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Original Article

Primary intracardiac leiomyoma arising from cardiomyocyte progenitors at the right ventriculoseptal interface: case report with literature review☆

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ABSTRACT

Primary cardiac neoplasms are rare and are usually benign myxomas and rhabdomyomas. Cardiac leiomyomas are usually seen as a part of the spectrum of intravenous leiomyomatosis or benign metastasizing leiomyoma. De novo occurrence of primary intracardiac leiomyomas (PICL) is a rarity. Herein we describe a 14-year-old boy presenting with intermittent dyspnea for 2 years, with a large right ventricular mass suggestive of myxoma on transthoracic echocardiography, without any extracardiac lesions. Histology and immunohistochemistry of the tumor excised under cardiopulmonary bypass confirmed a PICL arising at the cardiomyocyte–smooth muscle septal interface. A review of existing literature highlights an increased incidence in young patients and an overwhelming right ventricular anatomical predilection. Abnormalities in the multipotent cardiac progenitor cells of the second heart field may provide a potential microenvironment for the histogenesis of PICL.

1. Introduction

Primary cardiac neoplasms are rare and account for an incidence of only 0.001% to 0.03% in autopsy series [1]. Most are benign tumors, myxomas being the commonest, while rhabdomyomas are known to have higher incidence among children [2]. Primary intracardiac leiomyomas (PICLs) are exceptionally rare with only few prior reports [3–6]. We describe a case of PICL arising at the cardiomyocyte–smooth muscle septal interface in a young boy and analyze the possible histogenesis of this uncommon entity. Existing literature on the subject has also been reviewed in an attempt to elucidate the biological characteristics of this tumor.

2. Case report

A 14-year-old Asian boy presented with a history of intermittent episodes of breathlessness and palpitations for 2 years. No history of fever, weight loss, or growth retardation was elicited. There was no significant past or treatment history. Examination revealed normal vital parameters. A systolic murmur over the pulmonary area with a loud first heart sound (S1) and an apical mid diastolic murmur was appreciated. Laboratory investigations found unremarkable biochemical and hematological parameters. Chest X-ray showed cardiomegaly with clear lung fields. Electrocardiogram (ECG) showed an enlarged right atrium, left posterior fascicular block, and anterolateral ischemia. Transthoracic echocardiography demonstrated a large right ventricular (RV) mass attached to the interventricular septum and associated with moderate tricuspid valve stenosis and regurgitation (Fig. 1). Right atrial pressure was elevated, and right atrium and inferior vena cava were dilated. Further imaging studies (whole body computed tomography/positron emission tomography) did not reveal any other lesions elsewhere in the body.

Excision of the intracardiac mass was planned, and peroperatively, a large RV mass arising from the membranous interventricular septum, involving the tricuspid leaflets and right atrium, was noted. Anterior and septal leaflets were involved by the mass. On opening the RV, the firm encapsulated, globular mass measuring 6×6 cm was seen adherent to the RV free wall and arising from the membranous part of the interventricular septum. The lesion was approached via a median

Abbreviations: PHT, primitive heart tube; PICL, primary intracardiac leiomyomas; SHF, second heart field.

☆ Ethical adherence: The present work was performed after taking informed consent from the patient, and a sincere effort has been made to uphold patient confidentiality.

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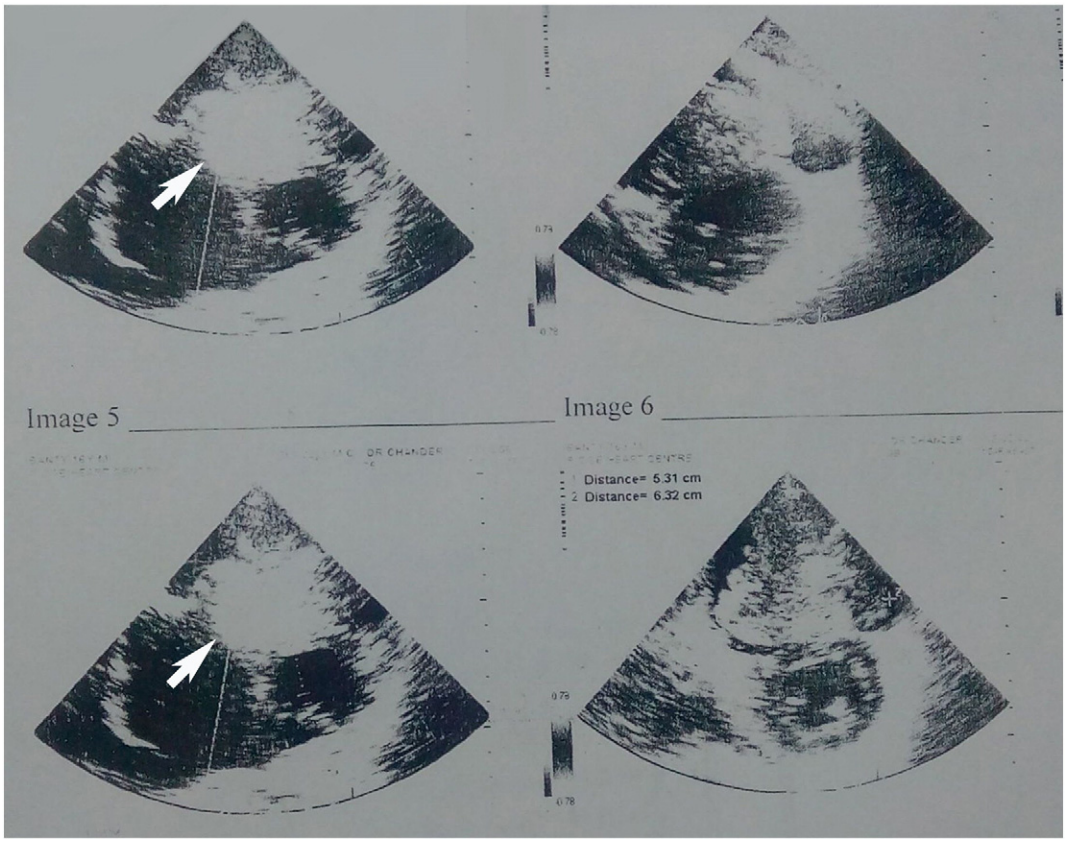

Fig. 1. Transthoracic echocardiography images showing a large mass (white arrows) of uniform consistency, present in the right ventricle, prolapsing into the right atrium; the mass appeared to be attached to atrioventricular septum.

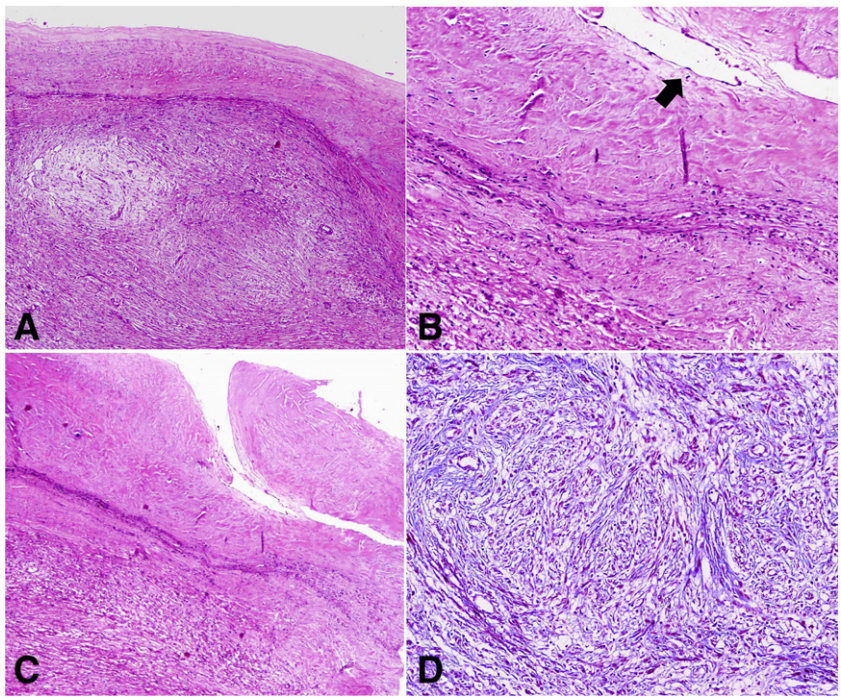

Fig. 2. (a) Low-power photomicrograph showing a spindle cell neoplasm admixed with blood vessels beneath the endocardium [hematoxylin and eosin (H&E), 40×]. (b and c) Overlying endocardial lining is also visualized (black arrow; H&E, 100× and 40×). (d) Masson's trichrome stain highlights smooth muscle cells admixed with collagen bundles (blue; Masson's trichrome, 100×).