CASE REPORT

Recurrent Embolic Stoke In Children Due To Giant Congenital Cardiac Diverticulum

Hayder Kadhum Hassoun*, Ali Hussein AL-Moussawi, Zuhair Salih Allebban*****

ABSTRACT:

One of the great challenges we face in clinical neurology practice is stroke among children, which is considered to be among the ten top causes of death in the United States. The cardiac diseases are the most common causes of strokes among children; But a recurrent embolic stroke among children is rarely reported to be due to giant cardiac diverticulum originating from the left cardiac chamber as presented in this paper.

CASE REPORT:

A 9-year old Iraqi right handed girl enjoying good health arrived in the emergency unit in May 2009 with a sudden attack of partial seizure as tonic-clonic movement affected the right side of her body for five minutes, which subsided spontaneously upon arrival to the hospital, followed by right sided weakness, facial asymmetry and complete loss of speech. She had two previous similar attacks of partial convulsive movement in 2005 and 2008, the latter being followed by weakness in her left upper and lower limbs with complete recovery over one week. Unfortunately and because of economic factors and lack of facilities, she did not investigated and treated in proper way. She has been admitted to the neurology ward for further evaluation. Her general examination two days after admission revealed an alert conscious and comprehending state, with expressive aphasia and controlled sphincters. She was febrile, with a pulse rate of 90/min, RR 16/min and BP of 120/70. Her medical examination was unremarkable. Cranial nerve examination revealed clear fundi with normal size pupils reacting to light with normal ocular motility. There is upper right motor facial palsy without affection of the rest of the cranial nerve. There were no signs of meningeal irritation. Upper and lower limb examination revealed dense right sided hemiplegia with impaired superficial modality sensations. The patient cannot walk to check her gait.

Detailed investigation of blood for complete blood count, sugar, renal function, liver function, PT, PTT, clotting time, and bleeding time were all normal. Her ECG was normal. CT scan showed medium size cerebral infarct in distribution of left middle cerebral artery (figure1). Her chest X-ray (see figure 2) revealed an interesting view of a large cystic lesion with rimmed calcification attached and protruded from the left cardiac margin. Detailed transthoracic echocardiography was done with color flow study revealing clear evidence of giant cardiac diverticulum attached to the apex of the left ventricle with 1.5 cm neck diameter (figure 3). The cyst and it's attachment to the left cardiac chamber was observed by CT scan of the chest (figure 4). Her MRI and MRA of the brain one week after admission revealed evidence of a medium size infarction at the left perisylvian, distributed in the left middle cerebral artery (figure 5).

The child was treated as an embolic stroke and discharged with partial recovery on antiepileptic drug , small dose warfarin with close monitoring of INR and an intensive course of physiotherapy. We sent the patient for further treatment at the cardiovascular center and arranged for surgical intervention. Cardiac surgery was done few months later ; a giant diverticulum (4 cm diameter) was removed from apical part of left ventricle .

*College of MedicineUniversity of Kufa, Najaf, Iraq Al-Sadder Teaching Hospital –AL-Najaf /Iraq.

**Al-Sadder teaching hospital –AL-Najaf /Iraq.

*** Department of Internal Medicine
Section of Cardiology St John Hospital and Medical Center.

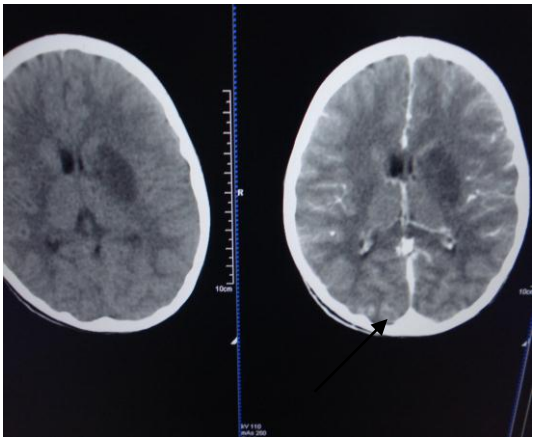

Figure 1: left medium size infarction in distribution of MCA(middle cerebral artery)

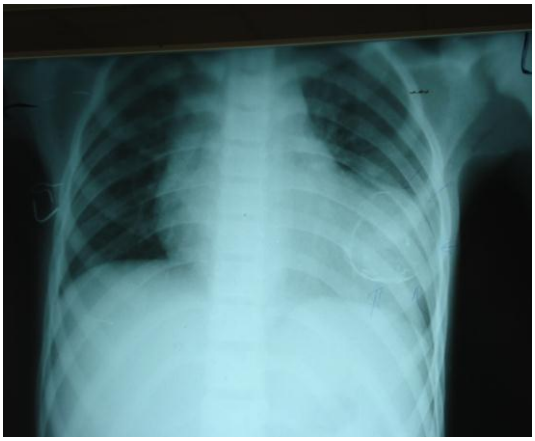

Figure 2: CXR shows the calcified cystic lesion that emerged from lateral cardiac border(arrows)

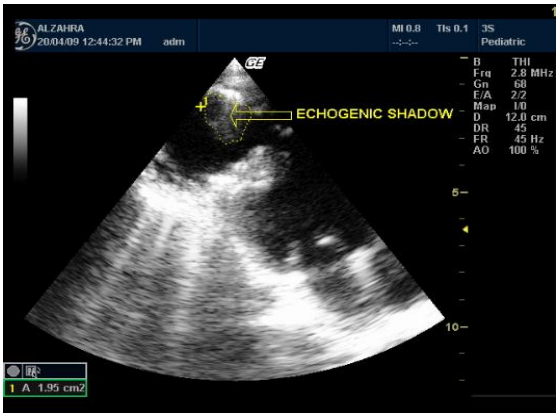

Figure 3: Echocardiograms shows large cystic diverticulum connecting with left cardiac chamberand thrombus formation clearly demonstrated(arrows).

DISCUSSION :

Stroke in children is a significant diagnostic challenge in our daily practice. It is among the top ten causes of death among children in the United States[1]. Recent data suggests that stroke in children is more frequent than previously recognized with multiple etiological and pathological factors. The incidence of stroke in children between the ages of 1 and 14 (from a pool of 100,000 children per year)

varies between 1.29 in a Baltimore Washington study to 27.1 in a Saudi Arabia study [1,4]. Underlying cardiac disease is an important etiological factor causing ischemic stroke in children [3,4]. The most frequent causes of stroke at the beginning of life is cardiac embolism for ischemic stroke , it is extremely rare to be due to congenital cardiac diverticulum (CCD) [5,6]. CCD is a rare congenital cardiac malformation which is either fibrous or muscular arising from either or both cardiac chambers as an isolated anomaly or associated with other congenital cardiac and/or somatic defect [5,10]. The association of CCD with major midline supra-umbilical abdominal defects, defect of lower part of sternum, deficiency of the lower anterior diaphragm, defect of diaphragmatic pericardium and congenital heart malformation constitute the Cantrell syndrome [7,11] . CCD was first described by O'Bryan in 1838. The incidence of left ventricular diverticulum has been reported as 0.05% of all congenital heart disease.[12]

It is important to differentiate between congenital diverticulum (muscular type) from congenital aneurysm (fibrous type) as these have been used interchangeably in the past. The wall of true CCD is formed by three cardiac layers that contracts normally and synchronously, whereas congenital aneurysm is generally a fibrous sacular sac with paradoxical contraction.[12,14] The other important differentiating feature is the neck of connection to cardiac chambers. The CCD is often tubular or sacular with a narrow orifice whereas aneurysm has a wide mouth which tends to occur near atria-ventricular valve apparatus[15]. The neck is defined as narrow if its diameter is less than 40% of the maximum diameter of the sac.[16] According to the above criteria; our case is an interesting case of an isolated apical giant left CCD in a 9 year old child who presented with recurrent embolic stroke and partial epilepsy. The neck diameter was 1 cm while the maximum diameter of the sac was 4cm. There are less than 100 reported cases of cardiac diverticulue world wide; but it is extremely rare to have giant left cardiac chamber diverticulum presented with recurrent embolic stroke with partial epilepsy in a young child [15].

CCD have a wide spectrum of presentation ranging from an acute rupture due to contraction of diverticulum against a closed orifice to life threatening arrhythmias to being completely asymptomatic [8,9,12,13]. A subgroup of patients may develop embolism, sudden death or severe valvular regurgitation [12,15,16,17]. It has been mentioned in literature that cardiac muscle is often recognized in infants and only rarely as an incidental finding in older children or adults[9]. The best treatment for CCD depends on the clinical setup of the case and the site of CD. Surgical resection is indicated as early as possible because of the risk and life threatening complication.[12,18,19] Surgery cannot be done if diverticulum is in close proximity to mitral valve apparatus.[15] We did base line INR. Warfarin was given two weeks after the onset of the present stroke. Further embolization is likely as long as we have the defect with stasis of blood inside with thrombus formation which was clearly shown in echocardiogram (see figure 3A and 3B). the surgery was performed in cardiac surgery center and the result confirmed our initial diagnosis.

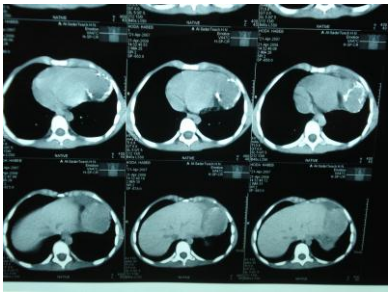

Figure 4: CT-chest shows clearly the calcified large cardiac diverticulum attached to left cardiac chamber

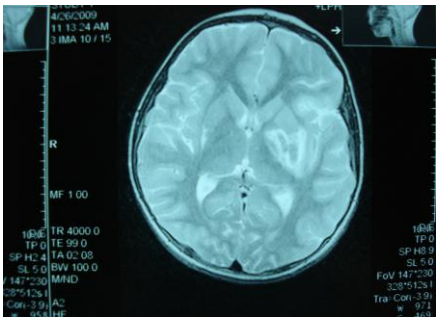

Figure 5: T2 MRI brain shows clearly site and size of infarction in left middle cerebral artery distribution.

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