Original Research Article

Coexistence of Giant cell arteritis with aortitis and Sweet's syndrome: is it a coincidence?

Abstract:

Sweet's syndrome (SS) is a rare disorder characterized by dermal infiltration by neutrophils. It was been reported in association with drugs, malignancies, infections, rheumatoid diseases, inflammatory bowel diseases. Its association with Giant cell arteritis hasn't been reported in our knowledge. The diagnosis of Giant cell arteritis was based on inflammatory biological syndrome with aortitis with negative infectious investigations. Herein, we present an unusual case of SS associated with GCA treated with steroids with good outcomes.

Key words:

Aortitis, Sweet's syndrome, Giant cell arteritis

Introduction:

Sweet syndrome (SS) is a rare disorder characterized by fever, neutrophilic leucocytosis, painful plaques on skin and dermal infiltration by neutrophils. Many associations have been reported in the literature, including malignancies, drugs, HIV infections, inflammatory bowel diseases, Behçet disease, rheumatoid arthritis, thyroid disease. Its association with Giant cell arteritis (GCA) hasn't been reported in our knowledge. This case represents unusual case of SS associated with GCA treated with steroids with good outcomes.

Case report:

A 67 year-old woman who had no medical history was hospitalized in April 2018 in our department of internal medicine, for exploration of aortitis.

The patient suffered since a month from general weakness, weight loss, pain of larges joints and insomniac paroxystic headaches. She had no visual

manifestations. In our department, her blood pressure was 100/60 mmHg, her pulse was at 80 battement / minute (bpm). Vascular examination including measurement palpitation and auscultation of pulses in all major vascular regions as well as the temporal artery found wide and symmetric pulses. Skin examination found bilateral infiltrated erythematous plaques on the palm of the hands. Skin biopsy confirmed the diagnosis of Sweet syndrome with evidence of dense dermal neutrophil infiltration: neutrophilic infiltrate with dermal edema, perivascular and interstitial infiltrate composed predominately of neutrophils and histiocytes **(Figure 1).**

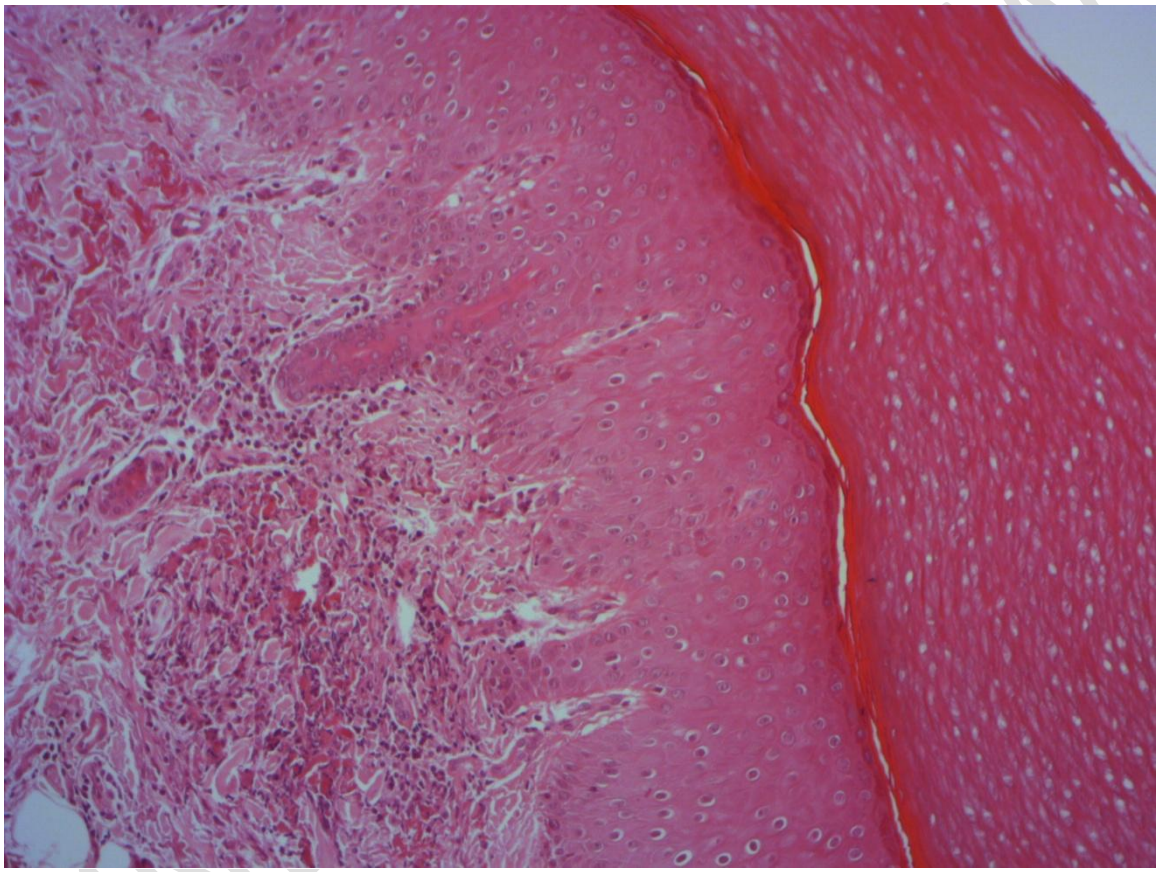

Figure 1: Diffuse neutrophilic and histiocytic dermal infiltrate in reticular dermis. Epidermis is normal (HE x 200)

Biological findings revealed increased Erythrocyte sedimentation rate (125mm/hour) and elevated CRP (146 mg/dL); Blood cell count showed white cell count at 11000/µL, haemoglobin was at 10.5 g/dL, platelets of 320000/µL. Liver function tests were normal. Abdominal Magnetic Resonance Imaging (MRI) revealed thickening of the aortic wall with late peripheral contrast uptake and peri aortic fat infiltration, with hyposignal T1 and hyper signal T2, from

T12 to L3. These findings were concordant with aortitis **(Figure 2)**. Infectious investigations including viral hepatitis, VHB, VHC, VIH, tuberculosis, syphilitis serology, wright and widal tests were negatives. Temporal artery biopsy was normal. The diagnosis of GCA with sweet syndrome was retained. Corticosteroids were started at high doses (1mg/Kg/day) for 1month then progressively digressed. The outcome was favorable and shehad no headaches. Inflammatory markers were within normal ranges (CRP=5). During her check-up, she was free of complaints (prednisone 10 mg/day) however she developed diabetes and osteoporosis. After total duration of 32 months of steroids intake, no flare up of her disease was noticed, decision to stop steroids was proposed. Radiological control was programmed before their stop.

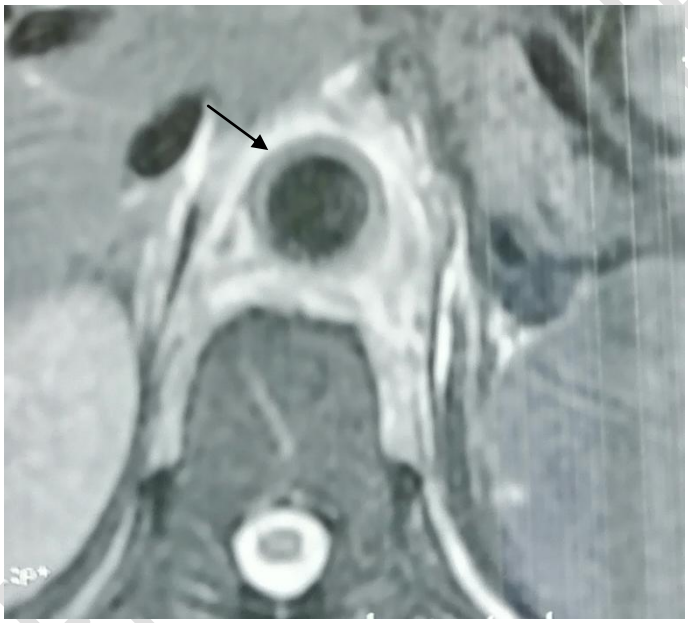

Figure2: Abdominal MRI, Axial section, aortitis

Discussion

GCA is a granulomatous vasculitis involving large and medium vessels, especially the extracranial branches of the carotid arteries. It's the most common vasculitis in adults affecting people more than 50 years [1].

The frequency of aortitis in GCA is under-estimated. Aortitis represents a serious complication because of the risk of anevrism, rupture and/ or dissection. It can be observed in initial presentation or occurs as a delayed complication. When it's present at the time of diagnosis of GCA, it seems to be associated with high risk of relapses and higher long-term vascular mortality rate [2]. Therefore, the screening of aortitis lesions at GCA diagnosis by an aortic CT-

scan was recommended [2]. Imaging studies showed signs of infra-clinical aortitis in 20 to 65% of cases at diagnosis [3]. Aortitis secondary to GCA is characterized in histological analysis by inflammation in media and adventitia with the presence of necrosis in the media associated and multifocal or transmural infiltrates of lymphocytes, histiocytes, neutrophils and polynuclear giant cells [4]. Only aortic biopsy and histologic findings could relate aortitis to GCA. In our case, clinical findings including headaches deteriorated general status, and inflammatory biological syndrome were suggestive of GCA. Temporal artery biopsy wasn't contributive. Good outcome with steroid therapy was an argument in favor of diagnosis. Our patient had also Sweet's syndrome (SS) which was diagnosed simultaneously with GCA. It's defined as an acute febrile neutrophilic dermatosis. SS is characterised by fever and erythematous painful nodules, plaques, and/or papules localized on the face, trunk and limbs [5]. It may be associated with systemic diseases such as dermatomyositis, Sjögren's syndrome, Behçet's disease, and takayasu arteritis. Its pathogenesis is still unknown. Autoimmune and infectious factors seem to be involved [6, 7]. This exceptionnal association of SS with GCA and involvement of the aorta has not been reported. The validity of the coexistence of these two diseases remained to be established. Few cases reported the coexistence of aortitis associated with SS had been published [8, 9]. In fact, vasculitis may be observed in SS [10] leading us to question if the aortitis is a part of SS or GCA or both. Arthralgia also could be multifactorial. They are observed inboth SS and GCA. Articular manifestation could be seen in 20 to 30% of adult patients with SS [11, 12]. Although the etiology of SS is not completely understood, an inflammatory component was involved. There is no consensus about treatment of aortitis in GCA or SS, but they are sensible for systemic steroids [2, 13]. In our case, we noticed persistent clinical and biological improvement after total duration of 32 months of steroids. Radiological control is scheduled before their stop.

Conclusion:

The association of inflammatory diseases such as Sweet's syndrome and giant cell arteritis is exceptional and even with involvement of the aorta. Biological inflammatory syndrome in eldery must lead us to suspect Horton arteritis and often normal temporal artery biopsy couldn't eliminate diagnosis. Both diseases had good outcome with steroids.

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