Biopsy-proven giant cell arteritis in a patient with ankylosing spondylitis: a rare coincidence of two diseases at different stages of life

Sirs,

Giant cell arteritis (GCA) is a large vessel vasculitis that typically occurs in individuals older than 50 years. GCA patients presenting with the typical cranial phenotype are generally older than those with a predominant extracranial pattern of the disease (1). Ankylosing spondylitis (AS) occurs in individuals generally younger than 40 years (2).

In this report, we describe a patient who was diagnosed with AS in the fourth decade of his life and with biopsy-proven GCA in his mid-eighties. The concurrence of these two distinct rheumatic diseases appears to be uncommon.

An 83-year-old Spanish man presented to the emergency department with sudden vision loss in his left eye. He had been diagnosed with ankylosing spondylitis (AS) 50 years earlier due to a history of long-standing inflammatory low back pain associated with grade IV bilateral sacroiliitis (Fig. 1) and HLA-B27 positivity. He had been treated with non-steroidal anti-inflammatory drugs (NSAIDs) for more than 30 years. However, in the last ten years, he had taken NSAIDs irregularly, depending on the pain. On admission, he reported a 48-hour history of hemicrania headache and jaw claudication. GCA was suspected, and due to the sudden visual loss, methylprednisolone pulses of 500 mg intravenously each day for 3 consecutive days were administered, resulting in improvement of the cranial symptoms but without visual recovery. A temporal artery biopsy showed typical findings of GCA, consisting of a mononuclear inflammatory infiltrate with the presence of multinucleated giant cells, disruption of the internal elastic lamina, and intimal hyperplasia. After the 3-day methylprednisolone pulse treatment, 60 mg/day of prednisone was administered for another 2 weeks. Later, prednisone was gradually tapered without any relapses of the disease.

Aortitis has been described in patients with spondyloarthritis. In this regard, data from a tertiary centre confirmed that positron emission computed tomography can disclose the presence of large vessel extracranial vasculitis in patients with AS (3). However, the co-occurrence of AS and biopsy-proven GCA with a typical cranial phenotype is rare and might pose diagnostic challenges due to overlapping symptoms such as headache, fatigue, and stiffness.

Although GCA typically affects older adults, whereas AS usually manifests in younger individuals (1, 2), the coexistence of both conditions in the same patient suggests a complex interplay of genetic, environmental, and immunological factors. In this regard, both AS and GCA are associated with the HLA region. However, AS is strongly associated with the HLA-B27 allele in the Spanish population (4). In contrast, as reported elsewhere, biopsy-proven GCA in Spain is associated with the HLA-DR4 genotype, specifically the HLA-DRB1*04:01 and HLA-DRB1*0404 alleles (5). A weaker association with HLA-B locus has been described in Spanish individuals with GCA. However, this association is mainly related to the HLA-B*15:01 allele (6). Therefore, no shared genetic predisposition may explain in this patient the development of two different inflammatory diseases separated in time.

This case highlights that having one inflammatory autoimmune disease, such as AS, does not necessarily reduce the risk of developing another, such as GCA. It reinforces the need for clinicians to maintain a high index of suspicion for overlapping or concurrent inflammatory autoimmune disorders, even when the presentation may seem atypical or unexpected based on the patient's age or prior medical history.

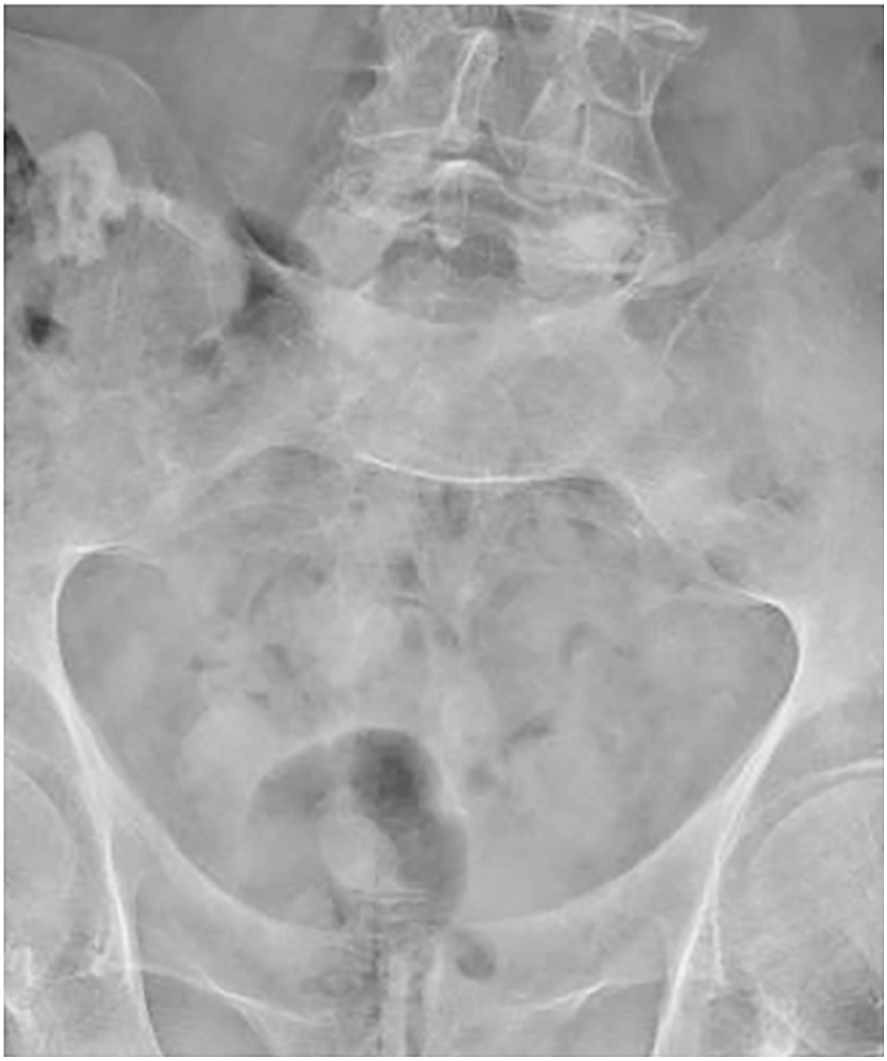

Fig. 1. Pelvis radiograph showing grade IV bilateral sacroiliitis with ankylosis of both sacroiliac joints.

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