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Case Report

Primary biliary cirrhosis associated pustular vasculitis

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Abstract

The association between primary biliary cirrhosis (PBC) and cutaneous vasculitides is well recognized. Pustular skin lesions though, have been described in association with hepatobiliary diseases other than PBC. Once the more common infective pustular rashes have been excluded, the differential diagnoses for a pustular skin rash are acute generalized exanthematous pustulosis (AGEP), Sweet's Syndrome (SS), pyoderma gangrenosum (PG) and pustular vasculitis (PV). We present a case of pustular vasculitis associated with PBC.

Key words: Primary biliar cirrhosis, cholestasis, vasculitis, liver.

A 74 year-old-woman presented with a two week history of malaise, arthralgia, fever and rigors. She suffered from chronic renal failure, while primary biliary cirrhosis (PBC) was diagnosed two years earlier; she had positive M2 anti-mitochondrial antibodies (AMA) and antinuclear antibodies (ANA-Sp100, titter > 1:640). She was on allopurinol, lansoprazole, simvastatin and sodium bicarbonate.

On admission, Erythrocyte Sedimentation Rate (ESR) was 105 mm/hr and C-reactive protein (CRP) 263 mg/L (normal range ≤5), associated with marked leucocytosis (WCC: 25.2 x 109/L – neutrophils: 23.3 x 109/L, with toxic granulation). Chest x-ray, urine microscopy, repeat blood cultures and echocardiogram were unrevealing. Ten days later, she developed red papules and pustules over her shins. Similar lesions appeared over her face, soles, chest and back within 72 hours. No mucosal involvement noted. Rheumatoid Factor, ANCA, syphilis and HIV serology were negative. The pustules were negative for bacteria, fungi, typical and atypical mycobacteria, Herpes Simplex and Varicella Zoster Virus.

Punch biopsies revealed collections of neutrophils in the dermis with central cavitation, surrounded by palisade histiocytes. The skin lesions evolved from papules to pustules and then localized necrosis. She remained pyrexial with rising CRP.

Based on histology pustular vasculitis (PV) was diagnosed. The patient was started on steroids (prednisolone 40 mg) and within 7 days there was complete resolution of the rash. Inflammatory markers and leucocyte count returned rapidly to the normal range. Eventually steroids were reduced without recurrence of vasculitis.

Although rare, the association between cutaneous vasculitides and PBC is well described.[1,2]

Pustular skin lesions have been described in association with hepatobiliary diseases other than PBC.[3] This is the first published case describing PV in association with PBC.

The differential diagnoses of a pustular skin rash are acute generalized exanthematous pustulosis (AGEP), Sweet's Syndrome (SS), pyoderma gangrenosum (PG) and PV, once the more common infective pustular rashes have been excluded, Skin biopsy can help in differentiating the possible causes, but since there can be significant histological overlaps, the ultimate diagnosis can only be made after the individual clinical history is taken in context.

AGEP shows very superficial pustule formation within the epidermis and is often associated with drugs (e.g. beta-lactams, macrolides and calcium channel blockers),[4] SS typically shows papillary dermal oedema with a widespread neutrophil infiltrate and is associated with female sex and haematological malignancy.[5] In PG there is often evidence of a deep and dense neutrophil infiltrate, associated abscesses, blood vessels with fibrin deposits and ulceration. Clinically, PG is associated with conditions such as rheumatoid arthritis, Crohn's disease and monoclonal gammopathy.[6] In PV

there is predominantly a neutrophilic vasculitis within the dermis, with the exclusion of fibrinoid necrosis of vessel walls and is often idiopathic. This is in contrast to 'pustular vasculitis of the dorsal hands', a recently described condition, where lesions histologically identical to Sweet's syndrome occur exclusively on the dorsa of the hands.[7]

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All patients with a pustular vasculitis should have haematological malignancy excluded with a blood film, and if clinically indicated, bone marrow examination. Like all of the neutrophilic dermatoses, lesions generally respond well to topical and/or systemic steroids. Other treatments include oral dapsone, ciclosporin and tacrolimus.

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