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 has been reported in association with drugs, malignancies, infections, rheumatoid diseases, inflammatory bowel diseases. Its association with giant cell arteritis (GCA) hasn't been reported to our knowledge. The diagnosis of GCA 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's syndrome (SS) is a rare disorder characterized by fever, neutrophilic leukeocytosis, 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's disease, rheumatoid arthritis, thyroid disease. Its association with giant cell arteritis (GCA) has not been reported to our knowledge. This case represents an 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 survey of aortitis.

The patient suffered from general weakness, weight loss tenderness over larges joints (shoulder girdle, knees) and insomniac paroxystic headaches for one

month. She had no ocular manifestations. In our department, her blood pressure was 100/60 mmHg in the two arms. Her pulse was at 80 beats per minute (bpm). Physical examinations including peripheral pulses in neck, arms and in temporal arteries showed wide and symmetric pulses. Skin examination found bilateral infiltrated erythematous plaques on the palms of the hands. Skin biopsy confirmed the diagnosis of Sweet's 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).**

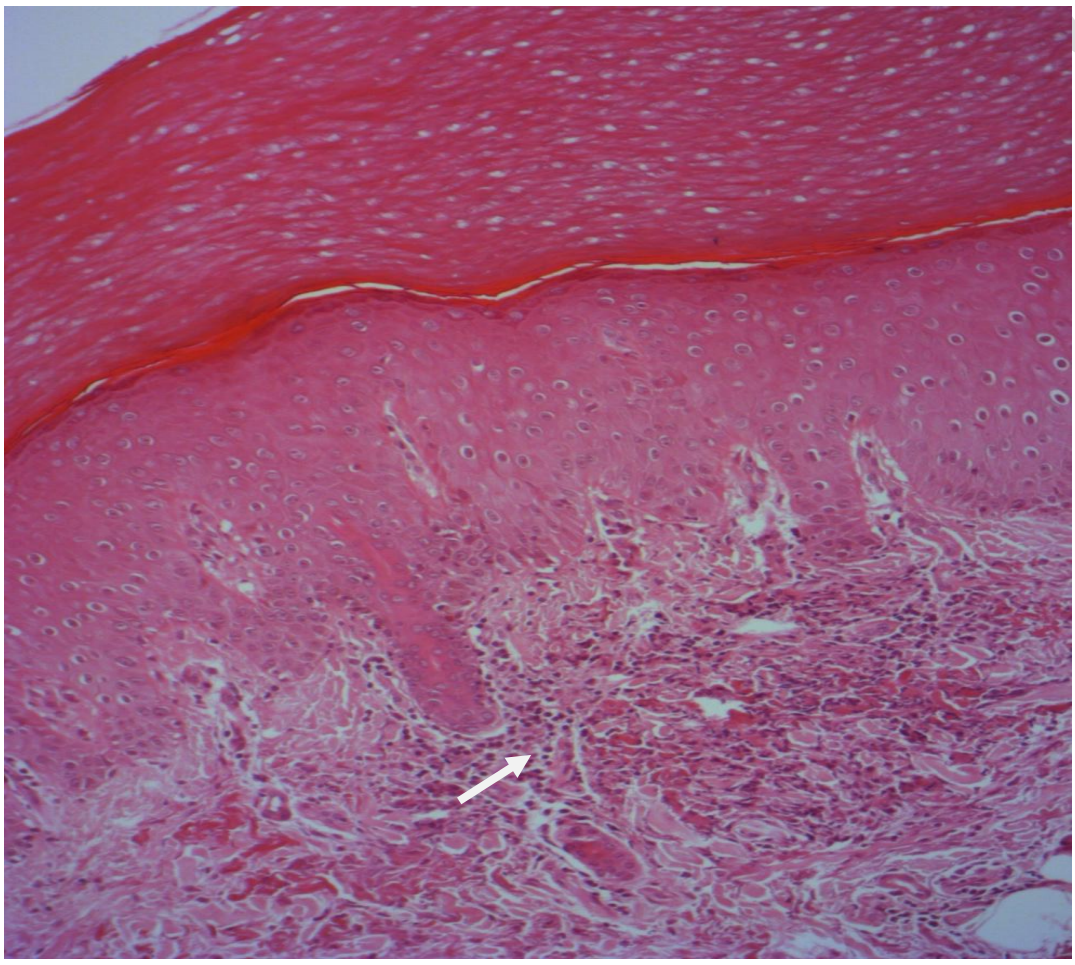

Figure 1: Diffuse neutrophilic and histiocytic dermal infiltrate in reticular dermis. Epideramis is normal. (HE x 200)

Biochemical findings revealed increased erythrocyte sedimentation rate (125mm /hour) and elevated C-reactive protein (CRP) (146 mg/dL); Blood cell count

showed white cell count at 11000/µL, haemoglobin 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-weighted sequence and hyper-signal T2-weighted sequence, from T12 to L3. These findings were concordant with aortitis **(Figure 2)**. Infectious investigations including viral hepatitis, hepatitis B virus (HBV), hepatitis C virus (HCV), human immunodeficiency virus (HIV), tuberculosis, syphiltis serology, and widal-wrights tests were negatives. Temporal artery biopsy was normal. The diagnosis of GCA with sweet's syndrome was retained. Corticosteroids were started at a high dosage (1mg/Kg/day) for 1 month then progressively digressed until 10 mg/day of prednisone. The outcome was favorable and she had no headaches. Skin lesions disappeared. Inflammatory markers were within normal ranges (CRP=5). During her check-up, she had no complaints. She developed diabetes and osteoporosis. After total duration of 32-months steroids intake, MRI will be done to control aortitis and steroids will be stopped.

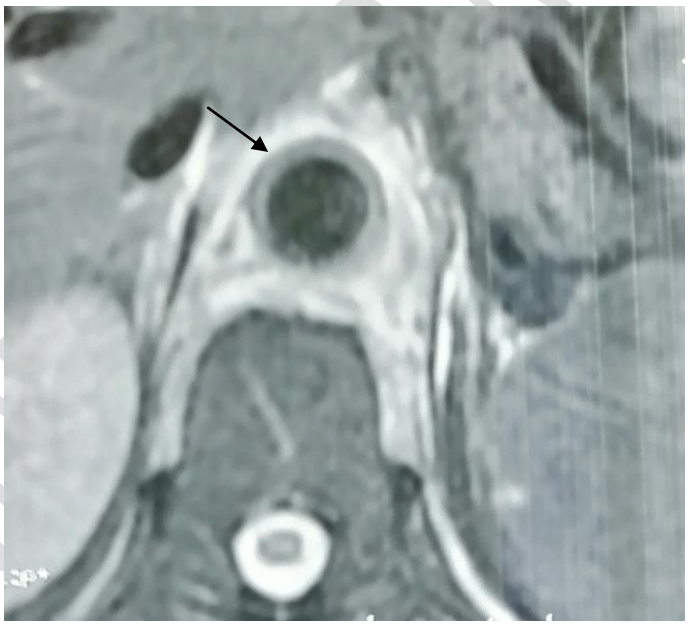

Figure 2: Axial section of abdominal MRI: thickening of the aortic wall with late peripheral contrast uptake and peri-aortic fat infiltration, with hyper-signal T2-weighted sequence, concordant with aortitis

Discussion

GCA is agranulomatous vasculitis involving large and medium vessels, especially the extra-cranial branches of the carotid arteries. It isthe 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 first presentation or occurs as a delayed complication. When it's presents 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 is 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 and multifocal or transmural infiltrates of lymphocytes, histiocytes, neutrophils and polynuclear giant cells [4]. 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 agreement in favor of diagnosis. Our patient also had sweet's syndrome (SS) which was diagnosed simultaneously with GCA. 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].Theassociation 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 [7, 8]. In fact, vasculitis may be observed in SS, [9] 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 [10-11]. 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 may be effectivefor systemic steroids [2, 12]. 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 unclear and even with involvement of the aorta. Neurological manifestations, abnormal blood pressure with specific skin findings in elders

must lead us to suspect giant cell arteritis, and normal temporal artery biopsy couldn't totally exclude the diagnosis. Both diseases had good outcome with steroids if diagnosed and treated early.

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