A Case Report of Pericardial Inflammatory Myofibroblastic Tumor Involving the Inferior Vena Cava Causing Hepatomegaly

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Radiology Case. 2024 July; 18(7):7-13 :: DOI: 10.3941/jrcr.5371

ABSTRACT

This case report discusses a 2-year-old child presenting with pericardial inflammatory myofibroblastic tumor (IMT) involving the inferior vena cava (IVC) causing hepatomegaly, which manifested as incidental upper abdominal distension without accompanying abdominal pain, nausea, or vomiting. Preoperative imaging revealed a soft tissue mass within the pericardium and pericardial effusion, causing deformation of the left atrium, and involving the IVC causing hepatomegaly. Subsequently, she underwent a mediastinal mass excisional biopsy by thoracotomy, which uncovered a smooth, resilient mass in the pericardium closely associated with the diaphragm. The tumor exhibited a fish-flesh appearance upon sectioning, with moderate pale red bloody fluid within the pericardium. The final pathology revealed an IMT. This case recapitulates the pathological classification, imaging findings, and differential diagnosis of the rare pericardial IMT.

CASE REPORT

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The patient was a 2-year-old female with no significant medical or surgical history. She presented to our institution after noticing upper abdominal distension for over 2 months. The patient was alert but appeared chronically ill, without jaundice in the skin or sclera, and with normal superficial lymph nodes. On examination, her abdomen appeared distended with abdominal respiration, the umbilicus was normal, and palpation revealed softness without ascites, significant tenderness, or rebound tenderness, without peripheral edema. However, an enlarged left lobe of the liver was palpable in the left upper abdomen, and the right lobe of the liver could be felt about one fingerbreadth above the umbilicus.

It extended one finger breadth below the umbilicus on the right side, was firm in consistency, and had normal bowel sounds. The patient denied any abdominal pain, bloating, decreased appetite, weight loss, or family history of malignancies. She also denied any history of medication use or allergies. She had not undergone any prior abdominal imaging.

The patient was vitally stable and had the following vital signs: oxygen saturation via nasal cannula at 98%, heart rate at 85-90 bpm, blood pressure at 90/68 mmHg, and temperature at 36.5°C.

Laboratory data showed normal alpha-fetoprotein (1.46 ng/ml; normal, <8 ng/ml), elevated alpha-1 antitrypsin (2100 mg/ml; normal range, 350-1200 mg/ml), elevated alpha-1 antitrypsin antibody (3030 mg/ml; normal range, 830-1990 mg/ml), elevated hepatitis B surface antibody quantitation (875.5 IU/L; normal range, 0-10 IU/L), prolonged prothrombin time (14.4 seconds; normal range, 9.6-12.8 seconds), elevated international normalized ratio (1.31; normal range, 0.88-1.15), elevated fibrinogen (5.87 g/L; normal range, 2.0-4.0 g/L), elevated aspartate aminotransferase (39 IU/L; normal, <35 IU/L), elevated alanine aminotransferase (172 IU/L; normal range, <45 IU/L), estimated glomerular filtration rate (205.89 ml/min; normal range, 56-122 ml/min), elevated red cell distribution width (22.4%; normal range, 11.5%-14.5%), elevated platelet count (62510*9/L; normal range, 100-30010*9/L), low creatinine (20.0 umol/L; normal range, 37.0-110.0 umol/L), low hemoglobin (88 g/L; normal range, 115-150 g/L), low mean corpuscular volume (68.4 fL; normal range, 82-100 fL), and normal cardiac biomarkers.

Upon admission, the patient underwent enhanced computed tomography (CT) of the chest and upper abdomen, revealing an irregular soft tissue mass within the pericardium with significant enhancement. This mass was compressing the adjacent left atrium and invading the segment of the inferior vena cava (IVC), resulting in its occlusion. Additionally, the liver showed

enlargement, hepatic congestion, decreased enhancement in the arterial phase, uneven enhancement in the portal venous phase, and indistinct visualization of the left hepatic vein (Figure 1). Considering cardiac assessment, cardiac Magnetic Resonance Imaging (MRI) with functional enhancement was recommended, along with evaluation for signs of malignancy. Imaging showed a soft tissue mass within the pericardium with low signal on T1-weighted imaging (TIWI) and high signal on T2-weighted imaging (T2WI), measuring approximately 4.7×4.1 cm with clear borders, accompanied by pericardial effusion, encircling the IVC (Figures 2,3).

For optimal treatment planning, a multidisciplinary consultation was led by the pediatric surgery department, with participation from vascular surgery, cardiac surgery, and anesthesia departments. Vascular surgery could not rule out the possibility of a primary liver tumor. Cardiac surgery advised against performing a percutaneous biopsy to avoid the risk of bleeding leading to cardiac tamponade. Anesthesia expressed readiness for surgical intervention. Ultimately, the pediatric surgery department summarized and formulated the final plan, considering that the nature of the mass was unclear, closely related to the IVC, liver, and heart. The risk of perfect excision of the mediastinal mass during surgery was high, so the doctor team opted for biopsy by thoracotomy to establish a definitive diagnosis, guiding the next steps in treatment. Following discussions with the patient and family, the patient underwent "mediastinal mass excision biopsy by thoracotomy", revealing a smooth, resilient mass in the pericardium closely related to the diaphragm. Tissue sampling revealed a fish-flesh appearance; pericardial fluid was moderate in volume and pale red. The patient underwent routine postoperative X-ray (Figure 4). The final pathological diagnosis included immunohistochemistry: the tumor is characterized by abundant lymphocytes and plasma cells in the background, interspersed with scattered proliferating spindle-shaped cells with visible nucleoli. This is indicative of proliferative spindle cell lesions, presenting as an inflammatory mesenchymal tissue tumor. Combined with immunohistochemistry results (showing partial positivity for P53, CDK4, and MDM2 in the lesion cells, while P16 and PD-L1 are negative), the diagnosis of IMT (intermediate/ borderline tumor type) is considered. After the operation, due to the family's decision to forego further treatment, the patient did not undergo complete surgical excision and was discharged. Unfortunately, the patient passed away in the second year postoperatively due to multiple organ failure.

DISCUSSION

Demographics

The IMT, formerly known as plasma cell granuloma or inflammatory pseudotumor, most commonly occurs in soft tissues and can be found in various locations such as the lungs, heart, bladder, kidneys, and skin [1]. Primary cardiac tumors are exceedingly rare, with an incidence rate ranging from 0.001% to 0.3% in the population. Among them, 75% are benign tumors, while 25% are malignant. In children, the most common primary benign cardiac tumors are rhabdomyomas, accounting for 40%-60%, followed by fibromas at 12%-16%, teratomas at 15%-19%, and myxomas at 2%-4%. In adults, primary benign cardiac tumors are most commonly myxomas, accounting for 75%, followed by fibromas and papillary fibroelastomas at less than 10% [2]. Primary malignant tumors are predominantly sarcomas, accounting for 75% of cases, with rhabdomyosarcomas being the most common in pediatric cardiac tumors, followed by vascular sarcomas. According to the WHO classification of soft tissue tumors in 2013, IMTs are now classified under the category of vascular pericytic tumors [3]. The tumor cells are well-differentiated contractile myoid cells, classified as benign tumors. The qualitative characterization of tumor biology is intermediate (locally invasive), with histological features being benign but exhibiting invasive and recurrent behavior [4].

Pathological type

IMT has an insidious onset and can occur across all age groups, predominantly affecting children to young adults [5]. The etiology and pathogenesis of IMT are still unclear, although recent studies suggest a possible association with anaplastic lymphoma kinase (ALK) gene rearrangement. The ALK locus is located on 2p23, with approximately 40%-60% of IMTs showing ALK positivity. However, only around 10% of cardiac IMT patients are ALK positive, and generally, ALK-positive patients have a better prognosis [2]. IMT lesions primarily consist of spindle-shaped myofibroblasts, fibroblasts, and inflammatory cells. Cofn et al [6]. categorized IMTs into three histological types based on their observations of 84 cases of extrapulmonary inflammatory myofibroblastic tumors: Mucoid type: characterized by loose arrangement of tumor cells in edematous, mucoid-like background, rich in blood vessels, with infiltration of various inflammatory cells, resembling granulation tissue and nodular fasciitis. Spindle cell dense type: characterized by densely arranged spindle-shaped tumor cells in bundles, whirls, or herringbone patterns, with varying degrees of mucoid or collagenous areas, clustered lymphocytes and plasma cells interspersed between spindle-shaped tumor cells, resembling fibromatosis or fibrous histiocytoma. Fibrous type: characterized by sparse spindle-shaped cell components, with lymphocytes and plasma cells infiltrating between transparent degenerated collagen fibers, resembling fibrous histiocytoma or scar tissue.

Imaging Characteristics

Due to the varying content of inflammatory infiltration and fibrosis, IMT presents with diverse imaging manifestations but lacks specificity. On ultrasound, IMT typically appears as a solid mass with clear or unclear borders, showing heterogeneous hypoechoic or hyperechoic patterns, with possible calcifications or central hypoechoic necrotic areas. CT imaging may reveal low/equal density masses with clear or unclear borders, occasionally showing calcifications. Different proportions of fibrosis and inflammatory components within the lesion result

in varied appearances, often demonstrating delayed and uneven enhancement (related to internal fibrous tissue) upon contrast enhancement.[7] On MRI images, IMT tends to exhibit low signal on T1WI and high signal on T2WI compared to skeletal muscle. When the tumor contains a large amount of fibrous matrix, it appears as low signal on both T1WI and T2WI. Enhancement scans show uneven enhancement, with delayed enhancement of fibrous components [8].

Differential Diagnosis

There are several entites which can potentially mimic the diagnosis of an IMT within the pericardium. These include rhabdomyomas, mesotheliomas, solitary fibromas, neurofibromas, and sarcomas.

Rhabdomyomas

When visualized on contrast-enhanced CT scans, typically present as low-density masses with minimal enhancement. On MRI, they exhibit a smooth surface, appearing isointense to myocardium on T1-weighted images or slightly hyperintense, and slightly hyperintense on T2-weighted images. In contrast to pericardial IMTs, rhabdomyomas tend to display significant delayed and uneven enhancement [9].

Mesotheliomas

Mesotheliomas manifest as pericardial effusions with thickening of the pericardial sac wall, early-stage irregularly enhancing masses, prone to cystic degeneration, necrosis, hemorrhage, and rich vascularity. They affect both the wall and visceral layers of the pericardium, with adjacent blood vessels and tissues often compromised. In contrast to pericardial IMTs, mesotheliomas typically exhibit delayed-phase enhancement [10].

Solitary Fibromas

Solitary fibromas may exhibit calcifications on chest X-rays and appear as homogeneous or heterogeneous enhancing masses on contrast-enhanced CT. On MRI, they display heterogeneous T1 and T2-weighted signals, with significant delayed enhancement and sometimes central enhancement attenuation, occasionally showcasing a serpentine vascular pattern. Unlike pericardial IMTs, fibromas show fewer internal blood vessels [11].

Sarcomas

Sarcomas can present as linear pericardial thickening or diffusely infiltrative masses, often larger than 10 cm in diameter, with indistinct borders, heterogeneous density, commonly accompanied by hemorrhagic pericardial effusion and necrotic tumor fragments. They exhibit notably heterogeneous enhancement, invasive and highly vascularized masses, with prominent vascular structures within. Compared to pericardial IMTs, sarcomas demonstrate a lower prevalence of internal blood vessels [12].

CONCLUSION

Once diagnosed, pericardial IMTs necessitate prompt treatment to prevent life-threatening complications. When involving cardiac valves or coronary arteries, sudden death is a possibility. Surgical intervention remains the primary treatment modality for IMTs. ALK-positive patients may benefit from targeted gene therapies, showcasing clear efficacy, while corticosteroids and anti-inflammatory agents can also be utilized.

In this case, since the medical team considered the nature of the mass to be unclear and its close relationship with the IVC, liver, and heart, the risk of surgically excising the mediastinal mass was deemed high. Therefore, the medical team opted for biopsy by thoracotomy to establish a definitive diagnosis and to maintain the patient's cardiac function as much as possible.

TEACHING POINT

Pericardial IMTs represent a rare diagnostic challenge, often accompanied by pericardial effusion and compression of the left pericardium, causing hepatomegaly. Timely treatment upon IMT confirmation is crucial to prevent fatal complications.

Authors' contributions

Xiaoqin Chen: writing original draft (lead); writing-review and editing (equal).

Guangxin Chen: writing-review and editing (equal).

Liqing Peng: conceptualization (lead); writing-review and editing (lead).

Disclosures

We are grateful for the funding provided by the Mudanjiang Medical University Science Foundation Torch Program Project (2022-MYHJ-007).

There are no conflicts of interest.

Consent

We obtained written informed consent from the patient for submission of this manuscript for publication.

Human and animal rights

This study does not involve.

QUESTIONS

QUESTIONS

Question 1: Which of the following statements regarding the pericardium is incorrect?

A. The pericardium can normally communicate with the outside environment. (applies)

B. The visceral and parietal layers are separated by less than 50 ml of fluid.

C. The pericardium originates from major blood vessels and envelops the heart cavity.

D. The pericardium covers part of the left atrium.

E. Normal measurement of the pericardium on CT and MRI does not exceed 2mm.

Answer 1: The pericardium can normally communicate with the outside environment. (The pericardium is a closed cavity that does not normally communicate with the outside environment.)

Question 2: Which of the following statements regarding pericardial tumors is incorrect?

A. Pericardial metastatic tumors outnumber primary tumors by 20-40 times.

B. Post-mortem examinations reveal pericardial metastases in approximately 10% of all malignant tumors.

C. The most common malignant tumors originate from the liver. (applies)

D. Colon tumors can metastasize to the pericardium.

E. Lymphomas can be found in the pericardium.

Answer 2: The most common malignant tumors originate from the liver. (The most common malignant tumors originate from the lungs.)

Question 3: Which of the following statements regarding pericardial IMT is incorrect?

A. CT can show low/equal density masses.

B. The borders of the tumor can be clear or unclear with infiltrative features.

C. Calcifications can be observed.

D. Delayed and uneven enhancement is seen.

E. All of the above. (applies)

Answer 3: All of the above. (The imaging manifestations of pericardial IMT are varied.)

Question 4: What are the complications of pericardial IMT?

A. Pericardial effusion

B. Compression of the atria

C. Pericardial hemorrhage

D. Pericardial tamponade

E. All of the above (applies)

Answer 4: All of the above. (Prompt treatment of pericardial IMT is necessary to prevent fatal complications.)

Question 5: Which of these statements is false?

A. Complete excision is the preferred treatment for pericardial IMT.

B. If the patient is intolerant surgery by thoracotomy, a needle biopsy can be performed.

C. During surgery, attention should be paid to prevent pericardial hemorrhage and tamponade.

D. The surgical approach for pericardial IMT involves complete excision of the tumor.

E. There is a risk of recurrence after surgery for pericardial IMT.

Answer 5: The surgical approach for pericardial IMT involves complete excision of the tumor. (The surgical approach should be tailored according to the patient's condition.)

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FIGURES

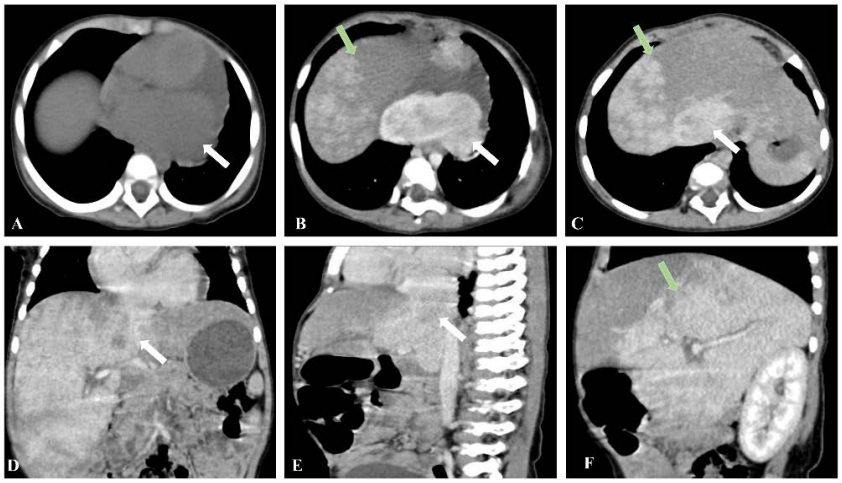

Figure 1: Contrast enhanced CT through level of IMT (1A: Axial section of Plain phase; 1B: Axial section of Arterial phase; 1C: Axial section of Portal venous phase; 1D: Coronal section of Portal venous phase; 1E: Sagittal section of Portal venous phase; 1F: Another sagittal section of Portal venous phase)
A soft tissue mass is observed within the pericardium (white arrowhead), showing persistent enhancement and extending downward to the IVC, with uneven enhancement of the liver (green arrowhead)

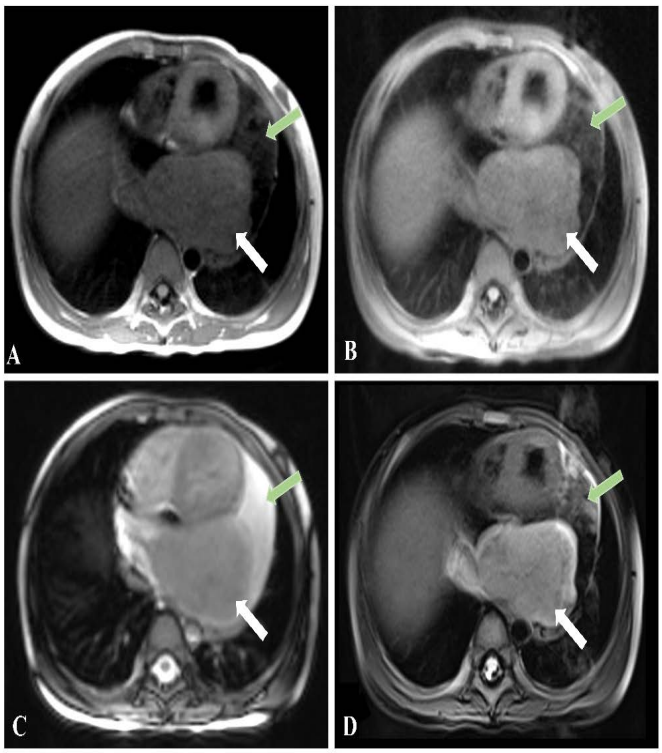

Figure 2: Axial section of MRI through level of IMT
(A: T1WI- Double IR Sequence; B: TIWI - Double IR+FATSAT Sequence;
C: T2WI Sequence; D: T2WI - Double IR+FATSAT Sequence)
A soft tissue mass is observed within the pericardium (white arrowhead), accompanied by pericardial effusion (green arrowhead)

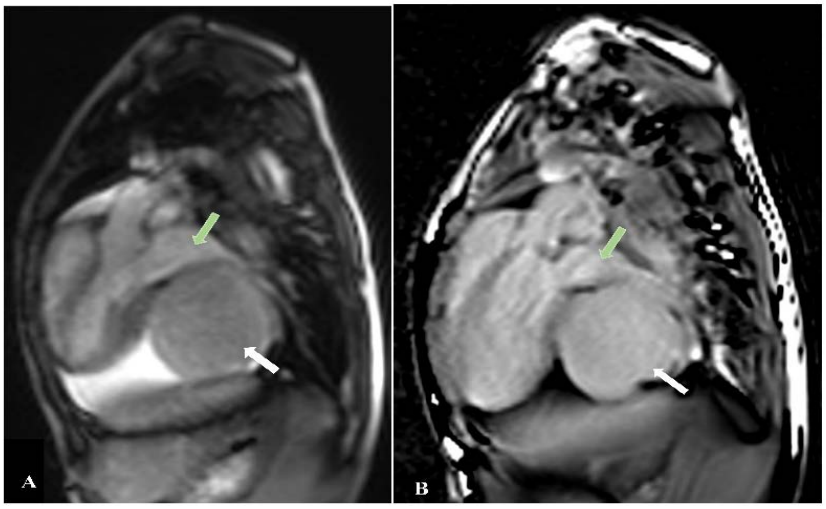

Figure 3: Sagittal section of cardiac magnetic resonance imaging (MRI)
(A: Fiesta Sequence; B: 2D-MDE Sequence)
A mobile mass in the pericardium (blue arrowhead) with compression of the left atrium (green arrowhead)

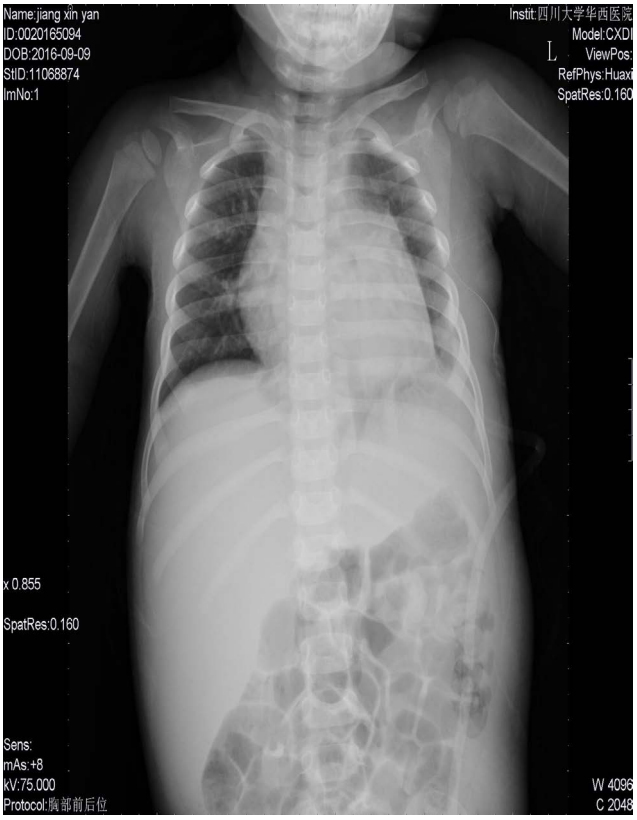

Figure 4: Postoperative X-ray image of the patient
The lung textures are slightly blurred, and the left costophrenic angle is indistinct, suggesting a small amount of effusion. A tubular shadow is observed in the left abdominal area

KEYWORDS

Pericardial Inflammatory Myofibroblastic tumor; Computed tomography; Magnetic Resonance Imaging; Inferior vena cava; Hepatomegaly