Giant primary pericardial synovial sarcoma in an adolescent with cardiac tamponade: Contrast-enhanced CT and ^{18}F-FDG PET/CT findings

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INTRODUCTION

Primary pericardial synovial sarcoma (SS) is an extremely rare and high-grade malignancy that presents a diagnostic challenge in clinical practice.[1] We report the case of an adolescent who exhibited primary pericardial SS with cardiac tamponade on contrast-enhanced chest CT and ^{18}F-FDG PET/CT.

CASE SUMMARY

A 16-year-old man was admitted to our hospital because of progressive worsening dizziness and a 2-month history of chest tightness and shortness of breath after exercise. The physical examination at admission was notable for neck vein distention and diminished heart sounds on cardiac auscultation. The chest x-ray

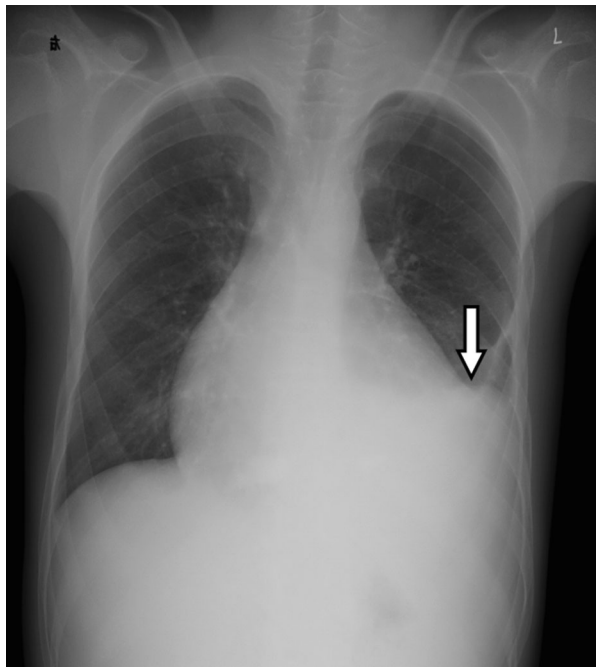

Figure 1. Chest x-ray (PA view) showing cardiac enlargement and left pleural effusions (white arrow).

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showed cardiomegaly and left pleural effusions (Figure 1). A transthoracic echocardiogram in an external hospital revealed a large mass in the pericardial cavity. The chest and upper abdomen CT showed bilateral pleural effusions and a lobulated mass in the pericardium, as well as massive pericardial effusion causing the so-called ''flattened heart sign'',[2] ''periportal halo sign'',[3] and dilation of the superior vena cava and inferior vena cava; these findings suggested cardiac tamponade (Figures 2, 3). Thus, a malignant pericardial tumor was suspected. To further evaluate the pericardial lesion and perform staging, ^{18}F-FDG PET/CT was performed; it revealed multiple foci of ^{18}F-FDG-avid lesions in the central solid component of the mass (Figure 4). Subsequently, the patient underwent tumorectomy; the final histopathological diagnosis was primary pericardial SS, monophasic type (Figure 5). Primary pericardial SS is an aggressive, high-grade malignancy characterized by local invasiveness and a tendency to metastasize; thus, it presents a significant challenge in clinical practice.[1] Pericardial tumors are frequently categorized as primary and secondary tumors in clinical practice. Secondary tumors are clearly diagnosed and the primary site of origin can be detected by ^{18}F-FDG PET/CT. However, the diagnosis of primary pericardial tumors is difficult and requires comprehensive analysis with multiple imaging modalities.

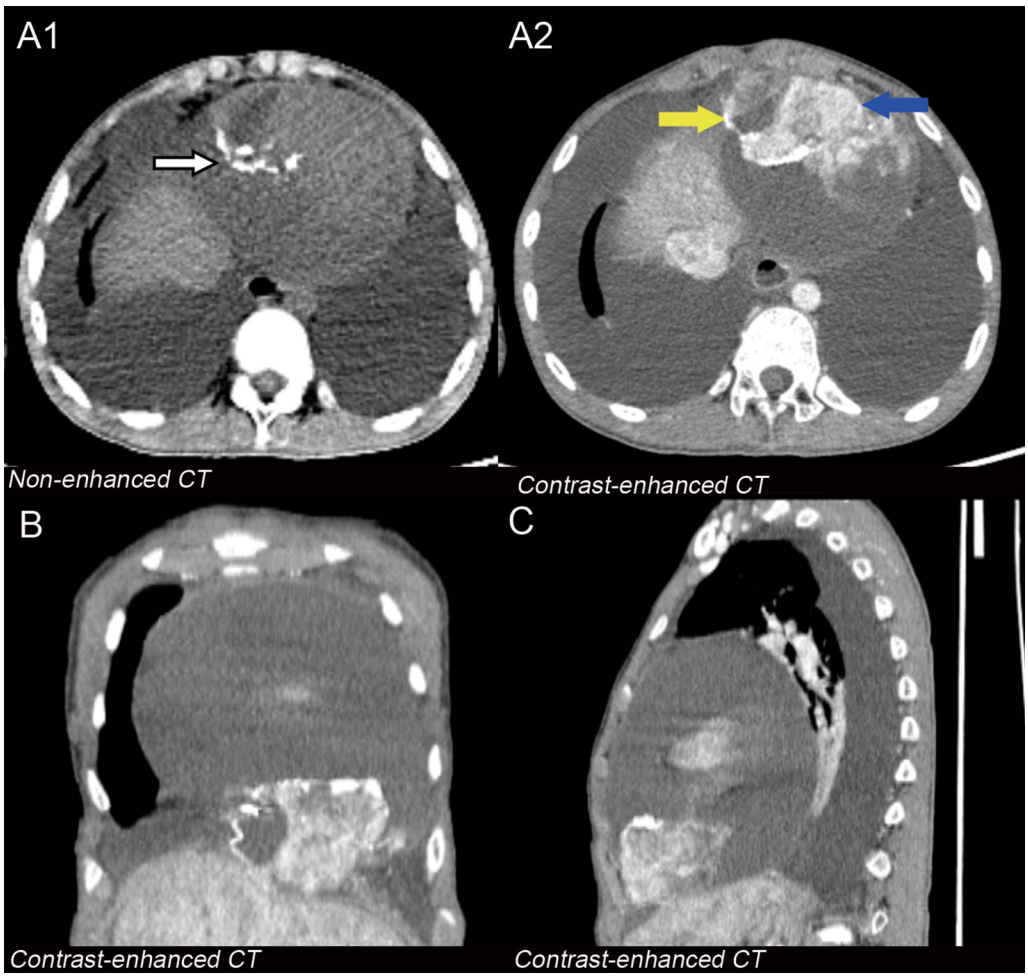

Figure 2. Computed tomography image of the chest in transverse (**A**), coronal (**B**), and sagittal (**C**) views: Non-enhanced CT (**A1**) showing an ill-defined mass with irregular calcifications (white arrow) in the pericardium, bilateral pleural effusions, and massive pericardial effusion. In contrast-enhanced CT, transverse view (**A2**) demonstrated a mass with cystic degeneration (yellow arrow) and solid components with moderate enhancement (blue arrow). Coronal (**B**) and sagittal (**C**) views showing that the mass originated from the diaphragmatic portion of the pericardium, which was separated from the myocardium.

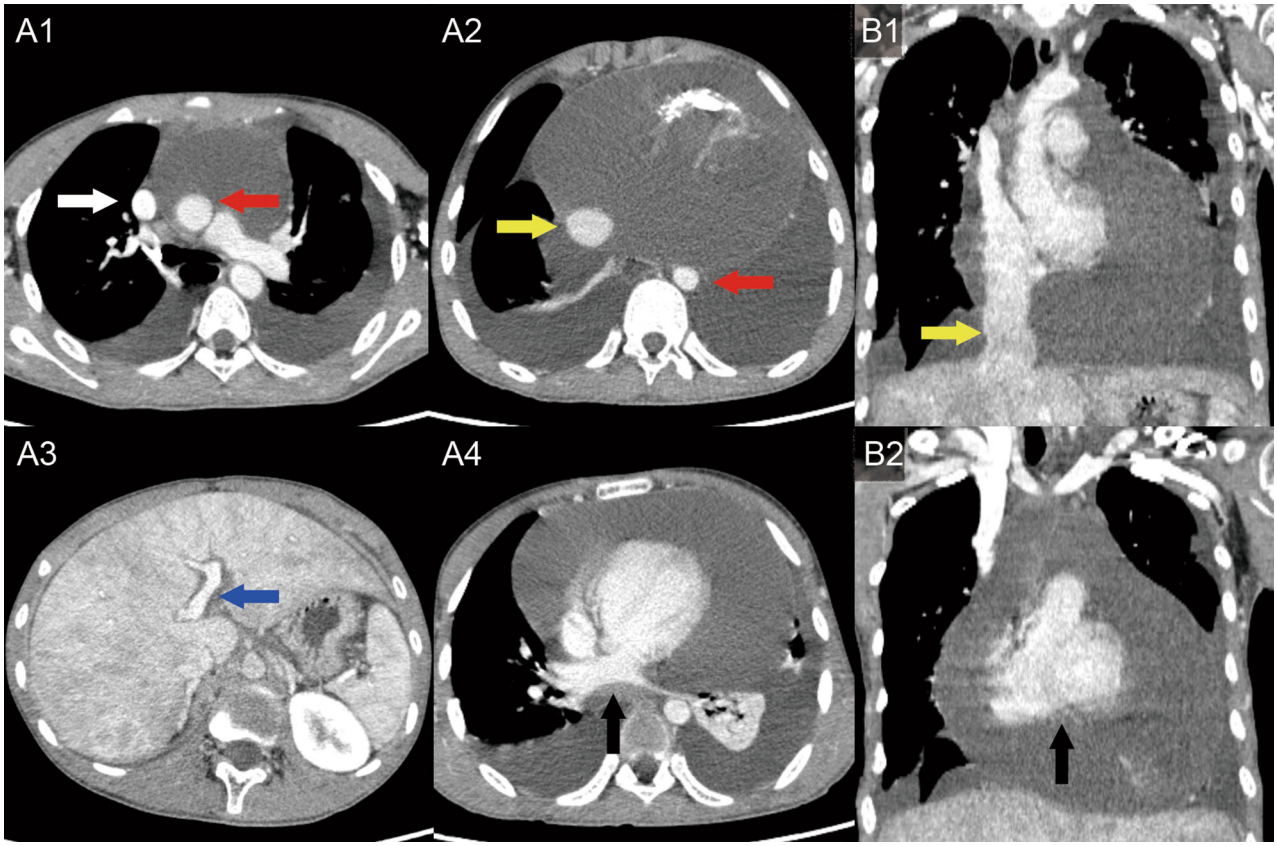

Figure 3. Contrast-enhanced computed tomography images of the chest and upper abdomen in the transverse (**A**) and coronal (**B**) views. Transverse view (**A1**) showing dilated superior vena cava (white arrow) with a diameter similar to that of the adjacent aorta (red arrow). Transverse (**A2**) and coronal (**B1**) views showing marked dilation of the inferior vena cava (yellow arrow), which exhibited a diameter greater than twice that of the adjacent aorta (red arrow). Transverse (**A3**) view of the upper abdomen showing low attenuation around the portal veins (blue arrow), due to distended periportal lymphatic vessels; this manifestation is known as the ''periportal halo sign.'' Transverse (**A4**) and coronal (**B2**) views showing concave deformity of the posterior wall of the left atrium (black arrow) and the inferior surface of the heart (black arrow), due to compression; this manifestation is the so-called ''flattened heart sign.''.

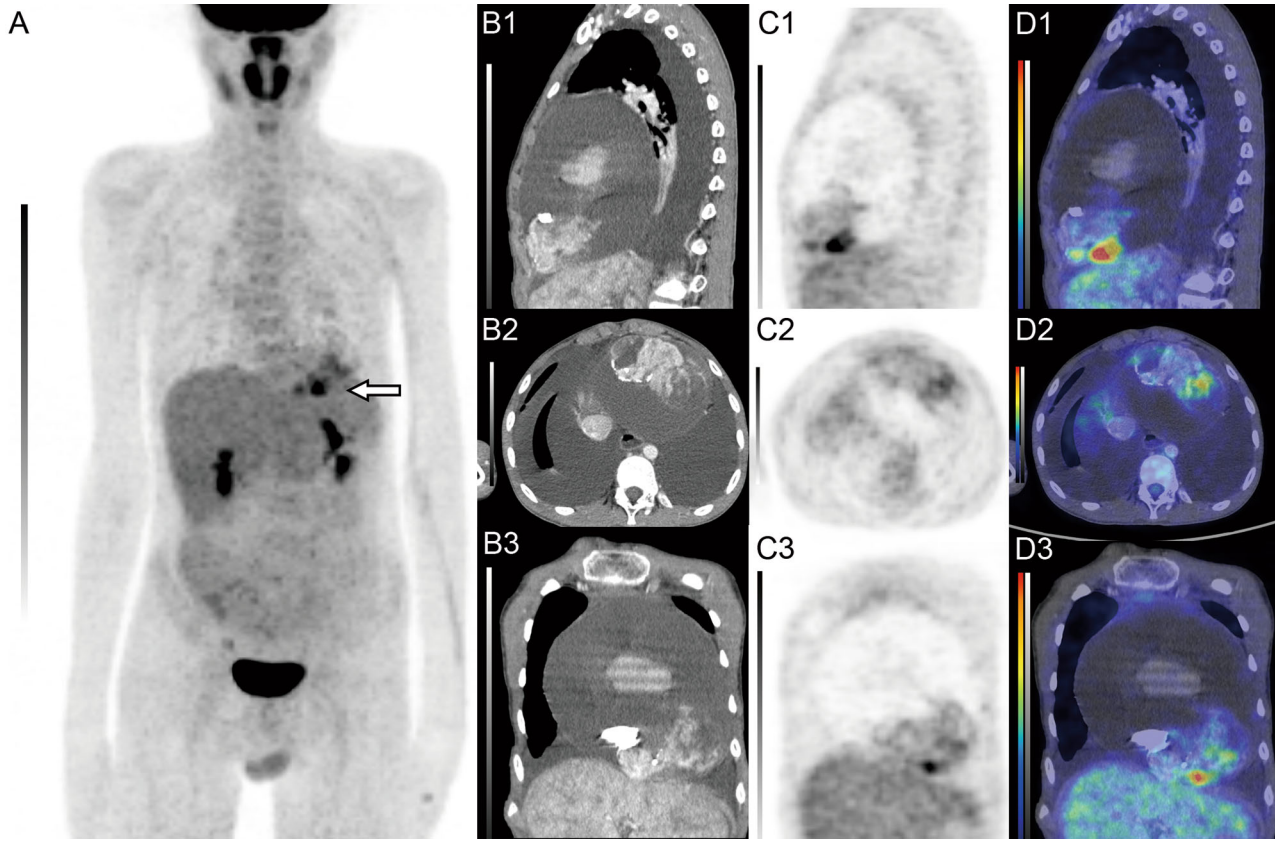

Figure 4. MIP PET image (**A**), contrast-enhanced CT (**B**), ^{18}F-FDG PET (**C**), and fusion images (**D**). MIP PET image showing multiple foci of small ^{18}F-FDG-avid lesions (white arrow, SUV_{max} 4.0) in the region of the heart. Sagittal (1), transverse (2), and coronal (3) views showing multiple foci with lesions exhibiting mild to moderately increased activity in the central solid component of the mass.

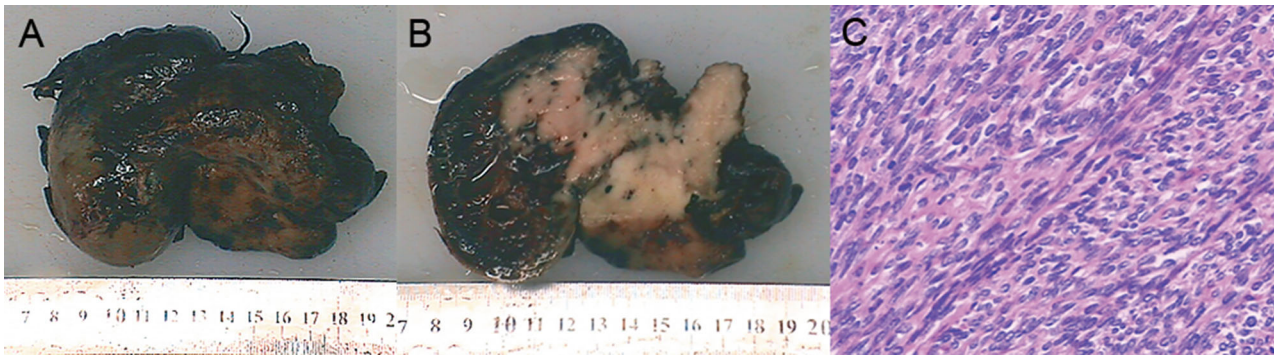

Figure 5. Gross image (**A**): The surgical specimen measured 12.0 × 8.0 × 7.0 cm, with a fibrous capsule. Image of cut surface (**B**): The cut surface was grayish white to yellow and mottled with dark red, exhibiting cystic degeneration and foci of ossification. The histopathological image (**C**) showing uniform spindle-shaped tumor cells with nuclear pleomorphism and mitoses (hematoxylin and eosin stain, magnification, × 400).

Disclosure

No conflict of interest and no disclosures.

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