ID: JCDR/2012/4039:2331

Case Report

Pathology Section

Congenital-Left Atrial Appendage Aneurysm in an 8-Year-Old Girl

JAYASHANKAR MARLA, NISHA JAYASHANKAR MARLA, GURURAJ TANTRI, CHANDRASHEKAR JAYAPRAKASH

ABSTRACT

Congenital aneurysm of the Left Atrial Appendage (LAA) is extremely rare because after the first case no was described by Diamond in 1960, only 50 cases have been reported in the literature. Congenital aneurysm of the LAA is rarely diagnosed during childhood. Generally it manifests during the second or third decades of life. We are reporting here a case of congenital left atrial appendage aneurysm in an 8 year old girl who presented with breathlessness on exertion which was present since 6 months. Pre-operatively, the diagnosis of aneurysm was made by chest radiography, echocardiography and magnetic resonance imaging. The surgical findings demonstrated a kidney shaped saccular aneurysm. The microscopic findings showed myocardial layers in variable numbers. Following an aneurysmectomy, the patient is now asymptomatic and in the sinus rhythm.

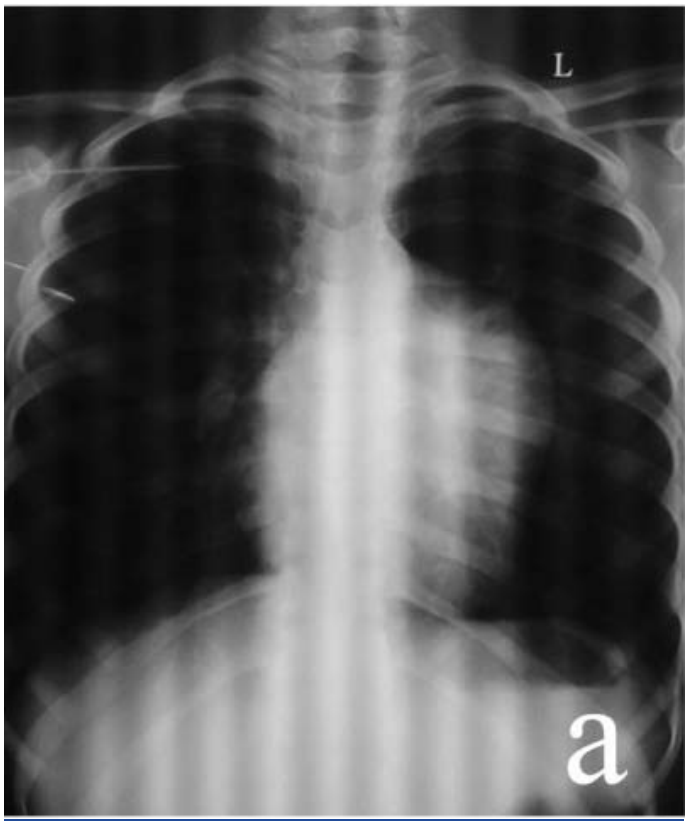
Key Words: Aneurysm, Congenital, Left Atrial Appendage (LAA)

INTRODUCTION

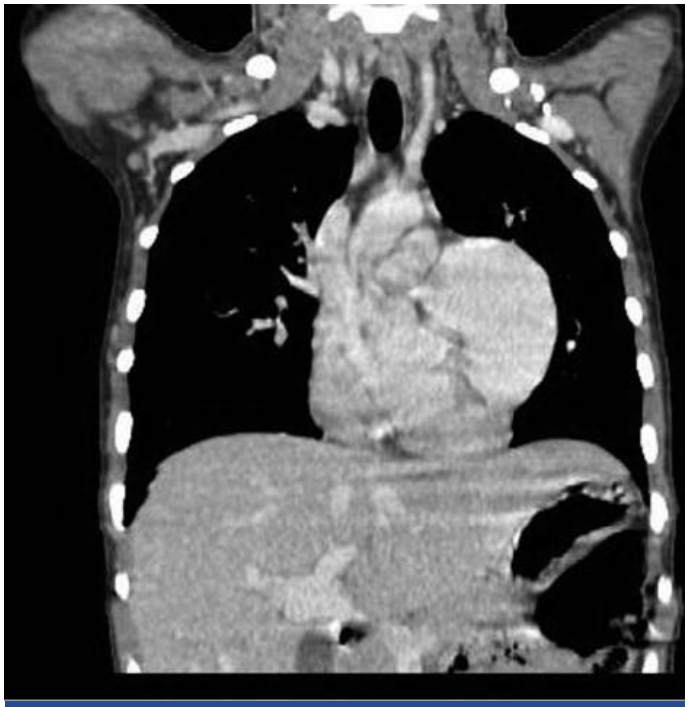
Congenital aneurismal dilatation of the Left Atrial Appendage (LAA) is an uncommon cardiac anomaly. Despite the fact that aneurysm of the LAA is thought to be congenital in nature; most patients have few problems early in life. It is rarely diagnosed during childhood. Generally, it manifests during the second or third decades of life. The early age at presentation of our case and the rarity of the condition made us to present it. The origin of a congenital aneurysm is unknown; some authors have attributed it to dysplasia of the musculi pectinati. The patients who are affected by aneurysms of the LAA are at a risk of significant morbidity and mortality. Therefore, an early diagnosis and a prompt surgical excision are essential, in order to eliminate the risk of complications. Aneurysmectomy is curative, with an excellent prognosis [1, 2, 3].

CASE HISTORY

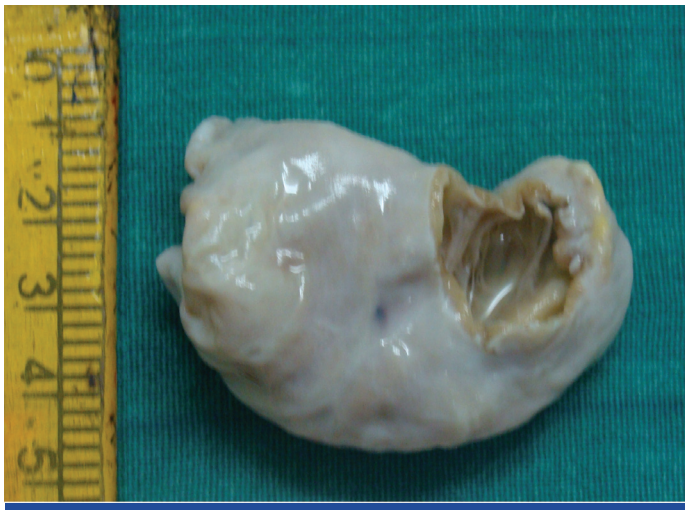
A 8-year old girl was referred to the Cardiothoracic Department with complaints of breathlessness on minimal exertion which was there since 6 months and a precordial bulge which was there since 1 year. The physical examination revealed no abnormalities other than the subtle pre-cordial bulge. A previous workup which was done by a paediatrician and a cardiologist revealed an aneurysm of the LAA. The Electrocardiogram (ECG) showed a normal sinus rhythm. Trans thoracic echocardiography demonstrated normal findings, except for an impressive dilatation of the LAA. Chest radiographs showed cardiomegaly with prominent convexity of the left basal aspect of the cardiac silhouette [Table/Fig-1(a)] and magnetic resonance imaging confirmed the left atrial appendage aneurysm [Table/Fig-1(b)]. The left atrial appendage was approached surgically through a median sternotomy and an under cardiopulmonary bypass. The pericardium was found to be intact. The surgical findings demonstrated a giant, kidney shaped, saccular aneurysm which was indenting the right ventricular out flow tract. The aneurysm was resected and it was closed with a continuous over and over suture in two layers. The gross specimen consisted of a sac like structure which measured 6×5×3cm. The external surface was pale white, with a glistening appearance [Table/Fig- 2(a)]. On cutting the cystic structure, the inner aspect revealed pectinati muscles [Table/Fig- 2(b)]. No thrombus was noted. The microscopic examination of the sac wall showed the myocardium with variable numbers of myocardial fibres [Table/Fig-3(a)]. Masson trichrome staining showed mild fibrosis in the subepicardial layer [Table/Fig- 3(b)]. The post-operative course was uncomplicated, and now the patient is asymptomatic and in sinus rhythm.



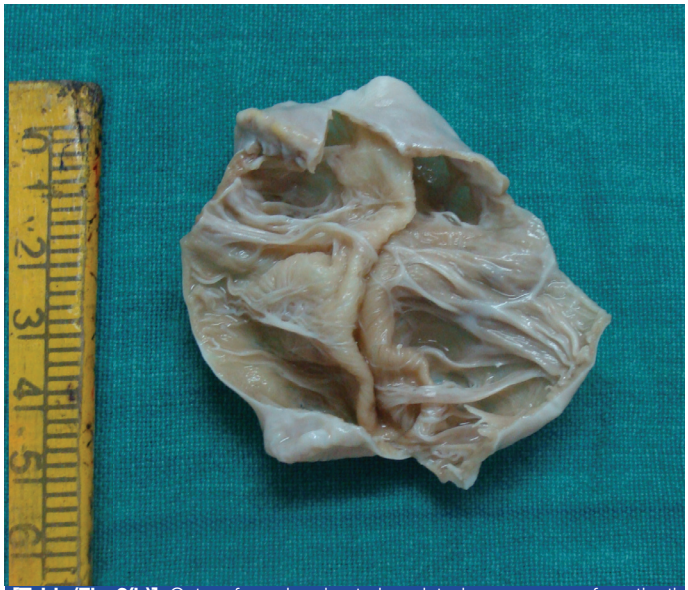
[Table/Fig-1(a)]: Chest radiography showing cardiomegaly with prominent convexity of the left Basal aspect of cardiac silhouette



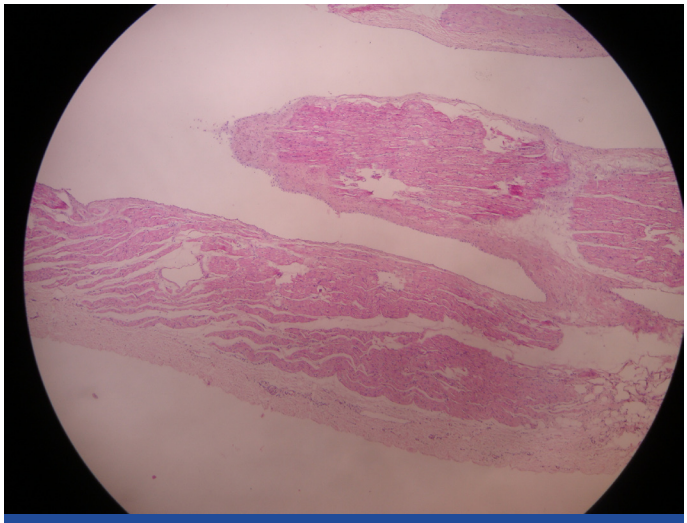
[Table/Fig-1(b)]: Magnetic resonance imaging showing left atrial appendage aneurysm



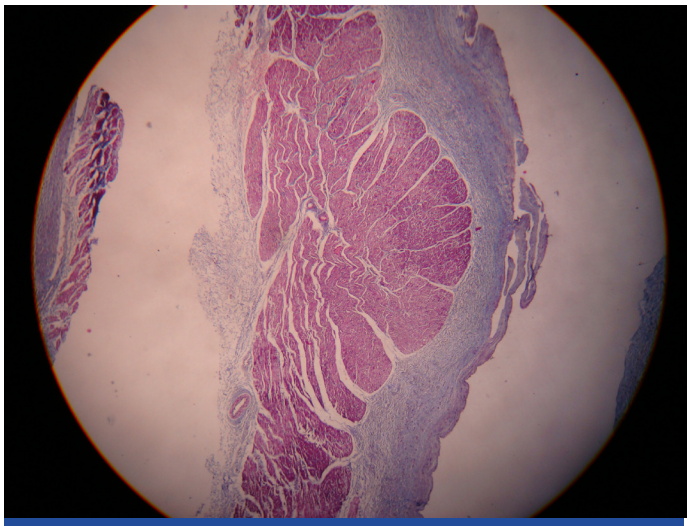
[Table/Fig-2(a)]: Gross specimen showing saccular structure with glistening outer surface



[Table/Fig-2(b)]: Cut surface showing trabeculated appearance of pectinati muscle



[Table/Fig-3(a)]: Microscopy of sac wall showing myocardial layers in variable number. (H&E, x200).



[Table/Fig-3(b)]: Masson trichrome stain showing mild fibrosis in subepicardial layer. (MT, x200).

DISCUSSION

An isolated aneurysm of the left or right atrium is a rare congenital abnormality which was first described by Semans and Taussig in 1938. It represents a diagnostic dilemma in patients with cardiomegaly. An aneurysm of the LAA is extremely rare because after the first case was described by Diamond in 1960, only 50 cases have been reported in the literature [1,2,3].

The LAA is derived from the left primary atrium which forms during the fourth week of the embryonic development. It has developmental, ultra structural and physiological characteristics which are distinct from those of the left atrium proper. The LAA is a long, tubular, hook shaped structure which has a narrow junction with a venous component of the atrium proper. Both the left and right appendages are trabeculated, with muscle bars largely running parallel to each other, giving a comb like appearance and hence they are termed as the pectinati muscles [4,5]. Victor and Nayak postulated that the cause of the aneurysm may be congenital dysplasia of the pectinati muscles. An LAA aneurysm can occur in two forms, one being the intra pericardial type with an intact pericardium as was seen in the case which is under discussion and the other being the extra pericardial type which occurs in association with pericardial defects[3,6]. The congenital LAA aneurysm arises from a weak appendage aneurysm wall, with no other associated pathologies such as mitral valve disease, tuberculosis, syphilis, or

pericardial defects, and it must fulfil the following criteria; presence of LA of normal characteristics, a direct continuity of blood flow between the LA and the appendage, the absence of pericardial defects, and the presence of the characteristics of a normal LAA in terms of the anatomical pathology [2,6].

In a majority of the cases (75%), the main symptom which leads to its diagnosis is recurrent or continuous supraventricular arrhythmias. Adults with the LAA aneurysm may present with shortness of breath, palpitation due to atrial tachyarrhythmias, embolic complications like stroke and even chest pain which is accompanied by a rise in the levels of cardiac enzymes. In infants and children, the aneurysm may be the cause of a cardiac arrest, an atrial fibrillation, or a heart failure and sometimes it can present with a cardiac tamponade [1,2,3]. The diagnosis of these aneurysms can be difficult. Physical examinations are non-contributory. In fact, the results of most of the cardiac examinations may be normal. ECGs may reveal arrhythmias in some patients. Chest radiographs always show a cardiac enlargement with a prominent convexity of the left upper border in the position of the left atrial appendage. The differential diagnosis which is made on the basis of the chest radiographs includes epicardial fat, a loculated effusion and a mediastinal mass. Transoesophageal Echocardiography (TOE) usually helps in the diagnosis of this condition. Intra operative TOE is a useful aid which helps in looking for a residual thrombus and in assessing the adequacy of the resection. In recent years, TOE and chest magnetic resonance imaging have proved to be useful in detecting and diagnosing this anomaly. A surgical treatment which is curative is usually recommended, even in asymptomatic patients [3,7].

Congenital aneurysmal dilatation of the left atrial appendage is rare, but it is a correctable lesion.

It represents a diagnostic dilemma in patients with cardiomegaly and it is commonly associated with supraventricular arrhythmias and life threatening systemic embolizations. The patients who are affected by aneurysms of the left atrial appendage are at a risk for significant morbidity and mortality, but with a prompt and low risk surgical resection, the prognosis is excellent [1,2,3,6,7].

REFERENCES

[1] Fuente A, Urchaga A, Sanchez R, Fernandez JL, Moriones I. Congenital aneurysm of the left atrial appendage . *AnnThor Surg*. 2008; 85: 2139-40.

[2] Munarriz A, Escribano E, Urchaga A, Olaz F, Beunza M, Fuente A, Cantabrana S.

[3] Congenital aneurysm of the left arial appendage. *European Journal of Echocardiography* .2008;9:152-54.

[4] Hammad AM, Abdel-Aziz O, Alshiek RGH, Wahby EA. Idiopathic giant aneurysm of the left atrial appendage. *IJTCVS* .2004; 20:186-88.

[5] Al- Saady NM, Obel OA, Camm AJ. The left atrial appendage: structure, function and role in thromboembolism. *Heart.* 1999; 82:547-54.

[6] Victor S, Nayak VM. Aneurysm of the left atrial appendage. *Tex Heart Inst J* 2001; 28:111-18.

[7] Pome G, Pelenghi S, Grassi M, Vignati G, Pellegrini A. Congenital intra-pericardial aneurysm of the left atrial appendage. *Ann Thor surg* 2000; 69:1569-71.

[8] Sawalha W, Badaineh Y, Samara Y. A giant left atrial appendage aneurysm: A case report. *JRMS*. 2005; 12:41-44.

[9] Shaheen F, Gojwari T, Ahmad N. Congenital aneurysm of the left atrial appendage presenting as a young stroke: CT and MR findings. *European Journal of Radiology Extra*. 2008; 68:e107-e09.

AUTHOR(S):

1. Dr. Jayashankar Marla
2. Dr. Nisha Jayashankar Marla
3. Dr. Gururaj Tantri
4. Dr. Chandrashekar Jayaprakash

PARTICULARS OF CONTRIBUTORS:

1. Consultant Cardiothoracic Surgeon, AJHRC.
2. Associate Professor, FMMC.
3. Consultant Cardiac Anaesthesiologist, AJHRC.
4. Professor, FMMC.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Nisha J Marla
Associate Professor, Department of Pathology
Father Muller Medical College
Kankanady -575004, Mangalore, Karnataka, India.
Phone: 9481148975
E-mail: nishajmarla@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Submission: **Jan 27, 2012**
Date of Peer Review: **Apr 09, 2012**
Date of Acceptance: **Jun 10, 2012**
Date of Publishing: **Aug 10, 2012**