Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 3 (Issue 12) : December 2016 ISSN: 2394-9414
DOI: 10.5281/zenodo.192488 Impact Factor- 3.109

ATYPICAL LOCATION OF A RIGHT ATRIAL MYXOMA- A CASE REPORT

Prof Naba Kumar Pattnaik, MS , MCh[*1] and Prof P K Mishra, MD[2]
[*1]Consultant, Cardio Thoracic surgery Apollo Hospital, Bhubaneswar
[2]Consultant, Anaesthesia, Apollo Hospital, Bhubaneswar

Corresponsing Author- Prof Naba Kumar Pattnaik, Consultant, Cardio Thoracic surgery Apollo Hospital, Bhubaneswar

Keywords:
Right Atrium, Myxoma, Surgery, Cardio Pulmonary Bypass,Intensive care \unit.

Abstract
Myxomas are the common type of cardiac benign tumour and most of them are located in Left Atrium, followed by, right atrium and have variable clinical presentation. We report a case ofin a 48 yr woman who had a large myxoma in right Atrium, which is an uncommon location, at the junction of right atrium and Superior Venacava.

INTRODUCTION

Cardiac Tumors represent 0.2% of all Tumours found in human(1). These tumors are divided into primary and secondary or metabolic. These secondary tumors are 20 to 40 times more frequent than primary tumors. Primary intracardiac tumors are rare. Approximately 75% are benign and out of which approximately 50%of them are Myxomas, which have an incidence of 0.0017% in general population. Histologically these are real tumors derived from multipotent mesenchyme cells of the suberdocardium(2)

Myxoma are located in left atrium in 75% to 80% of cases and are almost always present with signs and symptoms of Mitral valve disease. They may arise in other locations like right atrium (RA), 18% of cases (3) and rarely in aorta, pulmonary artery, ventricles and vena cava. Right atrial myxomas are sometimes associated with tricuspid stenosis and atrial fibrillation (AF). The differential diagnosis is performed between rhabdomyoma or thrombus. Myxomas affect patients within a wide age range (15 to 80 years) and the average age is approx 50 years. There is a female predominance in the sporadic from (4). Myxomas may be pedunculated, solitary and sporadic but may be associated with familial autosomal dominant syndrome 7 % of cases.(5)

About 10 % of myxomas are passed down through families (inherited) as in Carney syndrome, where several other abnormality are observed such as skin myxomas , pigmentation , endocrine hyper activity, schawannomas and epitheloid blue nevi. Such tumors are called familial myxomas. They tend to occur in more than one part of the heart at a time and often cause symptoms at an younger age than other myxomas. They are multiple and have a risk of recurrence after surgical excision (6). In regard the macroscopic feature the shape can be Oval rounded and irregular. Brownish color is predominant. Consistency is variable, firm to gelatinous.

Microscopic features of Myxoma is characterized by a myxoid matrix rich in mucopoly-saccharides in which polygonal cells with eosinophlic cytoplasm can be detected. The polygonal cells may appear as a star or may be multi nucleated. Microscopic features like mitosis, necrosis or pleomorphism are not detected.

The signs and symptoms caused by myxomas are atypical and highly variable and result in a difficult diagnosis of the neoplacia. (7). According to size mobility and location of the tumor as well as body position of the patient may follow an asymptomatic course or progress to thromboembolic event that may lead to sudden death.(6)

Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 3 (Issue 12) : December 2016 ISSN: 2394-9414
DOI: 10.5281/zenodo.192488 Impact Factor- 3.109

Symptoms- The classical triad found in cardiac myxoma is characterized by, (i) the obstruction of the blood flow,(ii) constitutional symptoms and (iii) thrombo embolic event (4) The obstruction to blood flow leads to intermittent heart failure. Constitutional and non specific symptoms like malaise, low grade fever of long duration, arthralgia, anorexi may occur. Thrombo embolic event may happen. RA Myxoma particularly may obstruct the Tricuspid valve, causing sign and symptom of right heart failure, peripheral oedema hepatic congestion and syncope. (4)

Cardiac auscultation may vary according to size, location, mobility and prolapse of tumor through Atrio-Ventricular (AV) valve and body position. Therefore a murmer may or may not be detected.Tumor plop is an typical auscultatory findings caused by presence of tumor inside atrial chamber that occur in 15% of cases.(4) . Routine laboratory examination may show nonspecific change such an anemia, increased ESR, increased level of globulin and of C Reactive Protein with leucocytosis. Recent studies show Cardiac Myxoma produce and release a substance called interlukin-6 which is responsible for inflammatory or auto immune phenomena.

ECG may be normal or shows arythmia and heart block, chest X ray findings may be nonspecific (2). Transthoracic Echocardiography is an excellent sensitive investigation in detecting 95% of of Myxoma, the sesitivity increases to 100% when followed by Ttrans Oesophagial echocardiography (TEE) (4). Computed tomography and MRI may be useful to demonstrate the point of attachment of myxoma and associated calcification. Once Myxoma is diagnosed surgical excision is done with out delay because of thromboembolic event.(1) Generally surgical treatment is definitive and recurrence is uncommon.

We report a rare case of large right atrial myxoma because of its unusual location.

CASE PRESENTATION

A 48 years old woman presented with a10 yeras history of palpitation, dyspnoea on exertion, arthralgia and weight loss which was intense in last two months. The patient was treated as chronic obstructive pulmonary disease by the primary physician. She was referred to our cardiac unit because of more frequent palpitation.

There was no anemia, pedal oedema and signs of heart failure. On auscultation heart sounds were normally heard, and without any murmur. Chest was clear. X-ray of the chest showed right atrial enlargement with clear lung fields (Fig:1) . ECG showed sinus rhythm with right bundle branch block (RBBB). Trans thoracic 2-D echo showed a moving mass at the junction of RA and superior venacava measuring 5cm x 4.0 cms.(Fig -2) ,with RA enlargement and RV overload. After due preparation patient was taken for surgical removal of the tumor under cardio pulmonary bypass.

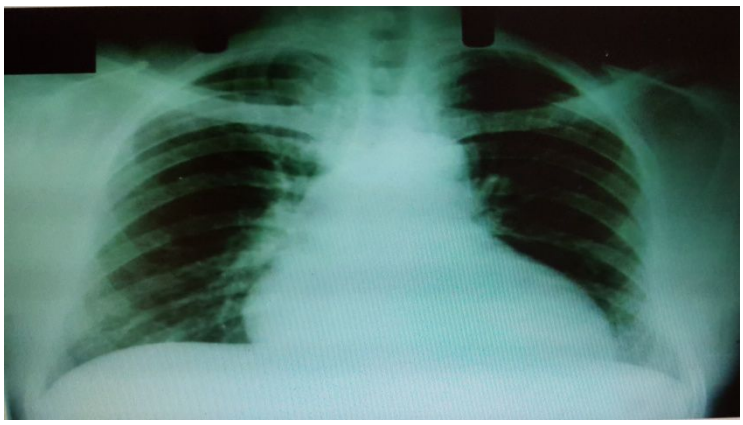


Fig -1 A chest Xay showing Right Atrial Enlargement

Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 3 (Issue 12) : December 2016 ISSN: 2394-9414
DOI: 10.5281/zenodo.192488 Impact Factor- 3.109

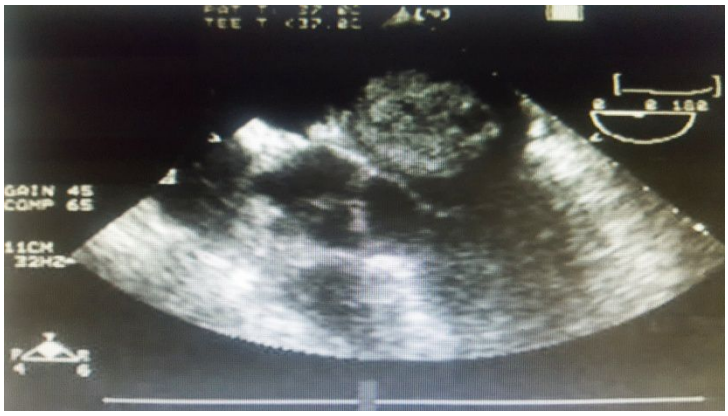


Fig-2 Trans Thjoracic Echo cardiogram showing mass in right Atrium

Under General Anaesthesia midline sternotomy was done . CPB was established with bi caval cannulation followed by conventional mild hypothermia of 34^{o}C. Cardiac standstill obtained with cold cardioplegia and aortic cross clamp. RA was enlarged and a right longitudinal atriotomy was done . On inspection there was a mass (5 cm x 4.5 cm), which was mobile , pedunculated and lobulated with implantation at right atrial and superior venacaval junction.(Fig 3, 4). The mass was resected in toto and sent for histopathological study.Weaning from CPB was uneventful. Patient was transferred to ICU with stable hemodynamic status. Post operatively patient developed Supra ventricular tachycardia (SVT) , which was controlled with intravenous Amiodarone. Sinus rhythm was restored after 6-7 hours. Subsequently patient recovered well and was discharged on 8th post op day.

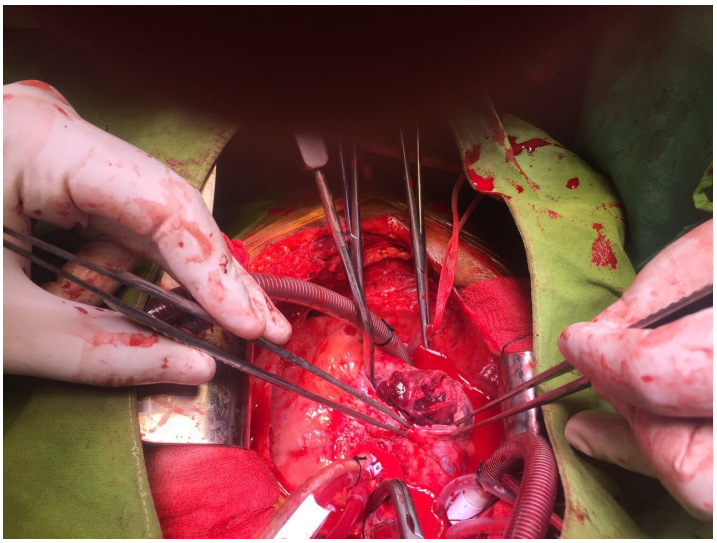


Fig – 3 Intra operative picture showing the right atrial mass being delivered.

Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 3 (Issue 12) : December 2016 ISSN: 2394-9414
DOI: 10.5281/zenodo.192488 Impact Factor- 3.109

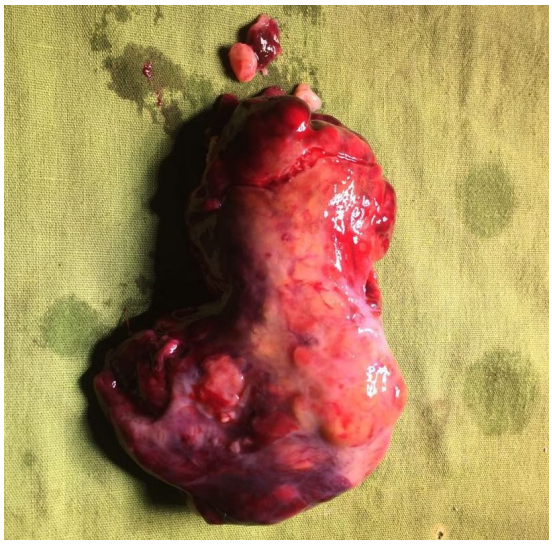


Fig- 4 A lobulate Right atrial mass with polypoid area

Histopathologically the mass had abundant myxoid stroma with stellate cells. Necrosis, mitotic activity atypia and pleomorphism wes not detected. (Fig- 5). This confirms the diagnosis of myxoma. The patient is on follow up for last2 years and has occasional palpitation but with out any other signs of heart failure .

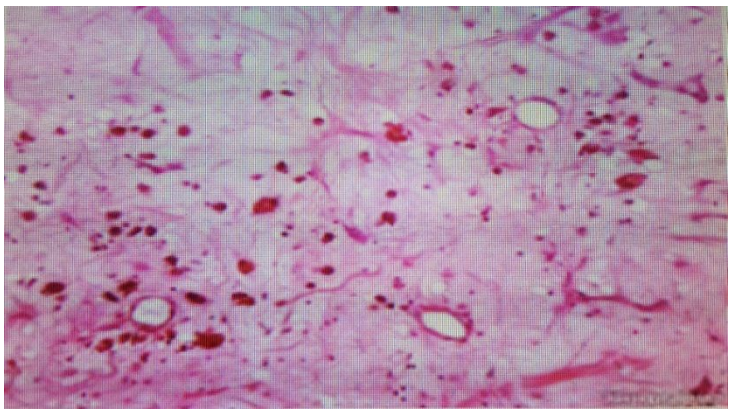


Fig-5. A microscopic view of myxoid stroma with stellate cells within.

DISCUSSION

Myxoma is the most prevalent primary cardiac tumor . RA is an unusual location and is the site of 15 to 20 % cases of Myxoma (3). About 70 % affected patients are women predominantly in 3rd and 6th decade of life (2), as in our case, who was 48 years and a female.

RA Myxomas originate in Fossa Ovalis or base of intra-atrial septum (7), but in our case Myxoma was implanted in the right atrial SVC junction. Myxomas are usually pedunculated, approximately in 83% of cases.(11). In our case the tumor was pedunculated and solitary. In a recent publication, reporting with 19 years of

Volume 3 (Issue 12) : December 2016 ISSN: 2394-9414
DOI: 10.5281/zenodo.192488 Impact Factor- 3.109

experience with surgical treatment of primary cardiac myxoma, 17 % cases , out of 41 originated from RA. Mean maximal diameter was 5.1 ± 1.8 cm (9).

The sign and symptoms of RA myxoma are atypical and depends on size, position and mobility of tumor and may change according to body position of patient. (4). RA myxoma may be asymptomatic and may cause symptoms like anaemis, weight loss ,fever and arthralgia due to production of interleukin-6. (11). Most common manifestation is dyspnoea (80%) and right heart failure. Our patient had palpitation probably because of location of the tumour in RA and SVC junction. These symptoms disappear after the tumor is removed.

Echo cardiography remains the best diagnostic method for locating and to asses the extent of the tumor. How ever this may not identify tumor smaller than 5 mm in diameter. So trans oesophageal echo cardiography (TEE) is done when there is suspicion of very small tumor.(12)

The treatment of choice is surgical removal (11). Complete resection of the tumor and its implantation base with a good safety margin is essential to cure the disease. In this case the tumor was located at the superior venaval junction with RA making with wide excision with large margin was not possible because of critical anatomical location and consequent high risk of conduction disturbance,

The recurrence rate of sporadic tumor is very low. (1-3 %) (4). The operative mortality ranges from 0-3 % in multiple series of cases (4). Some important aspect shoud be taken into consideration for the surgical treatment of myxoma. Care should be taken to clamp both Aorta and pulmonary trunk to avoid embolisation of myxoma before resection.

CONCLUSION

Though a rare location of a large myxoma, the RA myxoma should always be considered in the differential diagnosis of a right sided heart mass, especially when the patient shows signs and symptoms of heart failure with uncertain aetiology. The findings in our case suggests cariologists and cardiac surgeon need to make early diagnosis and treat patients with these tumors to improve prognosis.

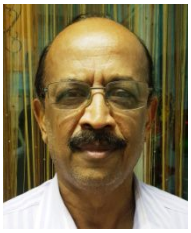
REFERENCES

1. *Arruda MV, Braile DM, Joaquim MR, Soares MJ, Alves RH: Resection of left ventricular Myxoma after embolic stroke, Rev. Bras Cir Cardiovas; (2008),* ***23****, 578-580, 10, 1590/50102 (Pubmed).*
2. *Jang KH, Shin DH LeeC, Jang JK, Cheong S, Yoo SY.. "Left atrial mass with stalk: thrombus or Myxoma ?" : J Cardiovas Ultrasound;(2010):****18****:154-156.*
3. *Diaz A, Di Salvo C, Lawrence D, Hayward M: "Left atrial and right ventricular myxoma: an uncommon presentation oJ a rare tumour". Interact Cardiovasc Thorac Surg. 2011, 12: 622-623Oliveira R, Branco L, Galrinho A, Abrean A, Abreau J: "Cardiac Myxoma: a 13 years experience in echo cardiography(2010);,* ***29****:1087-1100*
4. *Croti UA, Braile DM, Souza AS, Cury PM: "Right ventricle and Tricuspid valve Myxoma."; Rev Bras Cir Cardiovasc. (2008),* ***23*** *:142-144*
5. *Hermans K, Jaarsma W, Plokker HW, Cramer MJ, Morshuis WJ; "Four. Cardiac Myxoma diagnosed three times in one patient"; Eur. J. Echocardiogr; (2003),* ***4 :*** *336-338.*
6. *Murrayama H, Tamaki S, Kato N, Yuzi N, Yokote J, Motsuga M, Okada M;. "Right Atrial Myxoma associated with atrial septal defect: a case report and review of literature" Ann Thorac Cardiovas Surg.(2001)* ***7*** *; 166-169*
7. *Stalf NA, Venicio A, Moreina LF, Rossi E. "Right atrial myxoma originating from inferior venacavae, an unusual location with therapeutic and diagnostic implication." Rev. Bras Cir Cardiovas ,(2000);* ***15****; 255-258.*

Volume 3 (Issue 12) : December 2016 ISSN: 2394-9414
DOI: 10.5281/zenodo.192488 Impact Factor- 3.109

8. *Vale Mde P, Freire Sobrinho A, Sales MV, Teixeira MM, Cabral KC: Giant myxoma in the left atrium: case report Rev Bras Cir Cardiovasc. 2008, 23: 276-278.*
9. *Samanidis G, Perreas K, Kalogris P, Dimitriou S, Balanika M, Amanatidis G etal; "Surgical treatment of primary intra cardiac myxoma, 19 years experience." ; Interact Cardiovas Thorac surg; (2011);* ***13*** *: 597-600 (Pubmed)*
10. *Yuce M, Dagdelen S, Ergelen M ,Caglar N. " A huge obstructive myxoma located in right heart without causing any symptoms.". Int J Cardiol (2007);* ***114,*** *405-406. (Pubmed).*
11. *Guhathakurta S, Riordan JP; "Surgical treatment of Right Atrial Myxoma". Texas heart Inst. J. (2000),* ***27****, 61-63 (Pubmed)*
12. *Manfroi W, Vieira SR, Saadi EK, Saadi J, Alboim C. "Multiple recurrences of cardiac myxoma with acute tumoral pulmonary embolism".Arq Bras Cardiol (2001);77:161-163 (Pubmed)*

AUTHOR BIBLIOGRAPHY

	**Prof Naba Kumar Pattnaik, MS, Mch (CTVS)** Consultant, Cardio Thoracic surgery Apollo Hospital, Bhubaneswar Plot no. 1211/C, Sector- 6, CDA, Bidanasi, Cuttack -753014
	**Prof Prasanna Kumar Mishra , MD (Anaesthesiology)** Consultant, Anaesthesia, Apollo Hospital, Bhubaneswar