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Review

Aortic involvement in giant cell arteritis: Current data

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ABSTRACT

Aortitis due to giant cell arteritis (GCA) is rare but probably underestimated given the frequent paucity of symptoms. Thus, early studies relied on the occurrence of complications to estimate the prevalence of GCA aortitis. With this method, aortitis was a feature in 3 to 18% of GCA patients. Since then, the introduction of modern imaging techniques has established that aortitis is more common than previously thought. Aortitis should be considered in patients with atypical clinical presentations of GCA consisting, for instance, in isolated laboratory evidence of systemic inflammation or a relapse during treatment. Aortitis may be difficult to diagnose, as temporal artery biopsy has limited sensitivity in patients with predominant large-vessel involvement. Positron emission tomography (PET) and magnetic resonance imaging (MRI) are both highly effective for the early diagnosis of aortitis. Case-series evaluating PET in patients with GCA found evidence of aortitis in over half the cases, with predominant involvement of the thoracic aorta. To date, no evidence is available about the potential usefulness of PET or MRI in monitoring patients with GCA aortitis over time. Involvement of the aorta and other large arteries does not change the treatment strategy, which rests on corticosteroid therapy. Administration of a corticosteroid-sparing drug should be considered, most notably when a relapse occurs. Aortitis is associated with an increased risk of aneurysm of the thoracic aorta. Consequently, all GCA patients should be monitored for aneurysm at regular intervals, even after treatment discontinuation. The recommended strategy is an annual evaluation including a chest radiograph, echocardiogram, and abdominal Doppler sonogram; these imaging studies can be replaced by contrast-enhanced computed tomography of the chest and abdomen.

1. Introduction

Giant cell arteritis (GCA) is the most common form of large-vessel arteritis. The medium-sized and large vessels are affected. Onset occurs after 50 years of age, with a female-to-male preponderance. In Europe, the incidence of GCA is estimated at 32 to 290 million/year in individuals older than 50 years of age [1]. GCA typically involves the branches of the external carotid artery. The aorta and its branches are generally believed to be uncommon targets of the disease but their involvement is probably underestimated, as there are usually no symptoms until complications arise. Because aortitis can cause severe complications, routine screening for aortic involvement is mandatory. Modern imaging techniques show that aortitis is common even in the early stages of GCA.

2. Rate of aortic involvement in giant cell arteritis

GCA is the leading cause of inflammatory aortitis. Thus, in one study 48 (72%) of 66 patients with inflammatory aortic disease had GCA [2]. However, involvement of the aorta and other large vessels often goes undetected.

Early published data on aortic involvement in patients with GCA were based on the rate of aortic aneurysms diagnosed fortuitously or after acute events (aortic dissection and rupture of an aortic aneurysm). In retrospective studies, the prevalence of aortitis ranged from 3 to 18% [3–6]. However, data from surgical case-series [7–9] and autopsy studies suggested a high rate of aortic involvement in GCA. In retrospective studies of patients treated surgically for aneurysms of the thoracic aorta (Table 1), routine histological examination of the operative specimen indicated noninfectious aortitis in 4 to 12% of cases [7–9]. GCA was the second most common cause after idiopathic aortitis, with 8 to 30% of cases.

Subsequently, the introduction of new imaging techniques showed that aortic involvement was common, even before the development of structural abnormalities. For instance, of 145 patients with GCA, 33.1% had computed tomography (CT) evidence of aortitis [2]. In a prospective study involving routine CT at the

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Table 1
Rates of aortitis and giant cell arteritis in surgical case-series of patients with thoracic aortic aneurysms (TAAs).

	N of patients with surgery for TAA	*N* (%) of patients with noninfectious aortitis	*N* (%) of patients with idiopathic aortitis (%)	% of patients with GCA (%)	
Liang [9]	766	64 (8.4%)	81.3	7.8	Giant cells in 72% of patients with aortitis
Miller [8]	513	45 (8.7%)	47	31	
Rojo-Leyva [7]	383	45 (12%)	78	9	

TAA: thoracic aortic aneurysm; GCA: giant cell arteritis.

diagnosis of GCA, 45.4% of patients had thickening of the aortic wall compared to 13.6% of controls ($P = 0.02$). As discussed below, PET and MRI show aortic involvement in over 50% of GCA patients, with predominant involvement of the thoracic aorta.

3. Diagnostic challenges raised by aortitis in giant cell arteritis

The diagnosis of GCA relies on clinical, laboratory, and histological criteria as described in the 1990 American College of Rheumatology classification scheme [11]. The definitive diagnosis rests on examination of a temporal artery biopsy. GCA is readily diagnosed in patients with clinical symptoms related to arteritis of the branches of the external carotid artery. However, the temporal artery is not consistently involved, and the disease may be confined to the aorta and its main branches. When this is the case, the clinical manifestations are limited and nonspecific, and histological documentation may be difficult to obtain. However, the new imaging modalities discussed below may provide useful diagnostic information [12].

GCA aortitis may constitute the inaugural manifestation of the disease or develop later on, during the corticosteroid taper or after corticosteroid discontinuation [13]. The manifestations may be limited to systemic symptoms (fever, decline in general health, and laboratory evidence of inflammation). Pain in the back or lower back is an inconsistent symptom. Patients should be evaluated for abnormalities indicating involvement of the aortic branches, such as claudication of an extremity with an arterial bruit, a blood pressure difference between the two sides, decreased or absent pulses, and Raynaud's phenomenon. The diagnosis of aortitis may be missed initially, being established only during surgery or after acute complication of an aortic aneurysm.

It has been suggested that GCA may exist as two variants: the typical pattern responsible for temporal arteritis and an atypical pattern involving the large arteries (subclavian arteries, axillary arteries, and aorta) [14–16]. The atypical form may manifest only as a systemic inflammatory syndrome. In a study comparing these two variants, the patients with large-vessel GCA had a longer time to diagnosis; lower rates of headache, jaw claudication, and visual disturbances; and lower sensitivity of the temporal artery biopsy for the diagnosis of GCA [14].

4. Diagnostic imaging for aortitis

4.1. Arteriography

Arteriography was long the reference standard for the diagnosis of large-vessel arteritis, most notably Takayasu disease, seen as changes in the arterial lumen. In GCA, arteriography shows long, regular, smooth-walled stenosis; occlusions; and/or dilations. The subclavian, axillary, and brachial arteries may be involved.

Arteriography is an invasive investigation and does not perform well for the early diagnosis of vasculitis. The introduction of new imaging modalities has limited the usefulness of arteriography for the diagnosis of arteritis.

4.2. Computed tomography

Contrast-enhanced CT can establish the diagnosis of aortitis by showing concentric thickening of the aortic wall with postcontrast enhancement (Fig. 1) [10,17,18]. Regular circumferential thickening of the aortic wall to more than 3 mm indicates arteritis [18]. CT is less sensitive than MRI or PET for detecting early arterial inflammation.

4.3. Early diagnosis: positron emission tomography or magnetic resonance imaging

4.3.1. Positron emission tomography (PET)

PET is a nuclear imaging technique that reflects functional processes. Images are acquired after the injection of a radiolabeled synthetic glucose analog, FluoroDeoxyGlucose (FDG). FDG accumulates in tissues characterized by high glycolysis rates. FDG is taken up preferentially not only by malignant cells, but also by cells involved in the inflammatory response. Thus, PET has proven useful in patients with inflammatory and infectious diseases [19].

PET is effective for the early diagnosis of large-vessel vasculitis [19–22]. Vessel-wall inflammation can be detected in early-stage disease, before the development of the structural lesions whose presence is required for the diagnosis with other imaging modalities. PET images the entire body and can therefore detect lesions in many large and medium-sized arteries. The EULAR recommends PET, together with MRI, for diagnosing large-vessel vasculitis, most notably in patients with Takayasu disease, as histological documentation is difficult to obtain in the large-vessel forms of the disease [1].

Over the last decade, several case-series studies evaluated PET for the diagnosis of GCA (Table 2). In untreated patients, sensitivity ranged from 50 to 100% and specificity from 95 to 100% [23–34]. Sensitivity was higher in patients with elevations in markers for inflammation, particularly C-reactive protein [26]. One of the limitations of PET is the absence of standardized methods for interpreting the images [20]. Maximal standardized FDG uptake ratios have been computed for differentiating sites of interest in the aortic wall from the liver (taken as the reference); the selected cutoff had 88.9% sensitivity and 95.1% specificity for the diagnosis of GCA [24].

The arterial lesions are usually symmetric and predominate in the subclavian arteries (Fig. 2) [25]. Aortic lesions are found in more than half the cases, with selective involvement of the thoracic aorta

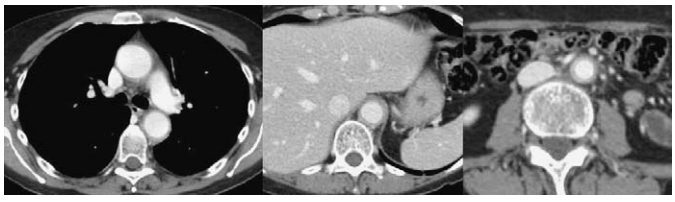

Fig. 1. Contrast-enhanced computed tomography of the chest and abdomen: regular thickening of the thoracic and abdominal aortic wall indicating aortitis in a 64-year-old woman with giant cell arteritis.