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IG A VASCULITIS PRESENTING AS HEMOPERICARDIUM

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A 36-year-old male presented with lower limbs vascular purpura, long-lasting fever of 10 days, joint pain, chest pain, and dyspnea. Transthoracic echocardiogram confirmed large circumferential pericardial effusion. Surgical pericardiectomy was performed, with 750 mL of hemorrhagic fluid removed. Cytology and bacterial cultures of the aspirate were unremarkable. Pericardial biopsy showed fibrinous pericarditis with a rich infiltrate of polynuclear neutrophils and skin biopsy concluded to leukocytoclastic vasculitis with dermal perivascular IgA and C3 deposits on direct immunofluorescence study. Thus, the patient received a 3-month course of colchicine and a 2-week course of aspirin. Follow-up transthoracic echocardiograms did not show recurrence of a pericardial effusion.

Introduction

Ig A vasculitis (IgA-V) is a systemic inflammatory disease affecting small vessels. While it is common and usually benign in childhood, it is less frequent in adults and has a more severe course. Its main manifestations are cutaneous purpura, arthralgia or arthritis, acute enteritis and glomerulonephritis. However, IgA-V rarely involves the pericardium and only few cases of pericarditis as primary presentation have been reported. We present a case of an adult patient with IgA-V with hemopericardium as inaugural presentation [1].

Case presentation

A 36-year-old male presented to emergency room with chest pain radiating to back, lower limbs vascular purpura, long-lasting fever of 10 days, diffuse joint pains with swollen wrists and knees. There was no history of abdominal pain or rectal bleeding. The family and personal past medical history were unrevealing. He did not report any preceding viral illness or sore throat. He reported central median chest pain that worsened on inspiration with shortness of breath. Physical examination showed temperature of 38°C, rapid heartbeats of 96 per minute, low blood pressure of 100/85 mmHg, paradoxical increase in jugular venous pressure during inspiration (Kussmaul's sign), and muffled heart sounds. The pulmonary auscultation was normal with elevated respiratory rate of 24 cycles / minute at rest. The abdominal palpation was painless and the digital rectal examination did not show bleeding in stool. Skin examination showed non-necrotic purpura on the lower limbs. The rest of physical examination was within normal limits. The patient was admitted for further work-up. The 12-lead electrocardiogram showed diffuse negative T-wave abnormalities, and cardiac enzymes (troponin and creatinine phosphokinase) were normal. Echocardiography showed normal motion of the left and right ventricles with a preserved left ventricular ejection fraction of 65% and a large circumferential pericardial effusion of 23 mm.

The chest computed tomography scan confirmed pericardial effusion **(figure 1).** Laboratory tests showed hemoglobin of 11.2 g/dL (normal range: 13 to 16 g/dl), platelet count of 260 G/L (normal range: 150-450 G/L), and white blood cells of 4.2 G/L (normal range: 3.5-5 G/L). C-reactive protein was elevated at 50 mg/dl. Liver, renal parameters and urine sediment were normal. Serum calcium, phosphates, and glucose were within normal range. Serum electrophoresis did not show monoclonal gammapathy. Immunoglobulin concentration showed an elective elevated level of Ig A at 5 g/dl (normal range 0.7-4 G/L). The fecal occult blood test was negative. The patient was referred to surgery for drainage. Pericardiocentesis was performed, and 750 mL of hemorrhagic fluid removed. A pericardial drain was placed for a few days until he could no longer bring back any serosity. Cytology, microscopic examination for acid-fast bacilli and bacterial culture of the aspirate, as well as flow cytometry were unremarkable. Pericardial biopsy concluded to highly intense fibrinous pericarditis with a rich infiltrate of polynuclear neutrophils, mesothelial hyperplasia **(figure 2).** There were no features of vasculitis, neoplastic cells or amyloid deposits. Thus, bacteriological workup consisting of repeated blood cultures and virological assessment including HBV, HCV, HIV serologies, and SARS-COV-2 oropharyngeal swab were negative. Immunological investigations showed negative anti-nuclear antibodies, rheumatoid factor, and anti-neutrophil cytoplasmic antibodies. Skin biopsies showed leukocytoclastic vasculitis and direct immunofluorescence showed dermal perivascular IgA and C3 deposits. IgA-V complicated with hemopericardium was diagnosed. The patient received a 3-month course of twice daily 0.5 mg of colchicine associated with a 2-week course of aspirin at dosage of 3g / day. Thus, the patient was regularly seen at our outpatient clinics. Follow-up transthoracic echocardiograms at 1 month, 3, and 9 months did not show recurrence of a pericardial effusion. He was free of symptoms. The current follow up is 2 years.

Discussion

Pericarditis is the most common disease of the pericardium encountered in clinical practice. It is responsible for 0.1% of all hospital admissions and 5% of emergency room admissions for chest pain [2]. The presentation of a patient with a pericardial effusion can range from an incidental finding to a life-threatening emergency. The causes of pericardial effusions are numerous and divided into inflammatory and non-inflammatory etiologies. It can be associated with systemic diseases such as infections (viral, bacterial or fungal), neoplasms and endocrine/metabolic disorders, or autoimmune diseases such as systemic lupus erythematosus, rheumatic fever, familial Mediterranean fever, and systemic onset juvenile arthritis [3].

In the present case, we diagnosed adult IgA- V based on purpura noted on the patient's bilateral lower limbs, elevated serum IgA levels, and typical histological findings on skin biopsy without gastro-intestinal (GI) or renal involvement (which are commonly affected). The patient fulfilled the American College of Rheumatology (ACR) established criteria for the diagnosis of IgA-V [4]. Our case illustrates an unusual inaugural presentation of a hemopericardium revealing an Ig A-V. Cardiac involvement is not regarded as a feature of IgA-V and may be more common than recognized. Limited cases reported Ig A-V involving the myocardium [1].

However, IgA-V rarely involves the pericardium, and only one published case of large or recurrent pericardial effusion as a primary presentation of IgA vasculitis has been identified [5]. Although pericardial biopsy did not demonstrate vasculitis and showed fibrinous pericarditis with a rich infiltrate of polynuclear neutrophils, we drew this conclusion because the patient showed improvement after anti-inflammatory course, exhaustive negative causal investigations, and met enough ACR classification criteria for Ig A-V [4].

Accordingly, we considered other types of small vessel vasculitis such as anti-neutrophil cytoplasmic antibody (ANCA) vasculitis, hypocomplementemic or cryoglobulinemia mediated vasculitis, which are associated with skin, joints and cardiac involvement. But our patient had no features indicating these causes. In addition, ANCA antibodies, complement fractions, and cryoglobulinemia dosage were normal.

Although there is no consensus regarding treatment, the administration of steroids is recommended for patients with IgA-V complicated by renal involvement or severe abdominal symptoms [6]. Treatment of Ig A-V complicated by pericardiac involvement is unclear. In our patient, we chose to use the association of aspirin and colchicine according to the European society of cardiology guidelines of pericardial diseases [7].

Ig A-V may involve pericardium and present as pericardial collection. Physician should be cognizant of such possible involvement". Based upon one case report, we may not be able to conclude anything related to therapy, as failure to observe recollection of fluid in the pericardial space, could be secondary to surgical pericardiectomy.

Novel Teaching Points

Cardiac involvement is not regarded as a usual feature of IgA vasculitis, but some patients may present with pericarditis and/or pericardial effusion.

Figure legends

Figure 1: Chest CT scan (axial and coronal planes) showing pericardial effusion with enhancement of the pericardium

Figure 2: Pericardium biopsy revealing fibrinous pericarditis with a rich infiltrate of polynuclear neutrophils on Hematoxylin, and Eosin stain (HES).

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