Unique Pearls In Synergy-Atrial Myxoma And Hypertrophic Cardiomyopathy

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ABSTRACT

Primary cardiac tumors are a rare occurrence amongst general population, myxoma being the most common benign primary cardiac tumor. Hypertrophic cardiomyopathy(HCM) is a genetic cardiovascular disorder which usually presents with syncope or palpitations. The case we present here is a unique amalgamation of both these pathologies which are uncommon as well as completely unrelated to each other as far as literature describes. The patient presented with symptoms of congestive cardiac failure without any history of syncope and palpitation ,however echocardiography revealed the presence of HCM as well as atrial myxoma. This case reveals that remote possibility of such coexisting conditions should be kept in mind in clinical practice and also the fact that HCM can notoriously remain undetected presenting only as heart failure instead of its usual clinical features.

Key words: Hypertrophic cardiomyopathy, Myxoma, Echocardiography, Heart failure

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INTRODUCTION

Primary cardiac tumors are very rare in the general population, with a reported prevalence of 0.001% to 0.03% in autopsy series. Secondary involvement of heart is much more common than primary cardiac tumors. Majority of primary cardiac tumors are usually benign (75%), Myxomas are most common amongst them.

Hypertrophic Cardiomyopathy(HCM) is the most common among the genetic cardiovascular diseases and is characterised by heterogenous clinical expression, unique pathophysiology and diverse clinical course. The prevalence of HCM in the general population is about 0.2%. HCM is a very rare occurrence in the common practise and cases usually go undetected. HCM is inherited as a Mendelian trait in an autosomal dominant fashion. Patients of HCM may present with symptoms of low output cardiac failure in case of obstructive variant of HCM. In the rest of the cases, patients may remain asymptomatic or develop symptoms of diastolic failure in late stages.

No definite association has been described between the presence of atrial myxoma and HCM.

Here we describe the case of a 65 yr old male patient who was incidentally detected to be having Right atrial myxoma with Hypertrophic Cardiomyopathy (non- obstructive) after the patient presented with symptoms of congestive heart failure. The patient finally suffered Sudden Cardiac Death(SCD) during hospital stay.

CASE REPORT

A 64 yr old male patient was on regular outdoor follow up for last 4 years for hypertension . Patient had history of 1 episode of hospitalisation for dyseclectrolytemia 4 yrs ago.Recently the patient had started developing lower urinary tract symptoms like frequency and hesitancy. he also suffered from one episode ofurinary retention.He was on silodosin for his symptoms to which he responded.

During his last outdoor visit in March 2015, the patient complained of swelling of bilateral lower limbs along with abdominal swelling, orthopnea for last 20 days along with high grade intermittent fever sometimes associated with chill and rigor. Urine output was decreased for last 15 days. The patient was advised for hospitalisation. At the time of hospitalisation, patient was consious and alert. Vitals were stable. JVP was raised and pedal edema was present. Heart sounds were muffled. No added heart sounds, murmur or tumor plop could be heard. Abdomen was distended. There was no organomegaly. The patient did not have any history of chest pain or syncope in the past. No history of sudden cardiac deaths in the family.

The patient was started on iv antibiotics with low dose diuretics and ace inhibitors as the patient had symptoms of right heart failure. Blood culture samples were sent along with routine investigations in order to rule out infective endocarditis. Investigations reveled cardiomegaly on Chest X ray along with right axis deviation and features of left ventricular hypertrophy with inverted T waves in lateral leads(V5 , V6) on ECG. Blood reports revealed a normal TLC with Hb 9.7g%. Serum electrolytes and creatinine leveks were normal. Liver function tests were within normal limit. ESR = 75.

Transthoracic Echocardiography revealed IVS 32 mm along with LVIDd 30 and LVPW 16. LVEF 63%. No SAM or MR could be seen. LVOT Gradient < 30 mm hg. Diastolic dysfunction present(E/A< 1). Mitral valve morphologicaly and functionally normal (EF Slope 40). LA was not dilated whereas RA was slightly dilated. A pedunculated mass was seen in the Right atrium attached to the atrial side of the tricuspid leaflet along with prolapse into the ventricular cavity during diastole suggestive of atrial myxoma. Tricuspid valve area slightly reduced (1.5 cm2) . Impression - HCM (non obstructive) along with Right atrial myxoma.

On the subsequent day(Day 2), the patient developed a feeble low volume pulse with rate of 100/min along with Blood pressure of 90/60. Considering this fall in Blood pressure as compared to previous day,diuretics were promptly withdrawn. Patient was symptomatically better though as his respiratory distress had decreased.

The following day(Day 3), patient was doing well with a blood pressure of 110/70 and all his vital signs were stable. Signs and symptoms of heart failure had decreased. Considering the presence of HCM, the patient was started on beta blockers at a low dose.

On Day 4, the patient suddenly became disoriented and agitated. His vitals were stable. SpO2 was 96% on room air. Blood sugar levels were normal. Plantar responses were normal bilateraly. No focal neurodeficit was elicited. Investigations revealed a normal electrolyte panel. (Na+ - 125 , K+ - 4.5 , Ca2+ - 8.1) . Urine output was normal. CT Scan Brain did not reveal any abnormality.

On Day 5, the patient's agitation and irritability decreased. He wasnt able to follow commands. He developed choreiform movements involving both hands along with head nodding. No other abnormality could be elicited in the neurological examination. On the same night after the patient had taken dinner, he complained of chest discomfort and collapsed immediately. He suffered Sudden cardiac death and could not be resuscitated.
His blood culture reports did not show any bacterial growth.

DISCUSSION

Primary tumors of the heart have a very low incidence among the general population. An incidence of 0.001-0.03% has been reported in autopsy series. Primary cardiac tumors can be both benign and malignant. The most common benign tumor is myxoma followed by rhabdomyoma,fibroma,lipoma and others. Amongst primary malignant tumors, sarcomas top the list with angiosarcoma being the most common. Secondary tumors are metastatic in nature, with melanoma having the highest incidence of mets to the heart.

Myxomas are the most common benign primary neoplasm of the heart. They have an intracavitary location with 75% located in the left atrium, 25% in the right atrium and <2% in the ventricular cavity. Myxomas are usually solitary but on some occasions can be multiple and can be present in >1 cardiac chambers.

Myxomas can be both sporadic and familial in origin. Sporadic myxomas occur in mostly women and the mean age of occurrence is usually above 56yrs. Familial myxomas constitute about 10% of all myxomas and they have an autosomal dominant pattern of inheritance. They occur in the younger population. Patients having familial myxomas usually have associated skin findings like freckling and skin pigmentation.NAME and LAMB syndromes are usually associated with familial atrial myxoma.

Myxomas are can be asymptomatic and clinically silent or they can have a varied range of symptoms. Myxomas,especially left atrial, can produce symptoms mimicking mitral stenosis. In some cases, the mitral valve damage may cause mitral regurgitation. Atrial myxomas can lead to pulmonary or systemic congestion depending on location. Peripheral embolisation can result from myxomas of which most dangerous is CNS embolization. Atrial myxomas can be clinically silent and can present with only constitutional signs and symptoms like

fever,weight loss, anemia,raised ESR. In such cases, they tend to mimic a collagen vascular disease or malignancy..

Our patient was also suffering from Hypertrophic Cardiomyopathy(non-obstructive) which was also an incidental diagnosis. HCM is mostly familial and has an autosomal dominant mode of inheritance. Several genes have been identified in association with HCM amongst which beta myosin heavy chain gene and myosin binding protein gene are the most important. Patients usually suffer from unexplained syncope, palpitation due to arrhythmias or episodes of SCD from which they have been successfully resuscitated. HCM is the commonest cause of SCD in young athletes.

The patient in our case report was incidentaly diagnosed to be having HCM with atrial myxoma after he presented with signs of congestive heart failure. The patient had no history of syncope,chest pain or any thromboembolic event in the past. There was no history of syncope or Sudden cardiac death amongst first degree relatives of the patient. No freckling or any other skin involvement was present. No documented episode of arrythmia was present in the past. Diastolic dysfucntion due to HCM was the cause of the heart failure. The fever associated with heart failue was probably a constitutional symptom related to myxoma . As the patient had no family history of myxoma, he had a sporadic atrial myxoma. The occurence of SCD was a natural event in the course of the disease which could have been prevented with ICD but there were no definitive indications for ICD apart from IVS thickness.

No definite association has been found between myxoma and chorea. No reports of such incidence have been found in the literature. However there are some case reports in which patients having myxoma have presented with features of vasculitis and have been misdiagnosed initially. Literature reports an interesting case of a 48yr old woman who had presented with a history of weight loss and superficial left femoral artery thrombosis. The patient had a history of collapse. During workup of the patient, the patient was seen to having critical stenosis of the involved vessel with high values of ESR and CRP and associated anemia. CT pelvis showed presence of inflammatory changes around femoral artery and thus a provisional diagnosis of large vessel vasculitis was made. It was only after the echocardiogram was done that the patient was found to be having a Left atrial myxoma. Thus myxoma can present with features of vasculitis. Other reports are present where a misdiagnosis of polyarteritis nodosa or Giant cell arteritis have been made in cases of myxoma. Our patient did not have any overt features of vasculitis apart from raised ESR. The occurrence of choreiform movements in the patient might have been due to vasculitis associated with myxoma but it is only a hypothesis as further investigations could not be done to confirm this theory.

Our patient had a combination of atrial myxoma and non obstructive hypertrophic cardiomyopathy. Very few cases have been reported in literature with this combination. No definite association or causal factors have been defined between these 2 conditions. The only known association that exists between these conditions is familial lentiginosis syndromes. In Carney complex and LEOPARD syndrome, atrial myxoma may co-exist with HCM. But in

our case no other features especially lentigines could be found in the patient. In the cases reported previously, patients were treated separately for both these conditions. Myxomas were resected out and most of the patient had ICD implantation for primary prevention against SCD.

CONCLUSION

This was a rare case of combined atrial myxoma and hypertrophic cardiomyopathy where no causative factors or association could be explained. One must keep in mind that such a combination may be present even in the absence of typical history like palpitation and syncope which is characteristic of HCM. This is a rare diagnosis which can present with a wide array of signs and symptoms.

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