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Rheumatoid Vasculitis

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ABSTRACT

Rheumatoid vasculitis (RV) is a protean, destructive inflammatory process that is centered on the blood vessel wall and is associated with substantial morbidity. Diverse and severe manifestation of this rare condition is a big diagnostic and management challenge indeed. This report describes a 47-year-old male who presented with digital gangrene, myocardial infarction and peripheral neuropathy (diagnosed as a case of rheumatoid arthritis with rheumatoid vasculitis admitted into the department of rheumatology BSMMU, January 2013). Patient is now under regular follow up, is being treated with methotrexate and doing well. Studying this case may help to develop insight into rheumatoid vasculitis

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INTRODUCTION

Systemic vasculitis has been a diagnostic challenge in the areas of clinical medicine and rheumatology for many years. Among these is rheumatoid vasculitis, a rare and serious complication of rheumatoid arthritis (with an annual incidence of 3.4/million),[1] characterized by inflammation of mid-size arteries and capillaries, is associated with a particularly dire outcome, [2-3] with up to 40% of patients dying within 5 years due to organ damage from vasculitis and/or consequences of immuno-suppressive therapy.[4,5] RV can be clinically heterogeneous and can simultaneously affect multiple vascular beds. Clinical manifestations may include deep cutaneous ulcers, peripheral gangrene, vasculitic neuropathy, inflammatory eye disease and visceral infarction, all associated with poor outcomes. Long-standing RA, male sex, smoking, rheumatoid nodules have been associated with an increased risk of RV.[6-8]

The present case is different in that, the patient had shorter duration of disease (5 years) and even rarer presentation of coronary vasculitis (high lateral MI).[9]

The Case

Mr A Bari, a 47- year-old farmer from Royganj, Sirajganj, got himself admitted into the department of rheumatology Bangabandhu Sheikh Mujib Medical University (BSMMU), January 2013. He had symmetrical polyarthritis involving small hand and feet joints (for 5 years) with deformity. Presenting complaints were aggravation of joint pain, low grade fever and digital gangrene with severe pain involving 2nd to 5th digits (bilateral) for about 2 months. Few days prior to admission in BSMMU he experienced severe retrosternal chest pain and was diagnosed as a case of acute myocardial infarction (high lateral MI) in National Institute of Cardiovascular Disease (NICVD) and was treated accordingly in that institute, from where he was referred to BSMMU. It may be mentioned that he had no other risk factors for coronary arterial disease except for a, 10 pack year smoking history (and of course, he is a male). He was normotensive, non-diabetic with normal lipid profile and had no family history of premature coronary arterial disease.

On examination, peripheral pulses were okay, dry gangrene with demarcation line on the aforementioned digits were present. Neurological examination revealed mild distal sensory neuropathy with bilateral carpal tunnel syndrome. Examination of other systems revealed no abnormality. Clinical disease activity index (CDAI) score was 52 (very high disease activity, his weight was 53 kg.

Investigations revealed, total count of WBC 16000/mm,3 Neutrophil 70%, ESR 90 mm in 1st hour, liver function test and renal function test were normal. ECG recent MI (high lateral), Echocardiography normal, rheumatoid factor, anti-cylic citrullinated peptide strongly (Anti-CCP) positive, anti-neuclear antibody(ANA) negative, anti -double stranded (ds) DNA negative, Anti phospholipid antibody negative, perineuclear and cytoplasmic antinutriphil cytoplasmic antibody (P & CANCA) negative.

Referral and opinion from orthopedic department was sought, digital amputation along with head the proximal phalanges under local anaesthesia was done, on four different occasions (last on 19.02.13). Histopathology report was consistent with vasculitis with no immunoglobulin deposits on direct immunofluorescence (DIF).

Among the drugs prescribed, was clopidogrel, aspirin, ramipril, atorvastatin, antibiotics and after wound healing, oral cyclophosphamide (2mg/kg/day), with plenty of water intake, prednisolone (1mg/kg), cotrimoxazole (960 mg) 3 tabs /week, calcium + vitamin D, non-steroidal anti-inflammatory drugs (NSAIDS) and omeprazole. Rituximab (anti CD-20 monoclonal antibody) was not considered, as patient could not afford its cost.

Vaccination against Influenza virus and pneumococcal pneumonia was administered. Patient was under regular follow up, at vasculitis clinic BSMMU. He took cyclophophosphamide for 18 months, gangrene did not appear or recur, neither was there any infection.

Patient is now under regular follow up at rheumatology clinic Shaheed M Monsur Ali Medical College Sirajganj. He had exacerbation of his primary disease (rheumatoid arthritis) and is on methotrexate (MTX), NSAID along with other medication now. He quitted smoking just after diagnosis of the disease. Last visit was a couple of months ago, when was doing well (with CDAI 12) (Figure 1 and 2).

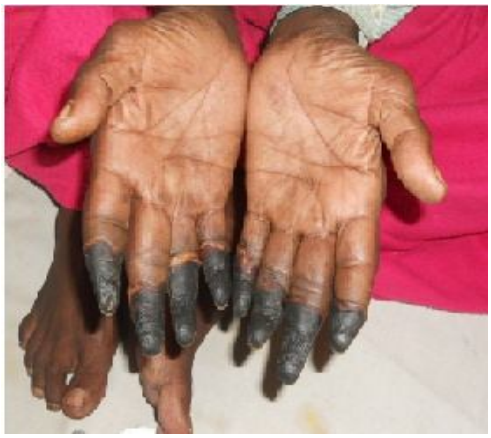

Figure 1: Palmar aspect with a closer look of the gangrenous digits

Figure 2: After digital amputation look of the gangrenous digits

DISCUSSION

Rheumatoid arthritis (RA) is a relatively common chronic inflammatory autoimmune disease that affects men and women of all ages, with worldwide presence. It affects 0.3–2.1% of the world population.[10] In addition to joint manifestations, several organs and specific systems can be affected in RA, enabling the onset of various extra-articular manifestations.[11] These manifestations are usually observed in individuals with high titres of rheumatoid factor and anti-CCP.[12] Rheumatoid vasculitis is one of the most important as well as grave manifestation of RA. The clinical presentation of RV continues to remain heterogeneous and rare, similar to that reported in older case series.[13-15]

Rheumatoid vasculitis typically affects small and medium-sized vessels, with associated sensory peripheral neuropathy (often motor), deep cutaneous ulcers, digital gangrene, nail bed infarcts and palpable purpura. CNS vasculitis, mesenteric vasculitis, scleritis/episcleritis, pulmonary angitis, necrotizing glomerulonephritis are rare presentation. The most common manifestation of RV is cutaneous vasculitis (as much as 90%), followed by neurologic (peripheral neuropathy, around 40%).[8-9] Mononeuritis multiplex may also occur. Mononeuritis multiplex has three clinical hallmarks: asymmetry, asynchrony, and a predilection for distal nerves. Motor mononeuritis multiplex may cause a devastating loss of function of the hands and feet, requiring assisted devices for feeding and leg braces (ankle-foot orthoses) for ambulation. Although cases of coronary vasculitis are well-documented in the medical literature, myocardial infarctions that are the direct result of coronary arteritis in RV are rare.[16-18] All except superficial cutaneous lesions are associated with poor outcome.

RV typically occurs in patients with longstanding, joint-destructive RA. In one study, the mean duration between the diagnosis of RA and the onset of vasculitic symptoms was 13.6 years.[9] Presentations of RV within five years of the RA diagnosis are very unusual. Patients with RV nearly always have rheumatoid nodules and are typically strongly positive for rheumatoid factor.[19] RV usually develops at a time when the inflammatory arthritis is “burned out”. Treatment of RV depends on the extent and the type of organ involved. Isolated nail fold vasculitis has a low risk for progression to systemic vasculitis and may be treated symptomatically without resorting to immunosuppressive therapy. Opportunistic infection should be excluded in RA

patients who appear to have developed RV before the initiation of more intensive immune suppression. Disseminated herpes zoster has been described as a mimic of RV in immune suppressed RA patients.

Systemic RV has a poor prognosis, should be treated with immunosuppressive therapy. Rituximab (375 mg/m ^{2} weekly times four doses) plus high-dose glucocorticoids (1-1.5 mg/kg body weight) are recommended first line treatment option for severe RV. This treatment is appropriate for patients with severe visceral involvement, including deep cutaneous ulcers, vasculitic neuropathy, scleritis, digital ischemia, and other significant manifestations of RV. Combination of daily oral cyclophosphamide (up to 2 mg/kg per day, assuming normal renal function) and high-dose glucocorticoids, can be suitable alternative to rituximab. Azathioprine can be used as maintenance therapy after remission induction with rituximab or cyclophosphamide. Prophylactic therapy against Pneumocystis pneumonia (trimethoprimsulph-amethoxazole) should be provided when patient is on cyclophosphamide and glucoco-rticoids in combination.

The strong encouragement of smoking cessation is important because of the well-established synergy of the risk factor with anti-CCP antibodies in contributing to severe RA.

Conflicts of interest: None.

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