Case Report:

Cutaneous Polyarteritis Nodosa without any subcutaneous nodules but with Pyoderma Gangrenosum like ulcers: A Rare Case Report

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Abstract

Cutaneous Polyarteritis Nodosa (cPAN) is a rare form of vasculitis of medium and small sized vessel of unknown etiology, first described by Kussmaul and Maier in 1866. The average age of onset is approximately 50 years. In both adults and children, males appear to be more commonly affected than females. cPAN is characterized by disease affecting primarily the skin without any major organ involvement. cPAN is demonstrated purpura, ulcerations, and tender recurrent subcutaneous nodules on the lower extremities as well as postinflammatory hyperpigmentation. Systemic symptoms may include fever, rash, joint pain and myopathy. Diagnosis is confirmed by histopathologic evidence of necrotizing inflammation of the medium and small-sized arteries. Treatment for cPAN includes the use of topical or intralesional corticosteroids for limited disease, systemic corticosteroids for extensive disease, and nonsteroidal anti-inflammatory drugs for symptomatic relief. cPAN is difficult to control and often requires longer courses of prednisone and steroid-sparing agents such as colchicine. Untreated polyarteritis nodosa has a poor prognosis, with a 5-year survival of 13%. Treatment improves this substantially, with current 5-year survival rates of approximately 80%. Subcutaneous nodules precede skin ulceration in a case of cPAN in 80% of times. But here we are presenting a case of cPAN who was admitted into Department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University with multiple sharply marginated hyper pigmented painful plaques over the left thigh with elevated vesicular margin and surrounding erythematous halo, without any subcutaneous nodules and with multiple pyoderma gangrenosum-like ulcers with necrotic slough with perilesional erythematous halo. Although only 20% of cPAN cases present without any subcutaneous nodules, and even fewer present with pyoderma gangrenosum like ulceration, this case report will reveal the rare possibility of presenting both and therefore will warrant the clinical suspicion of cPAN in case of lower limb ulcerations in a young lady.

Key words: Cutaneous Polyarteritis Nodosa(cPAN), subcutaneous nodules, Pyoderma Gangrenosum like ulcer in PAN.

Introduction

PAN is necrotising arteritis of medium or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules, and not associated with Anti-Neutrophil Cytoplasmic aAntibodies (ANCAs).[1] It can affect any organ but, for unknown reasons, it spares the pulmonary and glomerular arteries.[2] A less severe form called cutaneous polyarteritis nodosa (cPAN) has also been described. Its features include tender subcutaneous nodules, livedo reticularis, cutaneous ulcers, and necrosis.[3] It is often associated with streptococcal infection.[4] Hepatitis B infection is an important cause of secondary polyarteritis nodosa.[5,6] Peripheral nerves and skin are the most frequently affected tissues. The skin can demonstrate a range of lesions, including purpura, livedoid, subcutaneous nodules, and necrotic ulcers.[2] Neurologically, mononeuritis multiplex is the most common presentation.

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Involvement of the gastrointestinal tract, kidneys, heart, and central nervous system is associated with a higher mortality.[7] If there is renal involvement, patients may present with hypertension or acute kidney injury. Gastrointestinal symptoms occur in 14-65% of patients and postprandial abdominal pain from ischaemia is the most common symptom.[8]

Diagnosis of polyarteritis nodosa requires the integration of clinical, biopsy, and angiographic findings. According to the ACR criteria, polyarteritis nodosa can be diagnosed in a patient with vasculitis if three or more of the following features are present:6 weight loss greater than 4 kg, livedo reticularis, testicular pain or tenderness, myalgias, mononeuropathy or polyneuropathy, new-onset diastolic blood pressure greater than 90 mm Hg, renal dysfunction (blood urea greater than 14.3 mmol/L or creatinine greater than 133 µmol/L), evidence of hepatitis B infection, arteriogram showing the arteries that are dilated or constricted by the blood vessel inflammation and on biopsy, presence of granulocyte or mixed leukocyte infiltrate in the wall of a small or medium-sized artery. ANCAs are negative in PAN, and a positive ANCA in the context of necrotising vasculitis strongly suggests microscopic polyangiitis, granulomatosis with polyangiitis, or Churg-Strauss syndrome.[5] Inflammatory markers (including ESR and CRP) are elevated. Biopsy of small arteries will show evidence of necrotising inflammation.[6,9] Arteriography shows microaneurysms in the small-sized and medium-sized arteries.[10] FDG-PET/CT (Fluorodeoxyglucose positron emission tomography-computed tomography) is emerging as a potentially useful non-invasive imaging technique for diagnosis.[11] In cutaneous polyarteritis nodosa:3 Mild cases may require only non-steroidal anti-inflammatory drugs (NSAIDs) or colchicine. Prednisolone 30 mg daily or less is often effective in more severe cases but a dosage of 1 mg/kg/day may be required. Unfortunately, exacerbations occur with the tapering of the corticosteroids and adverse effects limit their long-term use. Immunosuppressive agents are frequently effective in cPAN resistant to high-dose corticosteroids.

Case report

A 23-year-old woman got admitted into Department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University with multiple sharply marginated hyper pigmented painful plaques over the left thigh. She gave a history of similar lesions on left arm three years back which ultimately ulcerated and healed subsequently leaving a scar after treatment with Prednisolone and Azathioprine for 6 months. She remained symptom-free for next three years without any medication. Review of systems was significant for myalgia, fatigue and unintentional weight loss. She denied any atypical limb pain, leg cramps, joint pain, photosensitivity, wrist or foot drop or any other skin lesions.

Physical examination on admission found a normotensive, afebrile woman in no acute distress, and there were multiple sharply marginated hyper pigmented tender plaques over the left thigh with elevated vesicular margin and surrounding erythematous halo. On 7th day of admission, the plaques on the thigh turned into deep tender ulcers with undermined edge covered with necrotic slough with perilesional erythematous halo. Evaluation of conjunctivae and sclera were normal.

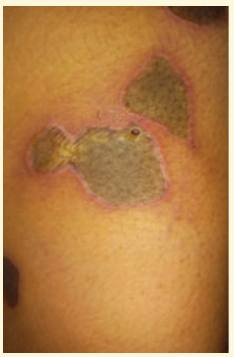

Figure A

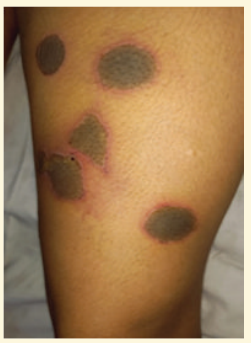

Figure B

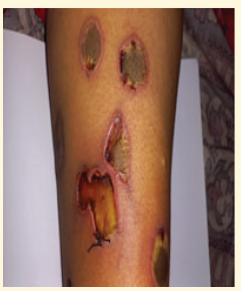

Figure C

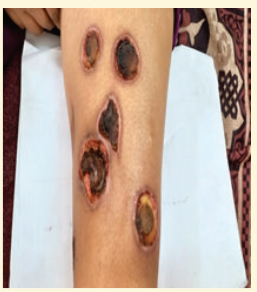

Figure D

Figure A and B: Erythematous Plaques during admission
Figure C: 7 days after admission
Figure D: 10 days after admission

Her complete blood count, C-reactive protein and comprehensive metabolic profile were all within normal limits. Hepatitis B and C panel were found negative. Wound swab culture obtained from ulcer revealed infection with Staphlycoccus aureus. A deep punch biopsy was obtained from the margin of an ulcer which revealed dense infiltration of lymphocytes in the reticular dermis and the subcutis with fibrinoid necrosis in the thick walled

medium-sized arteries. Based on histopathology and absence of systemic involvement, a diagnosis of cPAN was made. After addressing the infection with oral antibiotics, she was then started on Oral Prednisolone 1mg/KG/day and Methotrexate 15mg weekly. After 1 month of treatment with Prednisolone @1mg/kg/day, the ulcers show signs of healing as evidenced by appearance of granulation tissue with healthy ulcer margins.

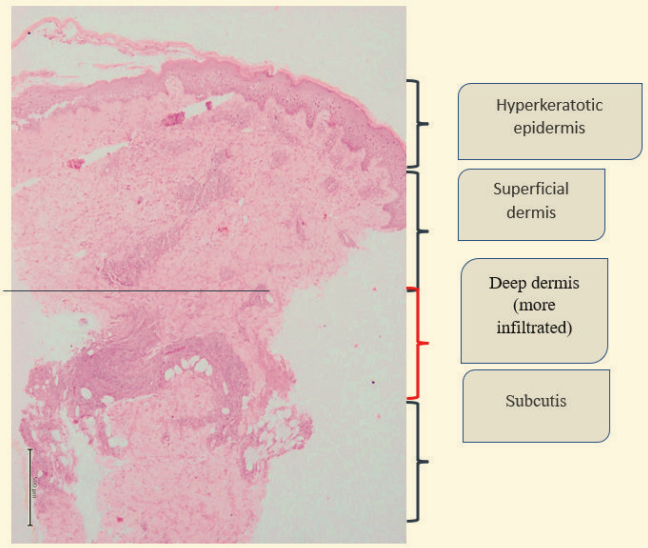

Figure E: Showing different layers of skin histopathology obtained from punch biopsy

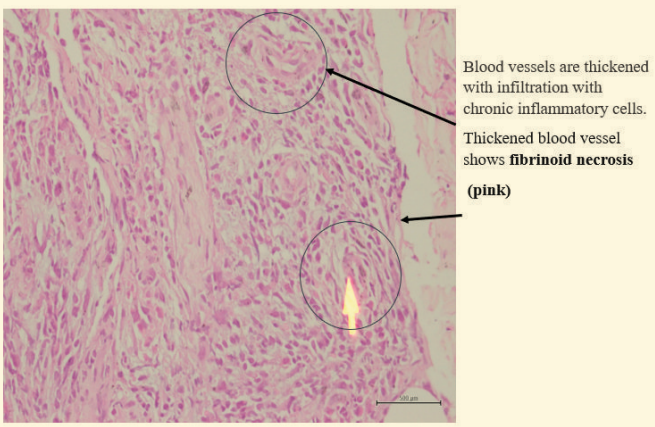

Figure F: Showing histopathological features of cPAN from the punch biopsy

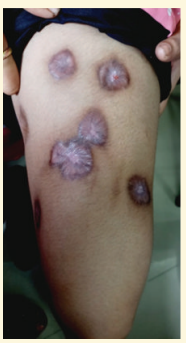

Figure G

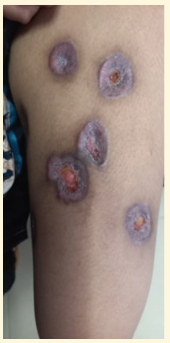

Figure H

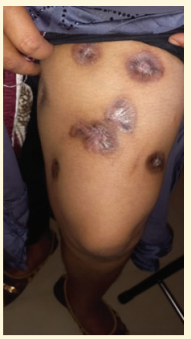

Figure I

Figure G: 1 month after starting Prednisolone
Figure H: 2 months after starting Prednisolone
Figure I: 4 months after starting Prednisolone

Discussion

Pineider J et al reported a case similar to our reported case except age and presence of painful black nodules of the patient. A 49-year-old white female smoker with a history of intravenous drug use and chronic hepatitis B complicated by early cirrhosis presented with painful ulcers on her right third finger and right second toe. She reported a 4-year history of recurrent spontaneous, painful black nodules, which ulcerated and then healed with scarring. She had been treated with multiple rounds of oral and intravenous antibiotics for presumed infections without resolution. Her liver transaminase levels were slightly elevated consistent with her known liver cirrhosis, but otherwise her comprehensive metabolic profile and complete blood count values were within normal limits. Magnetic resonance imaging of the right hand showed nonspecific arthritis of the third proximal interphalangeal joint without evidence of septic arthritis or osteomyelitis. Magnetic resonance imaging of the right foot showed mild skin irregularity over the second toe, interpreted as focal cellulitis. A punch biopsy was obtained from a purpuric papule on her right third finger. The day after biopsy, there was profound pathergy, creating a pyoderma gangrenosum-like lesion. Histopathology testing found a dense neutrophilic dermatitis with vasculitis, and near-complete obliteration of a medium-sized arteriole in the reticular dermis. Based on histopathology, lack of criteria to meet diagnosis for Behcet disease (BD) and absence of systemic involvement, a diagnosis of cPAN was made.[12] To the best of our knowledge, there is only one prior case reported in which a patient in whom pyoderma gangrenosum was initially diagnosed and was later

found to have systemic PAN after biopsy.[13]

Although usually cutaneous PAN presents with tender subcutaneous nodule on the background of livedo racemose and atrophic blanche-like lesions,14 our patient didn't show any of these signs. Rather she presented with Pyoderma Gangrenosum like deep, tender ulcers with erythematous halo. She also had negative P-ANCA, consistent with 80-90% of the patients of cPAN. We diagnosed the case ultimately based on the histopathological findings of dense lymphocytic infiltration with fibrinoid necrosis of the medium sized vessels in the reticular dermis and subcutis. There have been very few case reports where the initial clinical diagnosis of Pyoderma Gangrenosum (PG) was ruled out only after doing a histopathological analysis revealing features of PAN. cPAN patients who have elevated C-Reactive Protein and pretreatment ulcers have higher chances of relapse.[4] Delayed diagnosis and the inability to reduce severity of the flares resulted in prolonged skin, soft tissue, and joint damage leading to amputation of the affected digit. Therefore, early recognition is critical to control symptoms and prevent prolonged patient morbidity.

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