Case Report

CONGENITAL GIANT ANEURYSM OF THE LEFT ATRIAL APPENDAGE: DIAGNOSIS AND MANAGEMENT

[1]PARVEEN AKHTAR; [2]SULTANA HABIB; [3]MUSHTAQUE HUSSAIN; [4]HABIBA TASNEEM; [5]AKHTAR HUSSAIN; [5]ISHTIAQ RASOOL; [5]KHAN SHAH ZAMAN.

SUMMARY

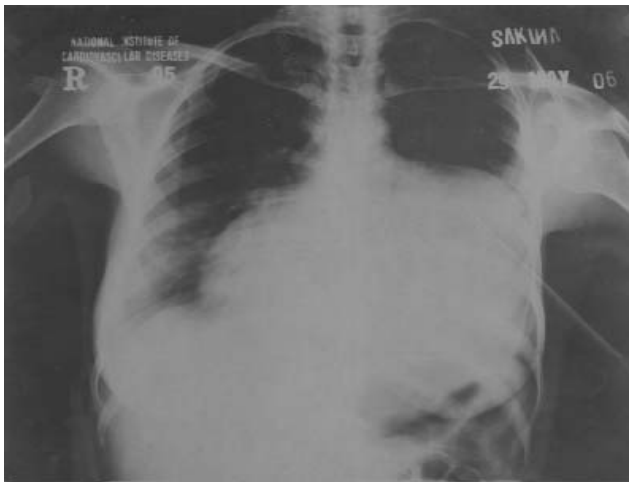
Left atrial (LA) aneurysm is a rare congenital abnormality[1] and associated with life-threatening complications.We report the natural history of Left atrial aneurysm which was complicated by supraventricular tachycardia (SVT) and stroke in early childhood and her condition deteriorate rapidly after development of progressive mitral regurgitation(MR) and heart failure and expired despite successful surgery.

Our purpose of this case report is to give the natural history of LA aneurysm and review it along its current literature, surgical strategy, and patient outcome.In literature there is a very limited description of such cases and their management.[1]

CASE HISTORY

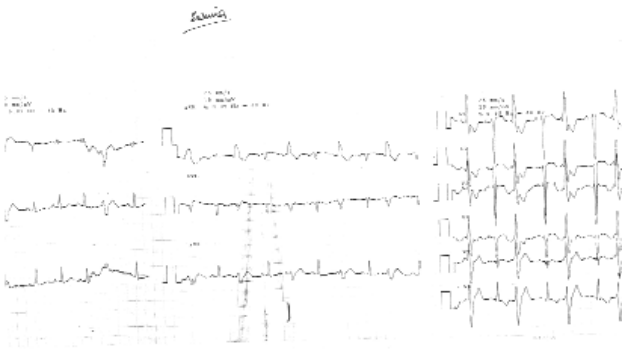
27 year old female resident of Nawabshah was admitted at National Institute of Cardiovascular Diseases(NICVD) with history of palpitation and shortness of breath for last 2 months. Her previous history revealed that she had stroke at the age of 3 year, which left her with limping gait and weakness of left arm.She had episodes of palpitation which increased in frequency and duration. For these problems she was investigated and was misdiagnosed as a case of cor triatriatum based on echocardiography and was advised medical follow up. She denied any family history of significant cardiac and non-cardiac disease. She was the mother of six healthy children and during her last pregnancy which was 18 months back, she started having shortness of breath associated with palpitation which progressively increased during last 2 months. For these problems she was bought to NICVD.

On examination she had shifted apex beat, left parasternal heave, irregular heart sounds and pansystolic murmur at apex. She had hemiplegic gait and decreased power and brisk reflexes on left side of body.On admission her ECG revealed sinus rhyhm with ventricular extrasystoles in form of bigemminies and left ventricular enlargement with intermittent atrial fibrillation.There was marked cardiomegaly on chest X-rays.Transthoracic echocardiography(TTE) showed severe MR with fluttering of anterior mitral leaflet and dilated with moderately dysfunctioned left ventricle and severe pulmonary arterial hypertension. There was markedly dilated (aneurysmal) left atrial appendage measuring 96x74mm with area of 43cm and volume on simpson's method of 183ml but LA dimensions were with in normal limits.There was spontaneous contrast in LA aneurysm.The diagnosis was subsequently confirmed by transoesophageal echocardiography(TEE) and MRI of chest. During her hospital stay she was not completely recovered despite appropriate medical management and she kept on having shortness of breath associated with palpitation so mitral valve replacement along with anuerysmectomy was recommended.



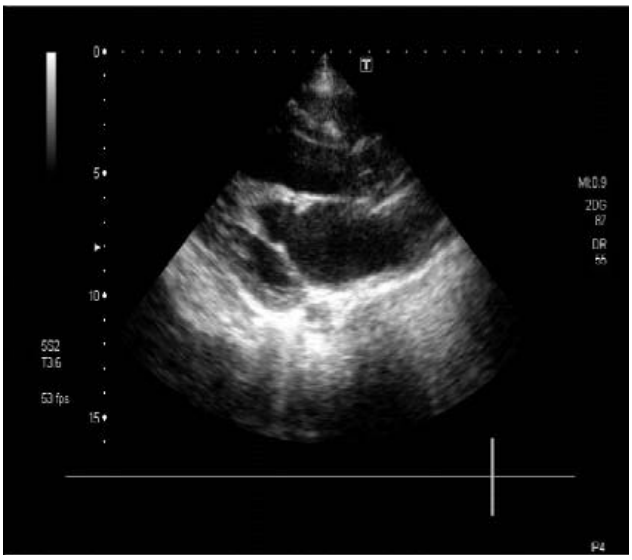
Massive cardiomegaly

* *[1] Post Fellow, [2] SR, [3] PG, [4] RMO, [5] Prof. at NICVD*

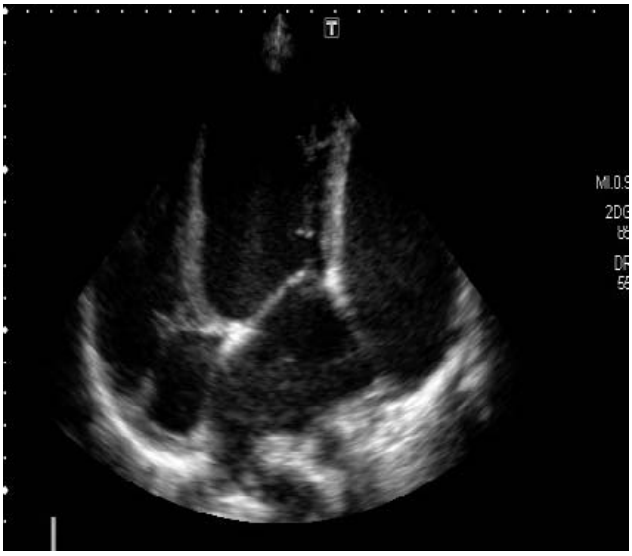


Ventricular bigemmies and LVH.

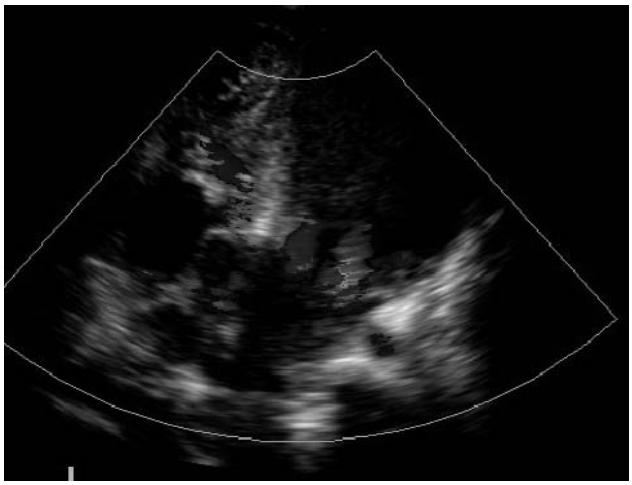
CHEST X-RAY



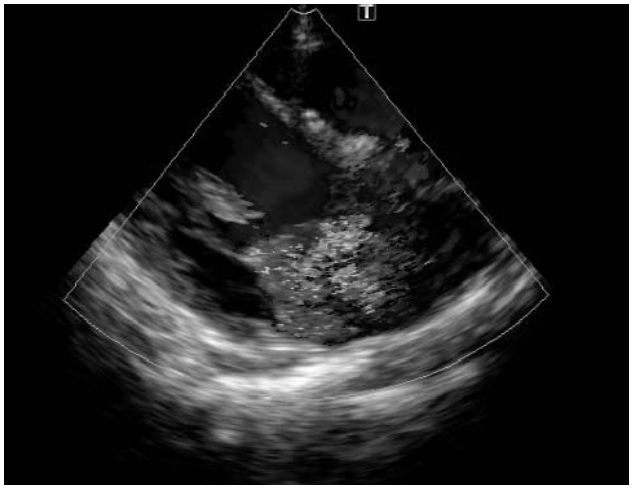
Parasternal Longaxis showing enlarged LV,normal sized LA,Ao and RV.



Apical LX showing huge LAA aneurysm



Apical LX showing huge LAA aneurysm with continous blood flow.



Parasternal LX showing severe MR.

ECG

ECHOCARDIOGRAPHY

SURGICAL NOTES

The surgery was done through a median sternotomy on cardiopulmonary bypass and with moderate hypothermia and aortic cross clamping. At the time of surgery it was found that pericardium was intact, left ventricle was enlarged, giant aneurysm extending from root of LA appendage going left laterally pressing the left ventricle and partly adhered to right ventricle.It was extended up to left diaphragm and with wide communication with the LA.There were multiple small thrombi in the aneurysm and the chordae were intact with myxomatous mitral valve which was then excised and

replaced by 29mm medtronic single disc valve and LA appendage aneurysm was partly excised and closed via left atriotomy.
Routine off bypass, but soon after the patient showed signs of RV failure and needed heavy ionotropic support which continued for next 16-18 hours and was expired despite all possible supportive measures.

HISTOLOGY

There was myxomatous degeneration of mitral valve but the appendage showed no signs of degeneration or inflammation and all three layers of atrium were intact.

DISSCUSION

Left atrial enlargement reflect remodeling associated with pathophysiologic processes and a predictor of common cardiovascular outcomes such as atrial fibrillation, stroke, mitral regurgitation MR[2], congestive cardiac failure and cardiovascular death due to rupture of LA aneurysm[3,4], cardiac tamponade[5].

LA enlargement is of 2 types;

1. Congenital
2. Acquired.

The acquired type is because of inflammatory or degenerative process involving the endocardium[6,7]. Some case reports have shown left atrial dissection as a cause of LA aneurysm after mitral and aortic valve surgery[8,9,10] and absent pericardium resemble LA aneurysm[11].

Congenital is a rare abnormality, first described by Semans and Taussig in 1938[12]. The enlargement may be of appendage or the body or both and size may be variable. Foale and colleagues[13] characterized the echocardiographic findings of LA aneurysms as:1.Origin from an otherwise normal LA,2.A well-defined communication with the atrium,3.A position within the pericardium ,and 4.Distortion of the left ventricular free wall by the aneurysm.In our case, all the 4 echocardiographic characteristics confirmed the diagnosis.

The age of presentation varies widely, from 1 month to 66 years and the mean age of presentation has been reported 23.5 years. Common manifestation of abnormal cardiac silhouette on chest x-ray, arrhythmias and systemic embolization. It may be associated with patent ductus arteriosis, atrial septal defect, myxomatous mitral value [3] bronchogenic cysts, diaphragmatic defects, anomaly of renal artery and, hypospadias [14, 15,16].

Echocardiography is usualy diagnostic.Magnetic resonance imaging (MRI)has been reported to aid in diagnosis[17]. In our case the of MRI was done to confirm the diagnosis and to delineate the pericardium as it is considered a useful modality to define the pericardium[18] and for that purpose short axis and coronal planes are needed to image the entire pericardium. There are different echocardiographic ways of measuring LA size, the more reliable is its volume than anteroposterir diameter[19,20] cardiac catheterization with cineangiocardiography is still recommended in order to evaluate pressures,shunts and biopsy if necessary.

Our patient was presented with all possible complications associated with LA aneurysm and it was associated with myxomatous mitral valve that was missed on echocardiography and MRI but was confirmed by both gross and histological examinations.Severe MR which was the cause of her rapid deterioration could be due to annulus dilation or involement of commissure of the mitral valve as a case report had shown related to LA aneurysm itself the other possibilities could be myxomatous mitral valve which could have ruptured during her last pregnancy.

Resection of the aneurysm is usually curative[21,22,23] with or without cardiopulmonary bypass[24]. However, there are no long-term follow-up studies on these patients[21]Besides traditional approches ,it has been demonstrated that a giant LA aneurysm can easily and safely be resected by a minimally invasive approach through an endoscope[25]. As our patient had myxomatous mitral valve with severe MR and LV dysfunction in addition to LA appendage aneurysm so it was prudent to perform mitral valve surgery along with aneurysmectomy.But due to delay in accurate diagnosis and associated complications ,the surgical outcome was disappointed.

CONCLUSION AND CLINICAL IMPLICATION

Early recognition of a LA aneurysm is important prognostically since SVT, systemic embolisation are common and it seems judicious to offer aneurysmectomy to otherwise healthy patient with congenital aneurysm of LA as a prophylactic measure. The natural history as judged by our case suggests that early accurate diagnosis and prompt surgical therapy would have obviated the risk of devastating result of this rare problem. .

REFERENCES

1. Kunishima T,Musha H,Yamamoto T,et al.Congenital giant aneurysm of the left atrial appendage mimicking pericardial abass case report.Jpn Circ J 2001;65:56-9.

2. J.Mark Morales,Sanjeet G.Patel,Josph H.Jackson,James A. Duff, and James W.Simpson. Left Atrial Aneurysm.Ann Thorac Surg 2001;71:719-22.

3. Dean A. Bramlet, Jesse E.Edwards.Congenital aneurysm of Left atrial appendage.Br Heart J1981; 45:97-100.

4. Ingram A, Macfie JWS.Two further cases of cardiac aneurysm.Ann Trop Med Parasitol 1922;16:119-25.

5 .Dimond EG,Kittle CF,Voth DW.Exreme hypertrophy of the left atrial appendage:the case of a giant dog ear.Am j Cardiol 1960;5:122-5.

6. Badui E,Delgado C,Enciso R,et al.Silent giant left atrium.A case report.Angiology 1995;46:445-8.

7. Terada H,Tanaka Y,Kashima K,Sannou K,Arima T.Left atrial diverticulum associated with severe mitral regurgitation.Jpn Circ J 2000;64:474-6.

8. Mikio Ninomiya,Tsuyoshi Taketni,Toshiya Ohtsuka,Noboru Moomura and Shinichi Takamoto. A rare type of left atrial dissection.JThorac Cardiovasc Surg. 2002;124:618-20.

9. Hiroshi Osawa, ShinpeiYoshii,Shigeru Hosaka, Shoji Suzuki, Samuel J.K.Abraham, and Yusuke Tada.Left atrial dissection after aortic valve replacement. JThorac Cardiovasc Surg.2003;126:604-5.

10. Martin C.Heidt, Ares K. Menon, Peter Roth ,Zan Mitrev, Hakan K. Akinturk, andPaul R.Vogt.Left atrial dissection after mitral operation mimicking severe mitral regurgitation. .JThorac Cardiovasc Surg.2004; 127:596-7.

11. Nasser WK.Congenital absence of the left pericardium.Am J Cardiol.1970;26:466-470.

12. SemansJH, TaussingHB.Congenital aneurysmal dilation of the left auricle.Bull Johns Hopkins Hosp1938; 63:404.

13. Foale RA,Gibson TC,Guyer DE,Gillam L,King ME,Weyman AE.Congenital aneurysms of the left atrium:recognition by cross-sectional echocardiography.circulation1982;66:1065-9.

14. Sagir T,Changani H, Yaqoob Z,Aziz K,Rehman M.Left Atrial Aneurysm or Absent Pericardium;A Diagnostic Dilema.Pakistan Paediatric Cardiology Journal 1999,Jan-Mar:27-31.

15. Parmley LF Jr.Congenital atriomegaly.Circulation 1962;25:553-8.

16. Sloman G,Hunt D,Hare WSC.Aneurysmal dilatation of the left atrial appendage. Med J Aust 1969;2:101-2.

17. Hoffmann U,Hamed N,Herold C,Globits S.Radiological signs of a left atrial aneurysm.Eur Radiol 2000;10(8):1332-4.

18. Schiavone WA James D,O Donnell.Congenital absence of the left portion of parietal pericardium demonstrated by nuclear magnetic resonance imaging.Am J.Cardiol 1985;55:1439-1440.

19. Walter P.Abhayarana,James B.Seward,Christopher P.Appleton,Pamela S.Douglas,Jae K.Oh,A.Jamil Tajik,Teresa S.M.Tsang.Left Atrial Size-Physiologic Determinnants and Clinical

Application.JACC Vol.47No.12,2006.

20. ng RM,Bierig M,Devereux RB,et al.recommendations for chamber quantification;a report from American Society of Echocardiograph? Guidelines and Standards Committee and Chamber Quantification Writing Group,developed in conjunction with the European European Society of Cardiology.J Am Soc,Echocardiogr 2005;18:1440-63.

21. Alok Mathur,MCh,Kenton J,Zehr,Lawrence J.Sink and Robert F.Rea.Left Atrial Appendage Aneurysm.Ann Thorac Surg.2005;79:1392-3.

22. Zhao J,Ge Y,Yan H,PanY,Liao Y.Treatment of congenital aneurysms of the left atrium and left atrial appendage.Tex Heart Inst J 1999;26(2):136-9.

23. Pome G,Pelenghi S,Grassi M,Vignati G,Pellegrini A.Congenital intrapericardial aneurysm of the left atrial appendage.Ann Thorac Surg 2000;69:1569-71.

24. Burke RP,Mark JB,Collins JJ Jr,et al.Improved surgical approach to left atrial appendage aneurysm.J Card Surg 1992;7:104-7.

25. Bob Kiaii,Nicolas Doll, Michael Kuehl and Friedrich W.Mohr.Minimal Invasive Endoscopic Resection of a Giant Left Atrial Appendage Aneurysm.2004;77:1437-8