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Inflammatory pseudotumor of the right atrium

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Abstract Inflammatory pseudotumor of the lung was first described by Brunn in 1939. Since that description, various extrapulmonary sites of inflammatory pseudotumor have been described. Review of the literature reveals five cases of inflammatory pseudotumor involving the heart, but no cases have been reported in the radiology literature. The present case involves a 7-month-old girl with inflammatory pseudotumor involving the right atrium, which was completely excised at surgery. Though rare, inflammatory pseudotumor should be considered in the differential diagnosis of cardiac tumors in children.

Case report

A 7-month-old girl with no significant past medical history initially presented with pallor, tachypnea and decreased oral intake. Blood gas analysis on room air showed a pO_2 of 80. Workup included laboratory evaluation for sepsis, which was negative, as well as chest X-ray, echocardiogram and cardiac MRI. The chest X-ray was normal. The echocardiogram showed a right atrial mass, abutting the tricuspid valve, and further evaluation with cardiac MRI showed a relatively homogeneous mass occupying most of the right atrium, contiguous with the inferolateral wall, with a clear plane of separation from the interatrial septum (Fig. 1a–c).

The mass was surgically resected; it was found to be based at the right atrioventricular junction just adjacent to, but not including the tricuspid valve. In the process of removing the mass, a 3 × 3-cm defect was created in the inferior wall of the right atrium, which was repaired with a patch of bovine pericardium.

Histopathological examination of the resected right atrial mass showed cellular spindle cell proliferation with scattered foci of inflammatory cells. No cytological atypia was found and the diagnosis was inflammatory pseudotumor of the right atrium.

Discussion

Inflammatory pseudotumor was first described in the lung by Brunn in 1939 and was so named by Umiker and Iverson in 1954 [1]. Inflammatory pseudotumor has been described in the literature by various names: plasmocytoma, plasma cell granuloma, xanthoma, xanthogranuloma, fibrous histiocytoma, solitary mast-cell granuloma and fibroxanthoma. The extrapulmonary sites of inflammatory pseudotumor reported in the literature include stomach, liver, pancreas, kidney, adrenal, retroperitoneum, bladder, thyroid, tonsil, fourth ventricle, spinal cord meninges and central nervous system [2]. The first description of cardiac inflammatory pseudotumor was by Gonzalez-Crussi et al. in 1975 [3]. Of the five cases described in the literature, the youngest was a 3-month-old baby and the oldest a 17-year-old boy. Cardiac sites involved by inflammatory pseudotumor included right and left atrium, right and

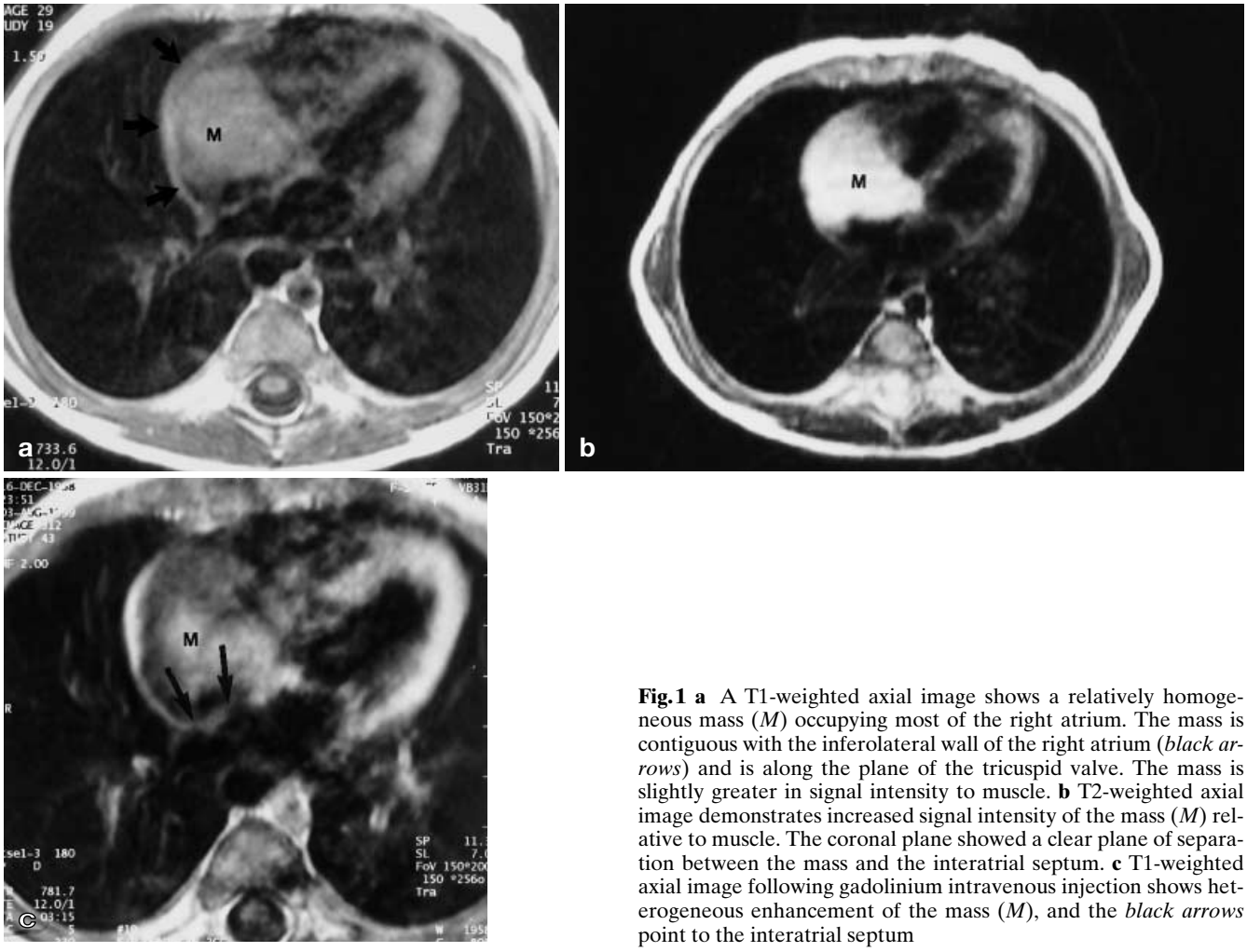

Fig. 1 a A T1-weighted axial image shows a relatively homogeneous mass (*M*) occupying most of the right atrium. The mass is contiguous with the inferolateral wall of the right atrium (*black arrows*) and is along the plane of the tricuspid valve. The mass is slightly greater in signal intensity to muscle. **b** T2-weighted axial image demonstrates increased signal intensity of the mass (*M*) relative to muscle. The coronal plane showed a clear plane of separation between the mass and the interatrial septum. **c** T1-weighted axial image following gadolinium intravenous injection shows heterogeneous enhancement of the mass (*M*), and the *black arrows* point to the interatrial septum

left ventricle, pulmonary and tricuspid valves, septum, right and left coronary arteries, superior vena cava, coronary sinus, and the atrioventricular groove.

In general, primary cardiac tumors are rare in children, with a prevalence of less than 1% at autopsy. More than 70% are benign, in the form of rhabdomyoma (seen in patients with tuberous sclerosis), fibroma, or myxoma.

The etiology of inflammatory pseudotumor is unknown; it may represent a low-grade neoplasm of fibroblasts with inflammatory reaction. This entity has been described in association with other malignancies. It can be a result of inflammation following trauma or surgery. In one patient, inflammatory pseudotumor was associated with vasculitis and inferior vena caval thrombosis, with anti- C3 and anti-fibrinogen deposits in the vessel wall, suggesting an immune/ autoimmune mechanism [4].

The role of cardiac MRI in evaluation of a possible mass includes confirming the presence of a mass. With its multiplanar capability, MRI can delineate accurate localization of the mass and is able to demonstrate motion of the mass with respect to adjacent cardiac structures. It can be used to characterize the composition of the mass and is useful in differentiating tumor from thrombus.

Differences in MR signal characteristics are helpful in distinguishing between fibroma, lipoma, and rhabdomyoma. Fibromas have a signal intensity lower than that of normal myocardium, and lipomas typically have higher T1 signal intensity than myocardium. The MRI signal-intensity pattern is nonspecific for rhabdomyoma, but in general shows a mass of similar or higher signal intensity compared with adjacent myocardium on short TE images, and isointensity with adjacent myocardium on long TE images.

Gradient refocused echo (GRE) imaging is more sensitive for differentiation of intracardiac thrombi than spin-echo imaging. Thrombi (except for hyperacute thrombi) show lower signal intensity than myocar-

dium on GRE images; tumor generally has increased signal on GRE images. Atrial myxomas are the exception, as they have also been shown to have lower signal intensity than adjacent normal myocardium on GRE images secondary to areas of hemorrhage, hemosiderin, and foci of calcification. Gadolinium-contrast enhancement of tumors may also play an important role in distinguishing a thrombus from tumor [5, 6].

The biological potential of inflammatory pseudotumor is highly variable, but it generally pursues an innocuous course with 25% local recurrence. Rare distant metastases have been reported [7]. Complete surgical resection, if possible, is the treatment of choice. Several reports of spontaneous regression in unresectable cases have been reported. Radiation therapy has been tried in unresectable cases. Chemotherapy has no role and response to steroids is unpredictable.

In conclusion, although rare, inflammatory pseudotumor should be considered in the differential diagnosis of masses in the heart in children. Although the natural history of inflammatory pseudotumor is quite variable, behavior is usually benign, with local recurrence being the most common complication. Surgical excision is the primary method of treatment.

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