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IMAGES IN CV APPLICATION

An unusual cause of pulmonary outflow obstruction: IgG4 deposition disease-MRI observations

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Introduction

Intraluminal filling defects in the right ventricular outflow (RVOT) are often due to a thrombus or occasionally due to rare lesions like sarcoma arising from pulmonary arteries [1]. Deposition disease is a very rare cause for a thrombus like intra-cardiac luminal defect. IgG4 deposition diseases occasionally present with focal deposition of immunoglobulin deposits at various organs, including the heart [2]. MRI imaging features in the specific clinical contest may provide clue to this rare phenomenon. It is important to recognise this uncommon entity, in view of the widely differing management options.

Case report

36-year-old male presented with exertional dyspnoea and progressive loss of weight of 16 kg, over the last 1 year. He had no history of fever or any chronic illness. His physical examination was unremarkable. On echocardiography, he was found to have a filling defect in RVOT, at the level of pulmonary valve. His lab investigations revealed following parameters: haemoglobin 12.2 g/dl; TLC 11,000 cell/c mm, ESR 56 mm/1 h, platelets 3.92 lac, total protein 8.6 gm/dl, albumin 2.9 g/dl, globulin 5.7 g/dl, AST 13 IU, ALT 28 IU CRP 36.5, HIV negative, serum creatinine 1 mg/dl, BUN 10 mg/dl. Routine examination of urine was normal. Specific tests for immunoglobulin revealed; serum IgG levels—16.2 gm/l and serum IgG4—4.43 g/l.

Gadolinium enhanced MRI examination performed for evaluation suspected intraluminal defect, revealed a well defined T2 hypointense, mildly enhancing mobile mass lesion in the RVOT, measuring 3.2 × 2.5 × 3.4 cm. Lesion was attached to the adjacent pulmonary valve leaflets. Larger part of the lesion was below the level of leaflets with a small component projecting beyond the valvular orifice causing obstruction to the pulmonary outflow (Fig. 1). Imaging features suggested the possibility of a benign lesion like a papillary fibroelastoma or an adherent thrombus. Both ventricular chambers were normal and showed normal systolic function. There was no evidence myocardial fibrosis or inflammation. Patient underwent open cardiac surgery, right ventriculotomy with removal of the lesion. On surgical assessment lesion was whitish in colour, firm in consistency, attached to RVOT and adjacent annulus further extending to anterior wall of pulmonary artery. Post operative period was uneventful. Follow up examination 2 months after the removal of the mass, patient was asymptomatic, gained about 10 kg of weight with improvement of appetite.

On gross pathology examination lesion was firm in consistency, measured 4.3 × 3.5 × 2.8 cm. It was homogeneously grey-white on sectioning. On microscopic examination lesion was composed of predominantly spindle shaped cells with ovoid to elongated plump nuclei, vesicular chromatin, indistinct nucleoli, moderate cytoplasm with indistinct borders admixed with areas of mature plasma cell aggregation (Fig. 2a). There was no necrosis or abnormal mitosis (Fig. 2a, b). On immuno-histochemistry cells were positive for vimentin (Fig. 2c), patchy SMA and CD 68 negative for CK, desmin, myogenin, CD 34, S100. Plasma cells were positive for LCA, CD138, Kappa, Lambda and IgG4 (Fig. 2d) hence, the diagnosis of IgG4 deposition disease with intra-cardiac IgG4-deposition was made.

Discussion

Cardiac IgG-4 deposition disease is essentially a diagnosis which can only be made conclusively with clinical, laboratory and histological evidence. This condition can be considered in the spectrum of cardiovascular lesions in patients

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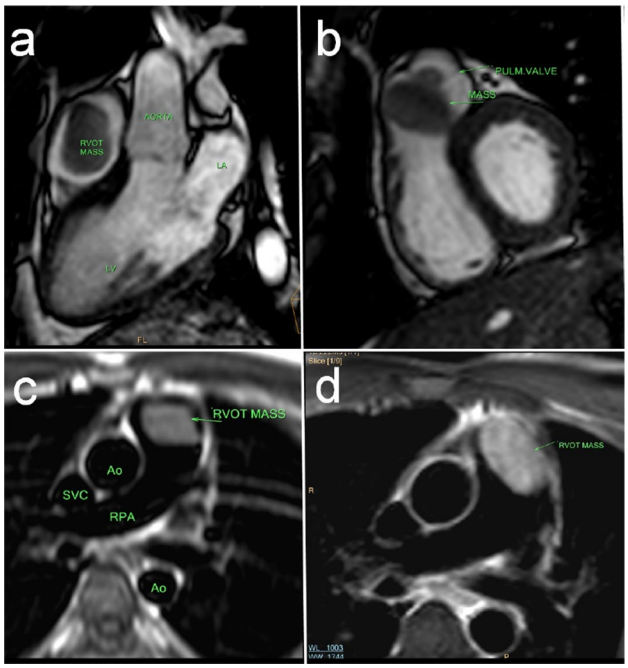

Fig. 1 Three chamber (**a**), RV outflow (**b**) bright-blood sequence demonstrate pedunculated filling defect in right ventricular outflow. **c, d** Axial Gd-enhanced T1 WI image at the pulmonary artery level showing uniform enhancement of the lesion

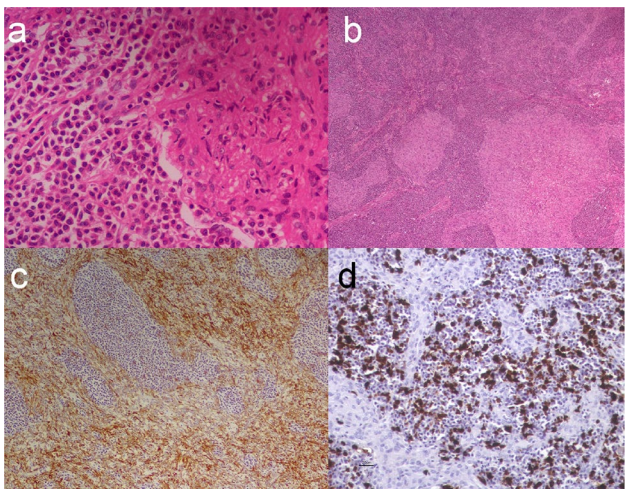

Fig. 2 **a, b** Microphotograph of H and E stain showing lesion matrix with aggregates of plasma cells (blue cells) and showing positive vimentin stain (**c**). Immunohistochemistry revealing +ve staining IgG4 complex (**d**)

with plasma cell dyscrasias [3]. Patients may also have ventricular hypertrophy or chamber enlargement. These patients are at risk for arrhythmias and diastolic dysfunction depending upon the site and extent of immunocomplex deposition. In suspected cases, immunofluorescent and electron microscopy (EM) studies are to be performed on the biopsy material. Coexisting amyloidosis may be seen in other organs.

On imaging findings are often non-specific. Screening echocardiography reveals filling defect and associated flow abnormality. MRI imaging is preferred as an additional examination as it provides characterising information on the filling defect, more comprehensive assessment of myocardial musculature and functional information of cardiac chambers. MRI findings are also useful in excluding malignant lesion like sarcoma. Imaging appearance mimics other benign entity like fibro-elastoma and cannot be distinguished. Hence final diagnosis can only be made with combination of clinical-laboratory parameters and histology. Specific immunological tests on the biopsy/resected specimen will provide the final proof. Heightened awareness of this entity is the key in considering the lesion in the differential diagnosis.

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Compliance with ethical standards

Conflict of interest There is no conflict of interest regarding the submitted work.

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