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Vasculitis and inflammatory arthritis

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A B S T R A C T

Vasculitis has been described in most types of inflammatory arthritis. The best described and most widely recognised form is rheumatoid vasculitis. The incidence of systemic rheumatoid vasculitis has declined significantly following the general early use of methotrexate in the 1990s, and it is now a rare form of vasculitis. Treatment of rheumatoid vasculitis is conventionally with glucocorticoids and cyclophosphamide, but there is an increasing role for rituximab similar to that in other types of vasculitis. Despite these developments the mortality of rheumatoid vasculitis remains high. Vasculitis in other types of inflammatory arthritis is less well described and the treatment remains empirical.

Introduction

Vasculitis is an inflammation of the blood vessel wall resulting in distal organ ischaemia or infarction. Vessels of all sizes may be involved in vasculitis. It has been recognised as a feature of inflammatory arthritis for over 100 years. Initially vasculitis was recognised with rheumatoid arthritis (RA), but as our concepts of the classification of inflammatory arthritis have evolved, it has been recognised that vasculitis may occur in association with other types of inflammatory arthritis, including spondyloarthropathies and psoriatic arthritis. The occurrence of vasculitis in spondyloarthropathies and psoriatic arthritis is much less well documented than in RA. The Chapel Hill Consensus Conference (CHCC) on the definition on nomenclature of vasculitis recognised the occurrence of vasculitis in association with RA as a distinct form of vasculitis [1].

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In this chapter, we will review the occurrence of vasculitis in association with the inflammatory arthropathies, focussing on the occurrence of vasculitis in RA.

Rheumatoid vasculitis

Systemic vasculitis occurring in the context of rheumatoid arthritis (SRV) was first described in 1898 by Bannatyne, in a patient with histological evidence of vascular inflammation of the vasa nervorum [2]. The early clinical descriptions in the 1940s and 1950s by Eric Bywaters and colleagues were of the classical features of peripheral gangrene and mononeuritis multiplex [3,4]. Since then, a wider spectrum of diseases have been recognised, including carditis, scleritis, nodules and systemic diseases [5]. All sizes of vessel may be involved, from the aorta to capillaries. Small vessel vasculitis can occur in isolation as small nail edge or nailfold lesions, which are considered to be benign but may herald or coexist with a major arterial disease [6]. Rheumatoid vasculitis usually occurs in patients with long-standing rheumatoid factor-positive erosive rheumatoid arthritis. Males with RA are at greater risk than females.

Classification and diagnostic criteria

The CHCC defined SRV as vasculitis occurring in association with and might be caused by RA [1]. There are no validated classification or diagnostic criteria. The Scott and Bacon criteria from 1984 remain in widespread use (Table 1) [7]. These have been criticised as not requiring a biopsy, and many recent studies have included only biopsy-proven cases.

Epidemiology

Rheumatoid vasculitis first became widely recognised and reported during the 1960s. In the UK, the first estimate of the incidence was from Bath/Bristol in the 1970s and suggested an incidence of 6/million [5]. In Spain, the annual incidence of biopsy-proven rheumatoid vasculitis during 1988–1997 was 6.4/million [8]. A detailed study conducted in Norfolk, UK, between 1988 and 2010, compared the incidence of SRV between 1988 and 2000 with the period 2001 to 2010 [9]. The annual incidence deceased from 9.1/million to 3.9/million. The decline was predominantly during the late 1990s (Fig. 1). Rheumatoid vasculitis is now less common than many other types of systemic vasculitis, such as the ANCA-associated vasculitides the overall incidence of which in Northern European populations is around 15–20/million [10,11]. The incidence in the Norfolk study was equal in both males and females, but other studies have suggested a male preponderance.

Several studies from the USA have supported this decline in SRV; a population-based study of the incidence of extra-articular RA reported a reduction in the 10-year cumulative incidence of SRV from 3.6% in 1985–1994 to 0.6% in 1995–2007 [12]. The reduction in the prevalence of SRV has also been reported in a serial cross-sectional study analysing both hospitalised and ambulatory patients from the US veteran population during 1985–2006, comprising a similar duration of cohort observation with our study of 22 years [13]. There has also been a decline in hospitalisation in California of SRV patients between 1980 and 2001 [14].

Table 1
Classification criteria for rheumatoid vasculitis.

The presence in a patient with rheumatoid arthritis of one or more of the following:
1 Mononeuritis multiplex or acute peripheral neuropathy
2 Peripheral gangrene
3 Biopsy evidence of acute necrotising arteritis plus systemic illness (e.g. fever or weight loss)
4 Deep cutaneous ulcers or active extra-articular disease (e.g. pleurisy, pericarditis, scleritis) if associated with typical digital infarcts or biopsy evidence of vasculitis

Other causes of such lesions, such as diabetes mellitus and atherosclerosis, should be excluded. Patients with nailfold lesions or digital infarcts alone are excluded.
Adapted from Scott and Bacon, 1984.