Recurrent Intracardiac Juvenile Xanthogranuloma in an Adult

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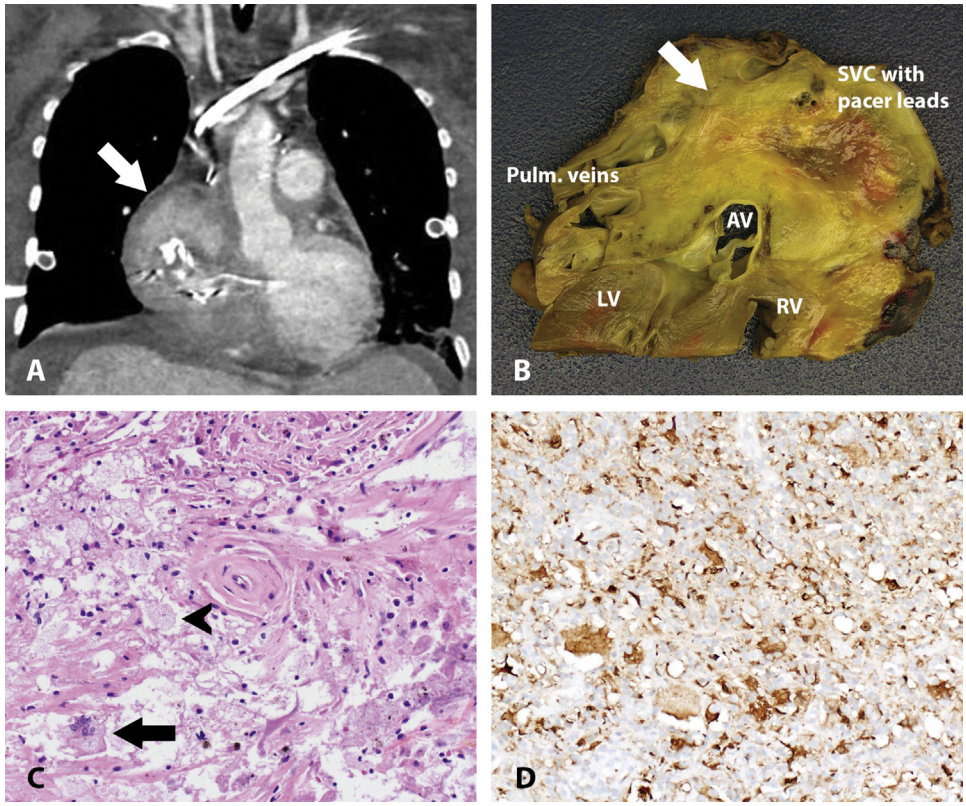

Fig 1.

A 50-year-old woman with fatigue and mitral regurgitation was admitted to the hospital for mitral valve surgery. Eighteen years previously, she had a 9-cm mass removed from inside her heart. More recently, intraoperative dissection revealed a firm inoperable mass encasing the right atrium (Fig 1A, arrow). After an iatrogenic preoperative superior vena cava tear, she passed away 2 weeks later from hypotensive multiorgan complications. At autopsy a 10.5-cm yellow-tan mass involved both atria and the left ventricle, with near obliteration of the right atrial chamber (Fig 1B, arrow; AV = aortic valve; LV = left ventricle; Pulm. = pulmonary; RV = right ventricle; SVC = superior vena cava). Microscopic sections of the mass showed dense sheets of foamy histiocytes (Fig 1C, arrowhead, original magnification ×200) and rare Touton-type giant cells (Fig 1C, arrow), identical to the mass previously removed. The histiocytes stained with factor XIIIa (Fig 1D, original magnification ×200), CD11c, and CD68; they did not stain with CD1a, S-100, or BRAF V600E. This scenario is consistent with recurrent juvenile xanthogranuloma, an indolent tumor of histiocytes that usually presents as a solitary skin lesion in children. Cardiac juvenile xanthogranuloma has rarely been reported in children, and to our knowledge only once in an adult [1]. Older lesions can create extensively fibrotic masses and can cause significant complications, especially in visceral sites. Cutaneous lesions usually regress spontaneously and require no treatment. When involving visceral organs, or causing dysfunction due to mass effect, these lesions should be treated by surgery, radiation, and/or chemotherapy.

Reference

1. Lehrke HD, Johnson CK, Zapolanski A, Kasatki A, Grau JB, Maleszewski JJ. Intracardiac juvenile xanthogranuloma with presentation in adulthood. Cardiovasc Pathol 2014;23:54–6.

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