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Clinical Case Report

A case of Castleman's disease and adult necrotizing aortitis: a co-incidence or a significance?

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ABSTRACT

We report a case of concomitant Castleman's disease and adult necrotizing aortitis, an association heretofore not reported. A brief discussion of the current state of our understanding of the pathogenesis of aortitis and possible link between these two entities is presented.

1. Background

Castleman's disease (CD) or angiofollicular lymph node hyperplasia is a rare lymphoproliferative disorder associated in a subset of cases with the human immunodeficiency virus (HIV) and human herpesvirus 8. CD comprises at least two distinct diseases (localized and multicentric) with very different prognosis. It is also associated with a number of malignancies, including Kaposi sarcoma, non-Hodgkin lymphoma, Hodgkin lymphoma, and POEMS syndrome.

There are no reliable estimates of its incidence given its rarity.

Aortitis is a term that describes inflammatory conditions of the aorta that are traditionally subdivided into infectious and non-infectious categories.

Our understanding of the pathology of aortitis has evolved over recent years with an increased understanding of the importance of autoimmune/connective tissue diseases in the pathogenesis. This includes the emerging role of IgG4-related systemic disease. A new classification of aortitis has been proposed by Burke et al. in 2008 that relies on histological features only, rather than the less robust clinical classification that has previously been employed.

2. Case report

We present the case of a 45-year-old Caucasian male with history of hypertension, asthma, dyslipidemia, and obesity. He had no smoking history, diabetes, and no marfanoid features. In 2012, he was referred to our hospital for surgical management of a thoracic aorta aneurysm with an aortic root of 56 mm. The patient had a six-and-a-half year history of atypical chest discomfort episodes, lasting for up to 2–3 days at a time and spontaneously resolving. He also had a 4-year history of recurrent episodes of presyncope, never progressing to syncope.

In 2008, he had an incidental finding of a 13.5-cm retroperitoneal mass while being investigated for dilated cardiomyopathy in hospital (EF of 37%). A core biopsy showed atypical lymphoid proliferation characterized by regressed follicles and nonspecific interfollicular changes but the sample was too small to give an unequivocal diagnosis. An excision biopsy confirmed CD (hyaline vascular type) (Fig. 1). He had no known recurrences.

At the time of cardiothoracic surgical referral, he had an EF of 54% with a mildly dilated, mildly hypertrophied left ventricle, severe aortic regurgitation, and an aorta with the following measurements: STJ and trans-sinus, 56 mm; ascending aorta, 49 mm; and aortic arch, 44 mm on echocardiogram (Fig. 2). Coronary angiogram showed no abnormalities. The decision was made to go ahead with surgery for severe aortic insufficiency and progressing aortic aneurysm. He underwent an aortic root and arch replacement on early 2013 with a 29-mm mechanical aortic valve conduit.

Intraoperatively, the left carotid artery and brachiocephalic artery had a common origin and the left vertebral artery arose directly from the arch. The dilated aortic root and ascending aortic aneurysm were seen to extend to the innominate artery and left carotid artery. The trileaflet aortic valve and aortic aneurysm were excised.

Unfortunately, due to preservation in formalin for histopathology, culture was not able to be conducted.

Histopathological examination showed plate-like zones of medial necrosis bordered by histiocytes and giant cells (Fig. 3). There was

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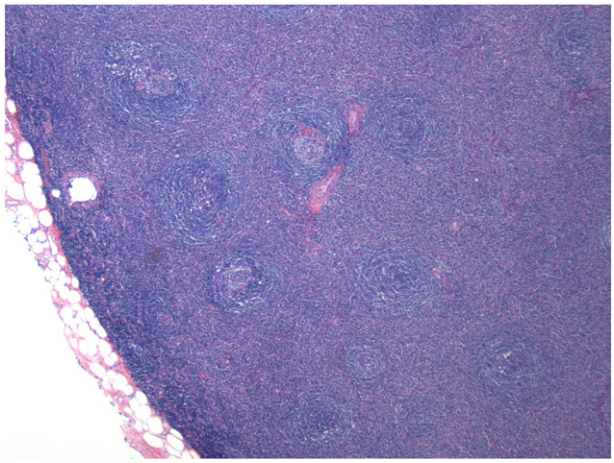

Fig. 1. Hyaline vascular CD (H&E, original magnification ×40).

extensive medial destruction with total loss of the elastic laminae in focal areas but the process was remarkably patchy. Intimal fibrosis (due to the aneurysm) was prominent. Atherosclerosis was mild to moderate.

Histology confirmed adult necrotizing aortitis (NA). HIV and syphilis serology were negative and thyroid function was normal. ANA, ENA, C-ANCA, P-ANCA, and rheumatoid factor were also negative. C3 was raised at 2.19 g/L (0.90–1.80) but C4 was within normal limits. ESR and CRP were elevated (102 and 63 mg/L, respectively) but unreliable given his recovery from major aortic surgery. On follow-up, CRP had reduced to normal levels but ESR was persistently elevated at 17H and 26H on subsequent visits.

The patient had no known family history of autoimmune conditions or aneurysmal disease and further history did not reveal any evidence of rheumatologic disease. Four or five years ago, he had been diagnosed with oral lichen planus presenting as mouth ulcers, which was managed with a course of prednisolone (5 mg/day) and hydrochloroquine for an unknown duration. There had not been further recurrence.

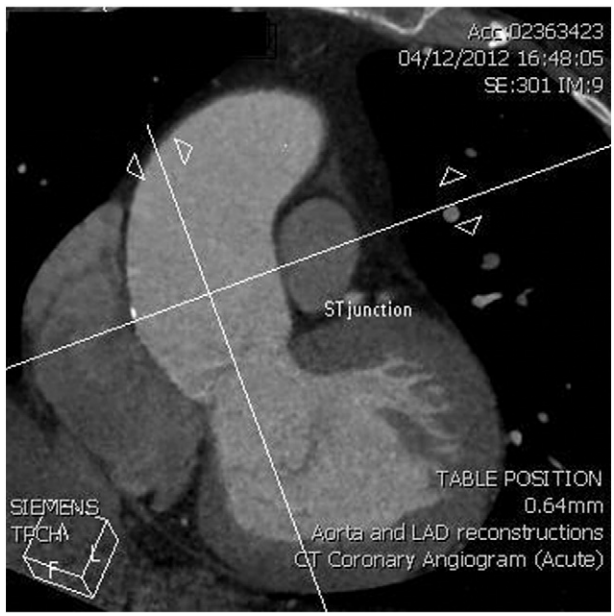

Fig. 2. Preoperative CT aortogram showing a dilated root and ascending aorta.

On follow-up, due to recurrent presyncopal episodes, bitemporal headaches, and a few episodes of amaurosis fugax in the left eye, he had a left temporal artery biopsy in June 2013 that showed nonspecific degenerative changes. An MRI brain showed multiple, bilateral lesions representing microhemorrhage and suspicious of embolic infarcts with no evidence of vasculitis. An echocardiogram demonstrated the valve to be well seated with normal hemodynamics.

He was found to be stable on the most recent clinic and will undergo long-term surveillance.

3. Discussion

In a significant percentage of cases, the diagnosis of aortitis is an incidental histopathological finding following surgery and the inflammation and aortitis are subclinical in nature [1–5]. Such cases of idiopathic isolated aortitis are typically localized to the ascending aorta and occur in association with an ascending aorta aneurysm [1,2].

In 2008, Burke et al. described 52 cases of aortitis and ascending aortic aneurysm and proposed a histological classification [6]. NA was the largest group and was characterized by zonal medial laminar necrosis, rimmed by giant cells (Fig. 4). It was usually isolated and showed a bimodal age distribution with a separation at age 65 years (adult vs. elderly). Elderly NA showed a female predominance. Adult NA had greater adventitial scarring and was histologically the same as Takayasu disease but limited primarily to the ascending aorta with no

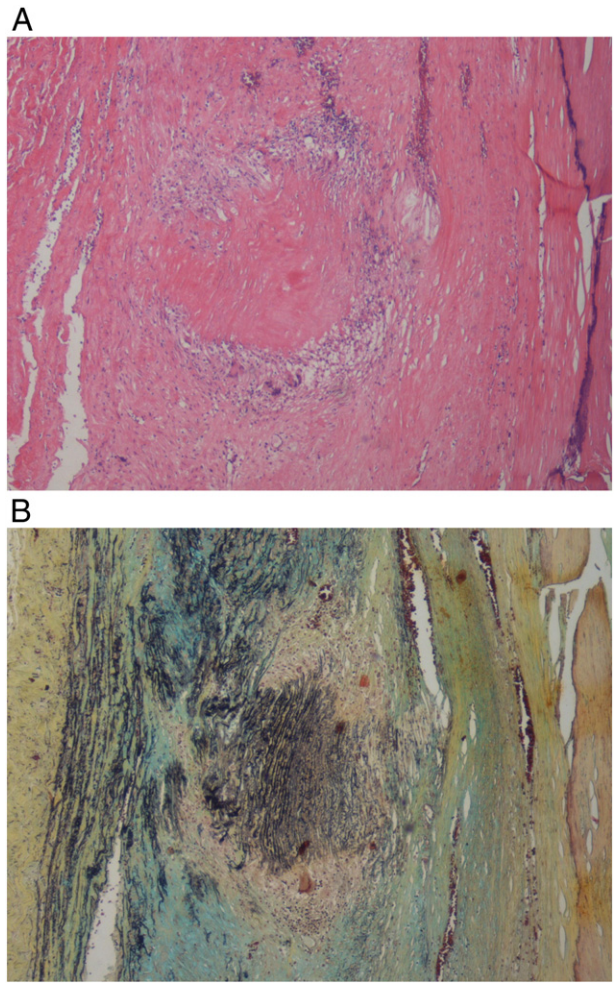

Fig. 3. (A) Plate-like zone of necrosis surrounded by histiocytes and giant cells. (H&E, original magnification ×100). (B) Same area: zone of necrosis highlighted nicely by Movat's pentachrome demonstrating retained elastic laminae that have collapsed after death of the smooth muscle component. The surrounding pale halo represents the inflammatory reaction. (Movat's pentachrome, original magnification ×100).