Inflammatory Pseudotumor of the Heart with Vasculitis and Venous Thrombosis*

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Inflammatory pseudotumor (IPT) is a tumor-like reactive lesion of unknown etiology. An unusual case of intracardiac IPT with multisystemic involvement, including leukocytoclasmic vasculitis, polyarthritis, and inferior vena cava thrombosis in a 17-year-old boy is reported. This unique combination may suggest that immune/autoimmune factors are important in the pathogenesis of IPT.

FIGURE 1. Two-dimensional echocardiogram shows two masses in the right atrium (M = mass; RA = right atrium; RV = right ventricle; TV = tricuspid valve).

FIGURE 2. Computed tomography shows low-density space-occupying lesion in the right atrium of the heart (M = mass; RA = right atrium; RV = right ventricle; LV = left ventricle).

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Inflammatory pseudotumor (IPT), a tumor-like reactive lesion, is an uncommon disorder of unknown etiology. In the majority of the reported cases, the disease was confined to the lungs. Other cases were found in the liver, stomach, lymph nodes, and other organs as well. Cardiac involvement was reported only twice, in 1975 and in 1988. We report the first case in the literature (to our knowledge) of intracardiac IPT associated with multisystemic involvement, including leukocytoclasmic vasculitis, polyarthritis, and inferior vena cava thrombosis.

CASE REPORT

A 17-year-old boy was admitted to the hospital because of fever, pain, and swelling of ankle, mouth ulcers, and a nonpruritic eruption over the heels, for a week's duration. He had been well until a year prior to hospital admission, when he began to suffer from intermittent painful swelling of several joints, responding to symptomatic treatment.

The patient appeared pale and sick, the temperature was 39°C, blood pressure was 110/60 mm Hg, and the pulse was regular, 80/ min. An erythematous-purpuric maculopapular eruption was seen over the heels. Aphthous ulcers were found in the mouth. Elastic, nontender cervical lymph nodes were palpated. The lungs were clear. The heart was not enlarged. The first and second sounds were normal. A middiastolic click was heard over the fourth intercostal parasternal space, and a blowing 2/6 systolic murmur was heard over the mitral valve area. The tip of a nontender spleen was palpated. The liver was not clinically enlarged. The left ankle was swollen, warm, and tender. Results of neurologic examination were normal.

The ESR was 40 mm/h. Hematocrit was 35 percent, white blood cell count was 13,400/cu mm with a normal differential count, and platelet count was normal. Results of blood chemistry, serologic, and immunologic studies were all normal. There was no laboratory evidence for a hypercoagulable state. Repeated blood, urine, and throat cultures were negative.

An electrocardiogram showed sinus rhythm, left atrial enlargement, and an incomplete left bundle branch block. Roentgenograms of the chest were normal. A two-dimensional echocardiogram, confirmed by computed tomographic (CT) scan, showed two masses in the right atrium: one attached to the interatrial septum and the

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Pathologic lymphocytes, and inflammatory cells were prominent in the atrial muscle fibers. The inflammatory infiltrate consisted of numerous foam cells as well as lymphocytes, plasma cells, eosinophils, and occasional multinucleated foreign body type giant cells. Focal neutrophil accumulations and very occasional hemosiderin-laden macrophages were observed. There was no atypia or marked mitotic activity (Fig. 3).

Immunohistochemical staining for immunoglobulins revealed a polycyclic pattern. Capillary and fibroblastic proliferation were prominent in a mildly myxoid background showing diastase-resistant-PAS and Alcian-blue positivity. The blood vessels within the lesion showed no significant abnormality. The submitted tricuspid valve showed no histologic abnormalities.

A diagnosis of IPT was made.

Discussion

IPT is a tumor-like reactive lesion that has many synonyms such as plasma cell granuloma, post inflammatory tumor and xanthomatous pseudotumor.1,2,8,11 During the last five decades, many cases of IPT were reported, most commonly in the lungs as well as in the liver, stomach, lymph nodes, orbit, and in other organs.6,8 Inflammatory fibrosclerosing lesions, eg, idiopathic sclerosing mediastinitis and retroperitoneal fibrosis, have occasionally been entered into this spectrum.2,13

The histologic findings described above are compatible with those described in the literature.1,2,8,9,11 Despite the gross tumor-like presentation, the lesion appears to be nonneoplastic due to the polymorphism of the infiltrate associated with capillary and connective tissue proliferation.

The etiology and pathogenesis of IPT are not known. Several mechanisms have been proposed, including immunologic,1,2,8,9,14 inflammatory,8 and infectious.11 In several cases, IPT was associated with inflammatory and oblitative changes of vessel walls within the lesion and other sites as well.2,4,5,13 The vascular oblitative changes, possible effects of interleukin 1 and fibrosclerosis secondary to methysergide therapy,10,18 have been contemplated.

Only two cases of intracardiac IPT were reported. In 1975, Gonzalez-Crussi et al described the first case associated with constitutional and systemic manifestations. The second case was reported by Pearson et al in 1988.

To our knowledge, we describe the first case in the literature of intracardiac IPT that also had vasculitis and inferior vena cava thrombosis. This unique combination and the demonstration of anti-C3 and antifibrinogen deposition in the vessel walls, as well as vasculitic and perivascular infiltrate, may highlight the process, suggesting that immune/autoimmune mechanisms are important in the development of IPT.

References


Figure 3. Polymorphous inflammatory infiltrate in cardiac inflammatory pseudotumor (IPT) (hematoxylin-eosin, ×200).