CASE REPORT

Pustular vasculitis

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SUMMARY

We present a rare case of pustular vasculitis secondary to streptococcal infection. The majority of cases described in the literature predominantly involve the dorsal hands, and may subsequently spread to involve the lower limbs. However, our case demonstrates a rare case where primary involvement of the lower limbs was present with secondary involvement at the dorsum of the hands. The patient responded well to intravenous methylprednisolone reflected both clinically and by his acute phase response.

BACKGROUND

This is a rare case of pustular vasculitis in a young patient and who presented to the rheumatology team with a wide differential diagnosis, and should always be considered.

CASE PRESENTATION

A 41-year-old man presented with an acute extensive painful pruritic papular rash which started on the medial aspect of his left thigh, and over 6 days spread to cover his legs, arms, back and lower abdomen. Two weeks prior to his admission, he had been bitten by insects over his right calf, which appeared to have become infected and so was started on flucloxacillin for 3 days. He had diffuse polyarthralgia with swelling of his right hand, but had no mouth or genital ulcers and no other systemic symptoms, no recent foreign travel, and had not been started on any other new medications. He had a background of asthma and depression. His daughter had psoriasis but had no other relevant family history. He smoked 20 cigarettes daily and drank alcohol moderately.

On examination, he had two 2 cm purple ulcerated lesions over his right calf and a widespread palpable purpuric rash over his legs, arms and trunk (figure 1). His joints were diffusely tender with no active synovitis. Systemic examination was unremarkable.

He was initially diagnosed as having a drug reaction secondary to flucloxacillin and was started on topical emollients, and a reducing dose of oral prednisolone (30 mg daily). A skin punch biopsy was performed from his back and he was discharged.

Two weeks later, he was readmitted with worsening of the rash, which had become necrotic with no improvement on steroids. He had also developed splinter haemorrhages in his left little and ring fingers.

Figure 1 Widespread pustular rash over lower limbs.

INVESTIGATIONS

Initial investigations revealed a markedly elevated white cell count of $21.80 \times 10^9/l$ (normal range 4–10.5), neutrophilia of $17.97 \times 10^9/l$ (normal range 1.8–7.5) and C reactive protein of 175 mg/l (normal range <5). Ferritin was also raised at 455 µg/l (normal range 30–400). Renal, liver and bone profile were normal.

Immunoglobulin (IgA) was raised at 10.4 g/l (normal range 0.7–4); however, no paraprotein was detected by electrophoresis or immunofixation. IgG and IgM were normal as were his Creatine-kinase, lactate dehydrogenase, ACE, and rheumatoid factor. Antinuclear antibody, ELISA and immunofluorescence were negative, with proteinase-3 of 0.5 U/ml (<5), and myeloperoxidase (MPO) of 0.3 U/ml (<5).

Antistreptolysin O titre was markedly elevated at 6000 (normal range <201) and streptococcal anti-DNase B titre was 400 U/ml (normal range <200 U/ml) suggesting streptococcal infection.

He had grown coagulase negative staphylococcus from a blood culture on initial admission although subsequent blood cultures were all negative. Genetic tests were negative for human leukocyte

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Figure 2 Skin punch biopsy at high power (x200) showing a dense dermal infiltrate of neutrophil polymorphs. Also a vessel is evident showing perivascular fibrin deposition and damage to its wall signifying vasculitis.

antigen -B27 and B51. Skin swabs from the ulcerated lesions showed a heavy growth of staphylococcus aureus and β -haemolytic streptococcus group B. Chest radiograph was normal and transthoracic echocardiogram showed no evidence of valvular vegetations. Urinalysis was negative for blood or proteinuria, and urine cultures were negative.

Skin biopsy histology demonstrated dense neutrophil polymorphs throughout the dermis mainly confined to the papillary dermis in an interstitial and perivascular pattern, surrounded by fibrin deposition and extravasation of erythrocytes (figure 2). Direct immunofluorescence showed non-specific deposition within some vessels of IgG and IgA suggestive of a neutrophil dermatosis or dermal vasculitis. Whole body CT did not reveal any significant lymphadenopathy, malignancy or focal lesions.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes pustular vasculitis, acute febrile neutrophilic dermatosis (Sweet's syndrome) and pyoderma gangrenosum.¹ The lesions share a histopathological appearance demonstrating neutrophilic dermatosis and leukocytoclastic vasculitis.^{2 3}

TREATMENT

Overall, the clinical, histological and biochemical findings were consistent with pustular cutaneous vasculitis secondary to streptococcus, and he was started on broad-spectrum intravenous antibiotics and oral dapsone. However, his acute phase response remained persistently elevated with no improvement in his dermal symptoms.

He was then given three 500 mg pulses of intravenous methylprednisolone with concomitant oral prednisolone (40 mg daily).

OUTCOME AND FOLLOW-UP

This resulted in a significant improvement both clinically with his skin lesions completely resolving (figure 3), and in his acute phase response with his CRP falling to 35 mg/l, and neutrophils to $9.7 \times 10^9/l$. He continues to remain in clinical and biochemical remission.

DISCUSSION

We present a rare case of pustular cutaneous vasculitis, which describes a heterogeneous group of disorders characterised by

Figure 3 Partial resolution of pustular rash over lower limbs.

papules, haemorrhagic plaque pustules on purpuric bases² and bullous lesions.³

The majority of cases described in the literature predominantly involve the dorsal hands^{1 3} and may subsequently spread to involve the lower limbs; however, our case demonstrates a rare case where primary involvement of the lower limbs was present with secondary involvement at the dorsum of the hands. The mean age of onset is 68.75 years.³ Fever, pathergy⁴ and peripheral neutrophilia can be present, but are not necessary to make the diagnosis.³ The lesions are often intensely painful, often disproportional to the lesion size⁴ and ulceration is a common feature.^{4 5} Systemic involvement, myalgia and arthritis may also be present.^{1 3}

The differential diagnosis includes pustular vasculitis, acute febrile neutrophilic dermatosis (Sweet's syndrome) and pyoderma gangrenosum.⁶ The lesions share a histopathological appearance demonstrating neutrophilic dermatosis and leukocytoclastic vasculitis.^{2 3} Vascular endothelial damage can range from endothelial swelling to fibrinoid necrosis.³ Subepithelial oedema, spongiosis, ulceration and neutrophilic microabscesses may also be present.⁵

The underlying pathogenesis is thought to be immune-complex mediated resulting in lysosomal product release and a pronounced neutrophilic inflammatory infiltrate.¹ The term 'neutrophilic dermatosis'³ is often preferred (particularly where there is no biopsy evidence of vasculitis). However, others argue that pustular vasculitis is a variant with important features to distinguish it from other forms of neutrophilic dermatosis.³

A variety of underlying systemic diseases may also be present, including Behçet's syndrome,¹ blind loop syndrome, inflammatory bowel disease,⁶ myeloproliferative disorders,⁴ visceral malignancies⁴ and chronic gonococcaemia.¹ Additional association conditions include thermal injury,³ exposure to chemical fertiliser,³ pregnancy,⁵ drug reactions⁵ and streptococcal infection^{3 7} as demonstrated in this case. Additionally, primary idiopathic pustular vasculitis has also been described.^{1 8}

Management is primarily with corticosteroids, either topical or systemic.³ This can be augmented with potassium iodide.³ Dapsone has been shown to be effective in some cases; however, recurrence of the rash is common.⁵ Minocycline, while useful in pyoderma gangrenosum, has been proposed as a cause of neutrophilic dermatosis, and recurrence often occurs even if lesions do show some improvement initially. Ciclosporin and tacrolimus can be used in resistant cases.³ Resolution of lesions takes place over a period of weeks.⁴ Recurrence of the lesions is variable with conflicting reports.^{1 3 4} Low-dose dapsone has been used to reduce recurrence risk.⁹

Learning points

- ▶ Pustular vasculitis is a rare cutaneous vasculitis, which predominantly involves the dorsal hands and may subsequently spread to involve the lower limbs. However, our case demonstrates a rare case where primary involvement of the lower limbs preceded hand involvement.
- ▶ Associated conditions include streptococcal infection, pregnancy, drug reactions and thermal injury.
- ▶ The mainstay of treatment is corticosteroids, topical or systemic.

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