A Case of Isolated Congenital Left Ventricular Diverticulum with Acute Myocarditis

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Isolated congenital left ventricular diverticulum is a rare congenital heart malformation with an incidence ranging from 0.05% to 0.4%[1]. This lesion was defined by a protrusion of the three-layer left ventricular free walls, and was first described in 1838[1]. Multiple clinical phenotypes were presented in literature, such as chest pain, arrhythmia, systemic embolisation, valve regurgitation, ventricular wall rupture and sudden death. However, most patients would be asymptomatic and detection usually occurred by accident, often during echocardiography. Cases of left ventricular diverticulum accompanying myocarditis were rarely reported. A nine-year-old boy diagnosed with left ventricular diverticulum accompanying acute myocarditis underwent surgical management and was followed up for two years after surgery in our hospital. We proposed that conventional surgical treatments are not suitable for our patient during the period of acute myocarditis, except for heart transplant. A nine-year-old boy who had been suffering from palpitation and edema for half a month was referred to our hospital in 2010. In physical examination a mild systolic heart murmur was auscultated at the apex and the liver was palpated 4 cm below the subcostal margin. Echocardiography revealed four enlarged cardiac chambers and a diverticulum, 55 mm in diameter and 70 mm in length, without other congenital cardiac malformations. Mild mitral valve regurgitation was detected by color Doppler flow imaging. The left ventricular ejection fraction (EF) and fractional shortening (FS), which mirrored cardiac function, was 33% and 16%, respectively. Magnetic resonance imaging confirmed that the diverticulum with a narrow short neck was connected to the left ventricle. The electrocardiogram showed a few ventricular premature beats. No evidence showed that the patient had a recent viral infection, particularly influenza.

The surgical procedure was performed. The diverticulum was observed to be full of thrombi upon incision. After cleaning the thrombi, the opening (8 mm × 8 mm) to the left ventricle was sewn and closed by a Teflon patch with 4/0 prolene through the incision. A myocardial specimen was harvested and sent for pathological examination. The examination revealed an active lymphocytic myocarditis with cell necrosis and thrombi. Upon surgical manipulation, the echocardiograph indicated exclusion of the diverticulum with only mild mitral valve regurgitation.

The postoperative course was uneventful and vasoactive drugs, like dopamine (6 μg/kg·min^{-1}) and adrenaline (0.05 to 0.1μg/kg·min^{-1}), were administrated for five days. The size of the four cardiac chambers did not change compared with their size before surgery during the two-year follow-up period; whereas more mitral and tricuspid valve regurgitations were observed. Cardiac function was significantly weaker (EF: 28%, FS: 13%) (Table 1 and Fig. 1). The patient suffered from chronic heart failure and had to

Table 1: Echocardiography results measured before and after surgery*

Object	Before	Post procedure				
		15 days	30 days	6 months	12 months	24months
Left atrium (mm)	26	29	24	24	28	42
Left ventricle (mm)	54	54	54	54	54	53
Right atrium (mm)	39	40	39	38	42	45
Right ventricle (mm)	37	39	38	37	39	40
Left Ventricular Fraction Shorten	22	22	23	16	15	13
Left Ventricular Ejection Fraction	42	43	46	33	30	28
Mitral Valve Regurgitation	mild	mild	mild	middle	sever	sever
Tricuspid Valve Regurgitation	mild	mild	mild	middle	sever	sever

*All the data was measured at the end-diastole dimension in the text and the table

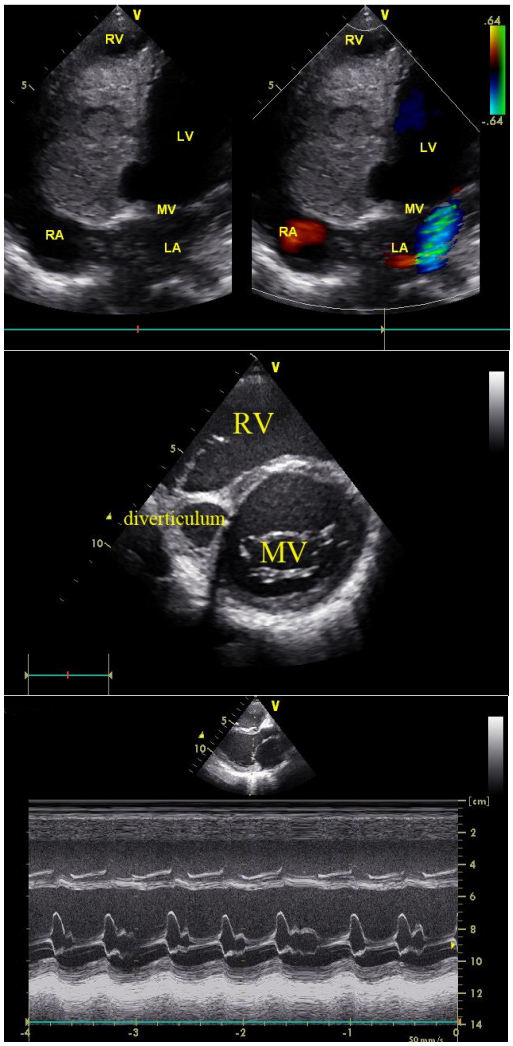

Fig. 1: (A) Color Doppler echo and B-echo showing the left ventricular diverticulum full of thrombi
(B) B-echo 2 and M-echo two years after the surgery showing the resident diverticulum and low heart function

receive digoxin interval treatment because of weaker cardiac function.

Congenital ventricular diverticulum is a rare malformation which is always accompanied by other cardiac malformations, as well as vascular or thoraco-abdominal abnormalities. This malformation can be differentiated from aneurysm, which is a consequence of coronary heart disease, cardiac trauma or hypertrophic cardiomyopathy according to the connection of the neck to the left ventricle. However, certain cases of aneurysm caused by viral myocarditis are similar to congenital diverticulum and are characterised by a narrow connection. These aneurysm cases are called micro-diverticula for their small size[3]. Endocardial biopsy is the golden standard for diagnosing viral myocarditis. In addition, hepatitis C and influenza A viruses are more commonly found in patients with aneurysm and myocarditis[4]. Our patient was subjected to pathological examination, which revealed acute myocarditis symptoms without any virus detected. Therefore, our patient was diagnosed with congenital cardiac diverticulum accompanying acute myocarditis based on morphological and clinical features.

Considering that only a few cases of such incidence have been reported or published in literature, controversies on the treatment and prognosis of congenital cardiac diverticulum accompanying acute myocarditis are ongoing[1]. Frustaci et al (1992) proposed that these combined lesions should not be subjected to surgical manipulation according to the mid-time follow-up[6]. However, majority of patients died because the diverticulum ruptured; therefore, repair was necessary once lesions were diagnosed[7]. Makoto (2003) performed a successful surgical management on a patient with left ventricular aneurysm accompanying acute myocarditis[8]. Considering the size of the diverticulum and the risk of rupture, our patient underwent surgical management. Unfortunately, the exacerbated cardiac function during the two-year follow-up period after surgery implied that conventional surgical procedures might not be suitable for our patient during the period of acute myocarditis, except for heart transplant.

Key words: Myocarditis; Cardiac Diverticulum; Ventricular Aneurysm

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Feminizing Adrenal Tumor: A Rare Presentation

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Malignant adrenocortical tumor (ACT), a rare entity with an incidence of 1-2/million, is uncommon in pediatric population[1]. These tumors are functional with varied endocrine syndromes, most frequent being rapidly progressing Cushing syndrome with/out virilization. In males estrogen-secreting tumors lead to gynecomastia and are invariably malignant[2]. The modality of treatment is surgical resection of tumor by laparoscopy or open laparotomy followed by chemotherapy in selected cases based on new Union International Contre Cancer (UICC) staging by WHO. Laparoscopic adrenalectomy as safe choice of surgical approach is being extensively described in recent literature. The prognosis depends on the tumor stage and five-year survival in different series ranged between 16 and 38%[2]. We report on a feminizing adrenal tumor in a six year old male child who presented with bilateral gynecomastia for its rarity.

A six year old male child with normal developmental history presented with bilateral breast engorgement since six months with no history suggestive of development of secondary sexual characters. His body mass index was 17.28 (>85th centile) with no mass palpable on per abdominal examination. SMR for testes, pubic hair and penis was prepubescent and breast maturity scale corresponded to stage 3 thelarche (Fig. 1A). Blood pressure, visual field and other systems were normal. His bone age was 12 years. Hemogram, liver and renal functions were normal. Contrast enhanced computed tomogram of abdomen showed well defined hypodense non-homogenous contrast enhanced right sided adrenal mass without calcification or fat density within. Well defined plane at upper pole of kidney was delineated. Blood vessels were normal (Fig. 2). Hormonal assay showed normal levels of serum FSH, LH, cortisol and testosterone with elevated levels of estradiol (28.94 pg/ml, ref range <10) and 17(OH)progesterone (2.2ng/ml, ref range <1.5) suggesting feminization. Tumor was resected by laparoscopy. Gross adrenal mass was partially capsulated measuring 4×3.5×2.5 cms, weighing 30gms with congested blood vessels and hemorrhagic areas on cut section. Histopathology showed tumor cells with pleomorphic nuclei with irregularly distributed coarse chromatin with>one mitotic figure/10 hpf. Capsule and vascular

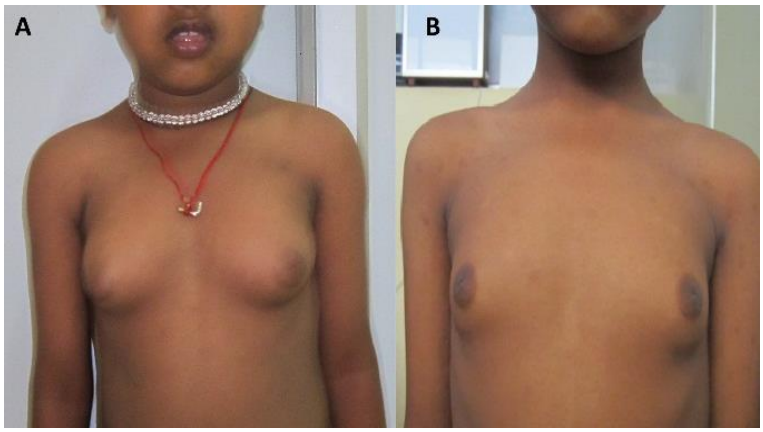

Fig. 1A & B: Clinical photograph of gynecomastia before (left) and after (right) surgery

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