

Uncommon cause of cardiac tamponade in a young man

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SUMMARY

We report a rare case of cardiac angiosarcoma in a young boy who presented with cardiac tamponade. His initial symptoms were non-specific. He was initially being managed in the line of fungal infection, with a possibility of malignancy. Cardiac imaging was also not conclusive and he worsened on antibiotics and antifungals and succumbed to the illness. After his death tissue biopsy from heart and lung was done and histopathological examination revealed the diagnosis of metastatic angiosarcoma. The case highlights the importance of considering the diagnosis of cardiac angiosarcoma in the patients presenting with haemorrhagic pericardial effusion and non-specific symptoms.

BACKGROUND

Cardiac angiosarcoma is a rare tumour and usually has a poor prognosis.¹ An early diagnosis and intervention is important in prolonging survival. Due to rarity of presentation and non-specific signs and symptoms it is often diagnosed late further complicating the course. Cardiac tamponade has been rarely reported as a presenting manifestation of angiosarcoma. Herein we report a case of cardiac angiosarcoma presenting with the cardiac tamponade.

CASE PRESENTATION

A 24-year-old man was rushed to emergency room with complaints of 2 days history of acute onset retrosternal chest pain, relieved on sitting position, dyspnoea at rest and with profuse sweating. The symptoms worsened with time. On initial assessment he was tachypenic, responding to calls, his pulse rate was 140 per min, blood pressure was recorded as 80 mm Hg (systolic) with cold extremities. His Jugular Venous Pressure (JVP) was raised and had prominent x wave descent. Heart sounds were muffled and respiratory sounds were normal with occasional scattered crepitations. Bedside ECG showed low voltage complex with diffuse ST segment elevation. Bedside transthoracic echocardiography showed pericardial effusion with features of cardiac tamponade. Immediate bedside drainage of pericardial fluid was done and a pigtail catheter was placed, which drained 500 mL of haemorrhagic fluid after which he was relieved of acute symptoms. After stabilisation, he gave history of similar episodes of dyspnoea on exertion and, chest pain 6 months back. He also gave history of on and off fever in the last 6 months for which he was started on four drug anti-tubercular therapy.

FURTHER COURSE

Transthoracic echocardiography, after fluid drainage, revealed a suspicious mass lesion with size 4×5 cm adherent to posterior wall of right atrium. Analysis of pericardial fluid showed total leucocyte count of 1000 (with 55% polymorphs), full field red blood cells, protein of 3.5 gm% and glucose 27 mg%. Fluid Adenosine deaminase was 35 IU/L. Given the history of fever, after drawing blood and pericardial fluid cultures, the patient was started on broad spectrum antibiotics. Fluid was also sent for cytological evaluation. His pericardial drainage had a daily output of 500 to 700 mL which was haemorrhagic in nature. His pericardial fungal culture grew yeast cells on two occasions on the basis of which the catheter was changed and antifungals were added. All the other cultures including blood culture were sterile. His total blood leucocyte count worsened from 14 000/μL to 55 000/μL over the hospital stay. Repeated pericardial fluid cytological analysis showed no abnormal cells.

IMAGING INVESTIGATION

On CT imaging (figure 1), both lung fields showed features of non-homogenous opacities suggestive of multiple ground glass opacities and consolidations suggestive of an infective pathology. Mediastinal window showed an intracardiac mass lesion in right atrium and multiloculated pericardial effusion.

On cardiac MRI (figure 2), there was presence of loculated pericardial effusion, with multiple haemorrhagic pockets. There was extensive nodular soft tissue thickening of the septations which showed enhancement on post contrast images. There was a heterogeneous soft tissue lesion, measuring 45×20×60 mm involving the right atria (RA) wall and adjacent interatrial septum, with frond like mobile protrusions into the right atrial cavity, and infiltrating the adjacent pericardium. The lesion showed mild patchy enhancement on late gadolinium enhancement sequences, however, some

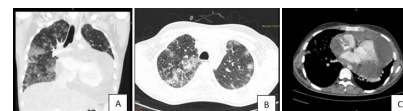


Figure 1 (A) Coronal CT chest (lung window) shows diffuse ground glass opacities with interspersed air space nodules showing basal predominance, gross pericardial effusion is also seen. (B) Axial CT chest (lung window) through the apices show patchy nodular consolidation as well as ground glass opacities with mild septal thickening. (C) Axial CT chest (soft tissue window) shows gross pericardial effusion.



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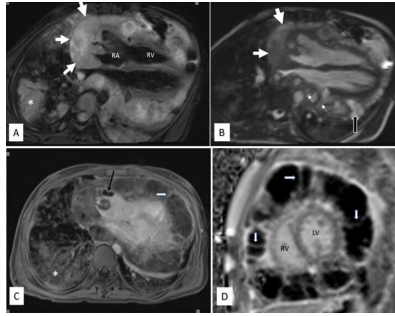


Figure 2 Four chamber cardiac MR images (A to C), T2 (A), Cine (B) and post contrast volumetric interpolated breath-hold examination (VIBE) sequence (C) show presence of an infiltrative right atrial wall lesion (white block arrows in A and B) with adjacent pericardial effusion (black block arrow in B) with haemorrhagic components, with multiple thick septations and soft tissue thickening (white thin arrows in B). On post contrast VIBE image (C) and short axis late gadolinium enhancement image (D), these septations show enhancement. Multiple lung lesions are also seen (white asterisk in A and C). LV, left ventricle; RA, right atrium; RV, right ventricle.

of the adjacent nodular lesions did not enhance, suggestive of adherent thrombi. There was presence of diastolic septal bounce of the intraventricular septum suggestive of constrictive physiology. Multiple extensive nodular lesions and lobar consolidations were seen in bilateral lungs. The MRI features were in favour of an infiltrative RA wall mass, most likely neoplastic or inflammatory (fungal) lesion, with extensive pericardial and lung involvement.

OUTCOME

He was planned for a positron emission tomography scan for the differentiation between an inflammatory mass and malignancy. However he developed new onset shock with fever. His shock worsened quickly and subsequently he got intubated and died within 5 days.

HISTOPATHOLOGICAL FINDINGS

After obtaining consent from parents, minimally invasive tissue sampling was performed with the help of bedside ultrasound machine. Samples were obtained from right atrial mass, lungs, liver and kidney, along with pericardial fluid and were sent for pathological analysis. The biopsy from right atrial mass showed histomorphological and immunohistochemical features of angiosarcoma (figure 3). Tumour cells were immunopositive for CD31, CD34, vimentin and increased MIB-1 index (30% to 40%) and negative for cytokeratin. Lung biopsy showed evidence of metastasis. Liver biopsy showed maintained lobular architecture with sinusoidal dilatation and focal cholestasis. The pathological findings suggested the diagnosis as cardiac angiosarcoma with metastasis.

DISCUSSION

Neoplastic invasions is one of the most common cause of spontaneous pericardial tamponade, more commonly associated with cancers of lungs, breast, melanomas and lymphomas. Pericardial involvement is however a late finding in such cases.² Primary cardiac neoplasms are extremely rare tumours.³ Metastatic tumours are more common than primary cardiac tumours.³ Cardiac tumours maybe asymptomatic or detected incidentally during evaluation. Primary angiosarcoma is one of the the most

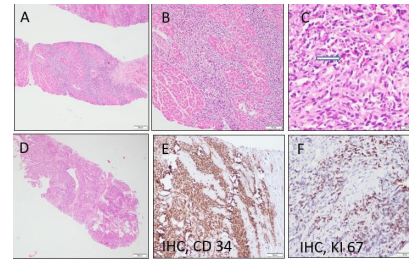


Figure 3 Histopathology of cardiac angiosarcoma: Trucut biopsy from the heart shows a (A) malignant mesenchymal tumour infiltrating into the myocardium; (B) higher magnification reveals that it is a vasoformative tumour with (C) significant nuclear pleomorphism (red blood cells in the vasoformative areas, arrow); (D) section from the lungs show presence of metastasis with similar tumour morphology; immunohistochemistry (IHC) reveals that these tumour cells are strongly positive for CD34; (F) tumour proliferating index (KI 67 labelling index) is high, approximately around 40% to 50%.

common intracardiac malignant tumours which usually has a poor prognosis owing to its aggressive nature and delay in diagnosis.

In primary cardiac angiosarcoma men are affected more than women in a ratio of 2 to 3:1.³ Most of the cases reported are those less than 65 years of age, though the age distribution is quite wide.⁴ There has been even reports of familial cases of cardiac angiosarcoma.⁵

The most common site of a cardiac angiosarcoma is right atrium, 90% of tumour occur in right atrium as a multicentric mass. Left-sided chambers contribute to 5% of cases⁴ and there has been even case reports of angiosarcoma arising from coronary vessels. If symptomatic, patients may present with complaints of shortness of breath, weight loss and fatigue.⁶ The non-specific symptomatology and a good amount of asymptomatic cases usually results in a delay in diagnosis. In the later stages of disease it may present with frank features of metastasis.

In a country where tuberculosis is endemic, the initial manifestations of this disease may be mistaken to be due to the mycobacterial disease. In an appropriate clinical setting it is important to consider a diagnosis of cardiac tumours.

The diagnosis of the tumour is suspected on imaging including echocardiography (ECHO), CT scan and nuclear imaging. A transoesophageal ECHO provides more information and is more sensitive compared with a transthoracic ECHO with sensitivity up to 97% in detecting cardiac mass.⁷ Cardiac MRI is more sensitive investigation compared with cardiac CT in detecting and characterising the lesion. MRI can differentiate a thrombus and tumour inside cardiac cavity.⁸ There are two different characteristic pattern defined in MR images, first of which is based on haemorrhagic and necrotic character of tumour and second is based on a diffuse involvement of pericardium.⁹ Pericardio-centesis and tissue biopsy can also be used in diagnosis though pericardial fluid cytology often has poor sensitivity.⁶ Endomyocardial biopsy can be diagnostic in approximately 50%.¹⁰ For an accurate diagnosis, the best method still is surgical exploration and frozen section biopsy.¹¹

Most of the patients with angiosarcoma has metastases at presentation. Most common site is lungs followed by liver, lymph node, adrenal and spleen.¹⁰ Brain metastases are uncommon. Immunohistochemistry of tumour shows positivity to CD31, CD34, cytokeratin, FLI-1 (FLI: Friend leukaemia virus integration), von Willebrand factor. In addition Ki-67, p53 and vimentin may also be positive in tumour.¹⁰⁻¹²

Histologically angiosarcoma consists of endothelium lined channels, which forms well differentiated vascular channels, which vary in size based on frequency of mitosis, solid spindle areas with minimal or no apparent vascular spaces, foci of endothelial tufting and lack of calcification.

The prognosis of the tumour is extremely poor with mean survival without surgical resection being close to 3 to 4 months.¹¹ Being a rare disease the therapy is not standardised. Considering the high fatality rate and aggressive treatment course is usually followed consisting of surgery, chemotherapy and radiotherapy. Surgery is the most common method followed in most of the cases with a median survival post-surgery of 14 months. Right-sided tumours are more bulky, infiltrative and more likely to undergo metastases. Even though chemotherapy and radiotherapy do not directly influence the tumour, they help in reducing the bulk of tumour, the adjuvant therapy also helps in reducing the spread of tumour.¹³ Orthotopic heart transplant has been tried in patients with high grade angiosarcoma but will not change long-term outcome.¹⁴

Learning points

- ▶ Cardiac angiosarcoma may present with non-specific features of dyspnoea, besides outflow tract obstructive symptoms and symptoms pertaining to metastases.
- ▶ This rare tumour can present with life-threatening emergency of cardiac tamponade.
- ▶ Importance of conducting a minimally invasive tissue sampling study in such cases of diagnostic dilemma which may give the correct diagnosis.
- ▶ Histopathological features are of paramount value in confirming the diagnosis in intracardiac mass lesions.
- ▶ In a country where tuberculosis is very common, it is important to consider diagnosis such as cardiac neoplasms in such atypical presentations.

Contributors AN and AR were actively involved in the clinical management of the case, VO assisted in getting the imaging investigations and reporting of images with clinical correlation. SA was involved in the pathological examination of the specimen.

AR came up with idea of reporting the case. AN compiled the clinical data, VO and SA did the radiology and pathology part, respectively. AN and AR did the literature search. AR gave the guidance throughout and did necessary additions.

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