A case of Rheumatoid Arthritis presenting with Vasculitis

RAJESH KANNAN SAMINATHAN  
Department of General Medicine,  
MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract:  
Rheumatoid vasculitis is a rare but well recognized complication of rheumatoid arthritis. Rheumatoid vasculitis typically affects small and medium-size blood vessels. It is associated with high rates of premature mortality with up to 40 of patients dying by 5 years as well as significant morbidity due to both organ damage from vasculitis and consequences of the treatment. We report a case history of a female with rheumatoid arthritis who presented with gangrene of the foot and footdrop due to vasculitis

Keyword: Vasculitis, Rheumatoid Arthritis, Mononeuritis Multiplex.

Case report:  
A 65 year old female presented with history of ulcer in the gluteal region of 1 month duration, discoloration of both the feet and inability to walk for a week. She gave history of joint pain and swelling during the past 2 years for which she was treated at a local hospital symptomatically. She had no history of oral ulcers, skin rash, cough, difficulty in breathing, sicca symptoms, Raynaud's phenomenon, dysphagia or constipation. She did not give history of abortions in the past. She was not a diabetic, hypertensive or a coronary artery disease patient.

She had no family history of connective tissue disorders. She was moderately built, moderately nourished, anaemic. Her vital signs were within normal limits. Clinical examination showed absent dorsalis pedis artery pulsations on both sides. There was a healing ulcer of size 2 X 2 cm seen in the right gluteal region. Gangrenous changes were seen in both feet up to the middle of the foot.
Gangrene and Foot Drop
System examination showed bilateral foot drop with no other neurological deficits. Other system examination was unremarkable. A clinical diagnosis of Rheumatoid arthritis with Vasculitis / foot drop ? Mononeuritis Multiplex was made and proceeded with the investigations. Investigation revealed normal renal functions and blood glucose. Urine routine examination was unremarkable. Complete Hemogram showed reduced hemoglobin with elevated sedimentation rate and thrombocytosis and the peripheral smear showed a hypochromic microcytic picture. An echocardiography was done which was found to be normal. Chest x ray was essentially normal. LFT showed a normal protein with AG reversal, enzymes and bilirubin were within normal limits. Doppler study of lower limbs showed feeble low volume flow noted in both dorsalis pedis artery. Rheumatoid factor was positive (555 IU/ml (immuno turbidometric assay)). CRP was elevated. ANA was negative (9.6 U/l), ACL IgG: 14.9 u/ml, IgM: 31.6 u/ml, C- ANCA: 10.55 u/ml, P-ANCA: 11 u/ml which were within normal limits. Viral markers were negative (Hepatitis B, Hepatitis C, HIV). X ray hand showed features of Rheumatoid Arthritis with Soft tissue swelling, Peri articular erosion, Juxta articular osteopenia, Joint space narrowing, Sub chondral cyst, in MCP and PIP joints.

Nerve Conduction Studies was done. The Left median nerve showed decreased amplitude and velocity suggestive of axonal neuropathy. Both Peroneal nerve Proximal and Distal not stimulated suggestive of severe axonal neuropathy and Both Tibial nerves showed reduced amplitude and velocity suggestive of axonal neuropathy.
A final diagnosis of rheumatoid vasculitis with foot drop due to mononeuritis multiplex was made. Patient was started on steroids and intravenous cyclophosphamide pulse therapy.

Discussion:
Rheumatoid arthritis is a symmetric inflammatory arthritis that mainly affects the small joints of the hands and feet. The prevalence of rheumatoid arthritis (RA) in most populations of around 1%, with an incidence in women twice that in men.
The criteria for rheumatoid arthritis is based on ACR revised classification criteria 1988. The classification criteria has been modified in 2010 by ACR/ EULAR joint committee.
RA is a multisystemic disease. It can have articular and extra articular manifestations. The articular changes involve MCP, PIP, and MTP joints followed by the wrists, knees, elbows, ankles, hips, and shoulders roughly in that order. Very rarely cervical spine C1- C2 is involved leading to atlanto axial subluxation. The extra articular manifestations involve cardiovascular, respiratory, neurological, blood vessels – vasculitis, ophthalmic and skin etc. these occur in the presence of positive rheumatoid factor. Rheumatoid vasculitis is a rare complication of the long standing disease and is associated with increased morbidity and mortality.
The diagnostic criteria for systemic vasculitis has been proposed by Scott et al. The criteria for rheumatoid vasculitis is still poorly codified. It is common in patients of male sex and long standing disease. Systemic Rheumatoid Vasculitis Classification Criteria: Patients with Rheumatoid Arthritis having one or more of the features

- Mononeuritis Multiplex or acute peripheral neuropathy
- Peripheral gangrene
- Biopsy evidence of necrotizing arteritis + systemic illness

Deep cutaneous ulcers or active extra articular disease + nail fold lesion or positive biopsy The vasculitis may limited nail fold vasculitis (NFV) or systemic rheumatoid vasculitis (SRV). Predictors of vasculitis in RA include clinical and genetic factors. These include smoking, presence of rheumatoid nodules in early disease course, HLA-DRB1 alleles, HLA-C*03 allele, KIR2D5 allele. 3–9 Presence of vasculitis related neuropathy and low complement (C3) predicts severity in rheumatoid vasculitis.3 It is sometimes noted that arthritis is not active when the features of systemic vasculitis occur in patients who are thought to have 'burned out' disease. Vasculitis may involve any blood vessel bed in the body, including cerebral, mesenteric, and coronary arteries. 11 More severe features of rheumatoid vasculitis are frank infarctions of the digits and mononeuritis multiplex. 10,12,13.

In our patient we could not confirm the associated antiphospholipid syndrome as the patient was not willing to do the repeat anticardiolipin antibody tests after 12 weeks.

Two well-established pathways leading to vasculitic nerve damage are immune complex deposition and cell-mediated immunity. 15,16 The presence of peripheral neuropathy, petichiae, number of extra articular manifestations and IgA RF and low C3 predicted the presence of rheumatoid vasculitis.

Rheumatoid vasculitis should be treated aggressively. High dose steroid, other immunosuppressant (cyclophosphamide) are used to treat it. 14 It is emphasized that RA needs to be diagnosed and treated early to avoid these complications.

**Summary:**
Rheumatoid vasculitis is a rare but well recognized complication of rheumatoid arthritis. This case is presented for its rarity.

**References:**
1 Kelly’s Textbook of Rheumatology 8th Edition


