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REVIEW ARTICLE

Coronary insufficiency in children: Review of literature and report of a rare case with unknown aetiology

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Abstract Myocardial ischemia due to coronary artery disease is a rare condition in children. The reported causes of this condition include vasculitis; commonest being Kawasaki' disease, premature atherosclerosis due to familial dyslipidemias, congenital coronary artery anomalies and post-operative complications of procedures requiring coronary artery re-implantation in children, e.g. arterial switch procedure and Ross procedure. Allograft arteriopathy after heart transplantation is a more recent addition to this list (Mavroudis et al., 1996). Surgical procedures required for the treatment of coronary artery disease in children include, but are not limited to coronary artery re-implantation, re-routing, augmentation of the ostium, and coronary artery bypass grafting (CABG). We present our experience of a rare case of triple vessel coronary artery disease of unknown aetiology in a child, treated with coronary artery bypass grafting procedure.

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1. Case report

A 12 year girl admitted through emergency room due to chest pain. She gave the history of four days, sudden onset chest pain and shortness of breath, for which she was admitted at a local hospital. She has no family history of heart disease. And on admission to the local hospital, the electrocardiogram (ECG), showed ST elevation in lead I, II, aVL, V4–V6 and the R wave has poor progression from V2 up to V6.

Her initial work-up showed that The Creatine Kinase (CK), level was 2714 U/L (units per liter), the CK–MB, were 343 U/L

and the troponin I were 110.16 U/L. All were suggestive of a severe myocardial injury and for which she was suspected of having severe viral myocarditis. However, the viral screen was negative. The other blood tests revealed slightly raised cholesterol (cholesterol 4.08, TG 1.86, LDL 3.35 and the HDL 0.58).

On admission to our unit, the troponin level dropped to 38.52 U/L (below 0.012) and the CK–MB, 213 U/L. However, she stayed complaining of chest pain and the echocardiography revealed depressed left ventricle systolic function and apical and mid segments hypokinesia of left ventricle. There were normal basal segment and normal right ventricle function with small pericardial effusion. The initial findings were suggestive of myopericarditis. The patient underwent coronary angiography just to exclude an ischemic insult. However, to our surprise the angiogram were of totally occluded left anterior descending artery (LAD), and totally occluded distal circumflex artery (CX) and severely stenotic right coronary artery (RCA). As shown in the Figs. 1 and 2.

She remained symptomatic and haemodynamically unstable, requiring ion-tropic support. Therefore, and in-view of the angio-finding, she underwent emergency coronary bypass surgery, using conventional general anesthesia, simultaneous arterial and venous pressure monitoring, and temperature monitoring. Since the internal mammary were of small caliber, and the critical condition on ionotropic support, saphenous vein were used as a conduit. Myocardial biopsy was taken from the right and left ventricle, which revealed myocardial cell edema and no suggestive evidence of viral cardiomyopathy.

The surgery was uneventful and the patient had smooth wean from cardiopulmonary bypass. The post-operative period was smooth and the patient was discharged home 2 weeks after surgery.

At 1 year follow up the patient is doing well and the cardiac echocardiography showed significant improvement in the left

Figure 1 Angio-illustration of totally occluded left anterior descending artery and circumflex artery.

Figure 2 Angio-illustration of severely stenotic lesion of the distal right coronary artery.

ventricle contractility and function with an ejection fraction of more than 50%.

2. Discussion

Myocardial ischemia in children leads to infarction or ischemic dysfunction and presents with symptoms of heart failure is rarely encountered. In contrast to adult population, the aetiology is varied (Mavroudis et al., 1996) and includes congenital anomalies of the origin and course of coronary arteries, in-born errors of metabolism e.g. homozygous and heterozygous hypercholesterolemia, vascular diseases like Kawasaki's vasculitis and iatrogenic problems of coronary trunks as a result of complex aortic root surgery. In the context of this rare case report, we will present a summary of literature review of these conditions with reference to their clinical course, the treatment options available and the outcome of surgical interventions.

2.1. Kawasaki disease

Kawasaki disease (KD) or mucocutaneous lymph node syndrome is an acute vasculitis of unknown aetiology, which may lead to myocardial ischemia and infarction due to aneurysm formation, thrombo-embolism and coronary artery stenosis (Hwong et al., 2004). It is the commonest cause of acquired heart disease in childhood in the Asian and western world (Hwong et al., 2004; Coskun et al., 2006). Most of the cases have, however, been reported from Japan (Fulton and Newburger, 2000). Up to 20% of untreated patients develop coronary artery abnormalities (Tsuda et al., 2004). However, early treatment with intravenous gamma-globulin may reduce this risk significantly (Coskun et al., 2006; Ishiwata et al., 1992). CABG is rarely required in KD and is only indicated in patients who demonstrate myocardial ischemia in the presence of aneurysms or stenotic lesions (Hwong et al., 2004; Wilson et al., 2004; Yamauchi et al., 2004). Early surgery (within 3–4 years) after acute KD has been demonstrated to result in better preservation of myocardial function and reduced number of major cardiac events (Wilson et al., 2004). In various series, attractive outcome has been shown with the use of single or bilateral mammary arteries, gastro-epiploic artery and saphenous vein grafts (Yamauchi et al., 2004; Gotteiner et al., 2002; Inoue et al., 2001; Kitamura et al., 2009; Kawachi et al., 1991).

2.2. Familial dyslipidemia

Children with familial dyslipidemia have shown a strong correlation between endothelium–platelet activation markers and pre-mature atherosclerosis and are at high risk of cardiovascular events (Kameda et al., 2001). Homozygous hypercholesterolemia is associated with highest risk (Guardamagna et al., 2009) and may also produce valvular and supra-valvular aortic stenosis (Guardamagna et al., 2009). Close surveillance regarding the progression of the development of coronary artery and aortic valve disease is recommended (Marais, 2004). Primary management is aimed at life style modifications which should be instituted as early as possible with limitation of total and saturated fat intake. Lipid lowering pharmacotherapy is recommended after 10 years of age, if low-density lipoprotein (LDL) levels remain very high despite dietary restrictions (Kolansky et al., 2008). LDL-apheresis may be needed in resis-

tant case in addition to drug therapy (Luggetti et al., 2007). A recent prospective study has shown long term clinical benefit of combination therapy with statins and LDL-apheresis in delaying the progression of atherosclerosis and preventing major cardiac events (Mehta et al., 2009). However, in a small number of patients, coronary artery bypass grafting may be required. Compared to the saphenous vein grafts alone, addition of one mammary artery graft has shown far superior results in terms of long term patency, survival and freedom from major cardiac events as compared. (Masaki et al., 2005; Kawasuji et al., 2000). Additional benefit of multiple arterial grafts has not been demonstrated. We could not use the arterial grafts in our patient, due to the high dose of immunoglobulin the patient requiring, the small caliber of the mammary artery and the severely impaired LV function.

3. Congenital coronary artery anomalies

Congenital coronary artery anomalies are a known cause of myocardial ischemia, contractile dysfunction and sudden death. Commonest conditions are anomalous origin from aorta (Takahashi et al., 1993) or from pulmonary artery (Youldelman et al., 2009), abnormal orifice (Alsoufi et al., 2008; Gabauer et al., 2008), intramural course or inter-arterial course (between aorta and pulmonary artery) (Gabauer et al., 2008). Various surgical options available which include augmentation of the coronary orifice (Takahashi et al., 1993), re-implantation with and without extension of coronary arteries (Youldelman et al., 2009), pulmonary artery translocation (Gabauer et al., 2008) and coronary artery bypass grafting (Takahashi et al., 1993; Youldelman et al., 2009).

4. Iatrogenic coronary artery anomalies

Complex surgical procedures in children including arterial switch procedure (Gulati et al., 2007; Bergoënd et al., 2008) and Ross procedure (Miyamoto et al., 2008; Da Costa et al., 2009) which involve surgical re-implantation of main coronary arteries carry an inherent risk of post-operative coronary problems. Early post-operative ischemia and myocardial dysfunction may be caused by technical problems like kinking of coronaries, intimal damage, coronary dissection and anastomotic stenosis. Late coronary ischemia may be caused by stenosis of coronary ostia due to fibrosis or accelerated atherosclerosis resulting from intra-operative intimal damage. Re-operation would be required if significant myocardial ischemia is demonstrated after such procedures. Patch arterioplasty of the origin of the main coronary trunk or coronary artery bypass grafting are available options; the former procedure has been shown to produce superior results (Gulati et al., 2007).

5. Conclusion

Myocardial ischemia in children requiring CABG is very rare. We presented the case of a 12 year old female, who presented with severe myocardial ischemia and cardiac failure of acute onset. Our case is unique in that, no aetiology for the development of pre-mature coronary artery disease was discovered despite thorough histo-pathological and biochemical work-

up. However, she underwent CABG and at 1 year follow up, she is doing well.

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