cm.

A subsequent CT aortogram confirmed the presence of a subacute proximal aortic dissection,
classified as a limited DeBakey Type II, with dissection flaps particularly close to the left
mainstem artery. Close to the posterior aspect of the sternal body, the ascending aorta
measured 7.8cm x 7.2cm. The patient was otherwise asymptomatic and was managed with
oral antihypertensives to optimise haemodynamic parameters. A further CT angiogram
confirmed an unobstructed left main stem and a compromised ostium, although a separate
small conal branch arising from the right-sided sinus was no longer identifiable.

Management

Cardiopulmonary bypass (CPB) consisted of left femoral-to-femoral cannulation and an
additional superior vena cava cannula. Thoracic cavity access required a repeat third
sternotomy. The dissection was repaired with resection of the dissected aorta, sandwich repair
of the base of the aortic root, repair of the left main stem coronary artery, and an interposition
tube graft (32mm Gel Weave). Due to post-operative right ventricular dysfunction his chest
wound was stented and closed three days later. A post-operative TTE revealed an ascending aorta of normal diameter and a moderately to severely dilated right ventricle with severely reduced systolic function. The aortic annulus measured 2.7x2.4cm and no aortic insufficiency was detected. A small VSD was noted.

Post-operative complications included arrhythmias, sepsis, acute renal failure, right ventricular failure, and stroke. He required pharmacological and electrical cardioversion for AF associated with runs of ventricular tachycardia and haemodynamic instability. Varied antimicrobial therapy was required for recurrent episodes of pyrexia and rising inflammatory markers of six weeks duration. Sputum cultures were positive for *Escherichia coli* and repeat bronchoalveolar lavage was positive for *Pseudomonas aeruginosa.*

Unfortunately, he was found to have a subacute left parieto-occipital stroke thought to be embolic in nature, with a small subarachnoid haemorrhage and a further small right occipital infarct on repeat imaging. The patient recovered well with almost no residual neurological deficit. He had a mildly raised C-reactive protein of 13 and was in rate-controlled AF at ongoing 6 monthly follow-up.

Discussion

Of the seven cases reported, five describe an acute presentation of an ascending aorta dissection on a background of TOF, whilst only two describe a subacute presentation. Ascending aorta dimensions ranged from 5.5cm to 9.3cm. Four cases were of males in their thirties, two in their sixties, and one 18-year-old (with 22q11 deletion syndrome). Whilst dissection remains a rare phenomenon, dilatation of the aortic root following repair of TOF is well reported in an enlarging population of patients. Since this is usually asymptomatic
routine follow-up is therefore crucial. If permissible, valve-sparing operations are favoured for this cohort of patients due to advantages such as anticoagulation avoidance and durability. Although an AVR may predispose to aortic dissection, a pre-existing dilated aortic root suggests that TOF aortopathy was the most likely contributing disease process. Cardiovascular risk factors such as uncontrollable hypertension, obesity, as well as underlying anxiety were likely contributing factors to TOF aortopathy in this case.

Conclusion

Continued surveillance of patients with TOF repair is crucial to identifying those with further aortic complications. Pathological aortic features present in this patient cohort seem to form the basis of dilated aortic pathologies post-TOF repair. The subject provided informed written consent for the publication of the study data; IRB approval was not required.
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