CASE REVIEW

Neuropathy and a rash

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A 26 year old woman was referred to dermatology with a one year history of a rash on her lower legs, abdomen, and arms, and a six month history of paraesthesia in her right hand and left foot. She had no history of fatigue, weight loss, fever, or arthralgia.

There was palpable retiform erythema and broken livedo on her abdomen, arms, and legs (fig 1). She had reduced sensation to light touch in the right ring and little fingers, the lateral palmar and dorsal hand, the left lateral calf, and the dorsum of the left foot. She had a feeling of clumsiness in her right hand that caused her difficulty with writing, but there was no weakness.

Blood tests were normal, including a full autoimmune screen. Skin biopsy showed medium vessel vasculitis with fibrinoid necrosis. Electromyography showed sensory damage in the right ulnar nerve and asymmetrical superficial sensory damage in the peroneal nerve, with the left side being affected to a greater extent.

Questions

1. What is the most likely diagnosis?
2. How can other causes of this neurological condition be ruled out?
3. How should this condition be managed?

Answers

1. What is the most likely diagnosis?
   
   Short answer
   
   Mononeuritis multiplex due to cutaneous polyarteritis nodosa.

   Discussion
   
   Mononeuritis multiplex is a type of peripheral neuropathy affecting two or more nerves. It can be asymmetrical, asynchronous, and can affect sensory and motor function. Mononeuritis multiplex is commonly caused by vasculitis. Results from the electromyogram show simultaneous asymmetrical involvement of three non-contiguous nerve trunks. This is consistent with mononeuritis multiplex, and differentiates it from mononeuropathy, where a single nerve is affected, or polyneuropathy where many nerves are affected in a more symmetrical manner.

   This patient had a six month lag between the onset of her rash and the onset of her neurological symptoms; however, associated neurology can also manifest simultaneously with the rash. The nature of the rash helps to identify the cause of mononeuritis multiplex. Broken livedo reticularis is suggestive of cutaneous polyarteritis nodosa, a form of vasculitis associated with mononeuritis multiplex.

   The differentials for a livedo eruption include systemic lupus erythematosus, antiphospholipid syndrome, and thrombocytopenia. Cutaneous polyarteritis nodosa affects small to medium sized arteries but does not involve internal organs. Medium vessel vasculitis with fibrinoid necrosis is seen on histology (fig 2).
Fig 2 Histology of deep skin biopsy from left lower leg showing medium vessel vasculitis with fibrinoid necrosis

2. How can other causes of this neurological condition be ruled out?

Short answer

Full blood count, glycated haemoglobin, hepatitis screen, anti-nuclear antibody, extractable nuclear antigens, anti-neutrophil cytoplasmic antibodies, rheumatoid factor, and complement levels can help to rule out other vasculitides, diabetes, connective tissue disorders, infections, and malignancy as possible causes of the mononeuritis multiplex.

Discussion

Mononeuritis multiplex can be associated with other forms of vasculitis such as granulomatosis with polyangiitis and Immunoglobulin A vasculitis. It also occurs in diabetes mellitus, connective tissue disorders (rheumatoid arthritis, systemic erythematous, scleroderma), infections (hepatitis B and C), malignancy, amyloidosis, and cryoglobulinaemia.

Detailed history, examination and investigations are important to identify the underlying cause. Relevant laboratory studies include full blood count, glycated haemoglobin, hepatitis screen, anti-nuclear antibody, extractable nuclear antigens, anti-neutrophil cytoplasmic antibodies, rheumatoid factor, and complement levels.

Patients presenting with a rash should have a skin biopsy to help confirm the diagnosis. Nerve biopsy might be required for diagnosis in some cases.

3. How should this condition be managed?

Short answer

Oral corticosteroids are required to treat the inflammatory response. Cyclophosphamide or azathioprine might be required to treat the vasculitis.

Discussion

Treatment of mononeuritis multiplex depends on the underlying disease.

When there is underlying cutaneous polyarteritis nodosa, early intervention with immunosuppressants might be beneficial as the rash often precedes neurological symptoms. Early immunosuppression can prevent the development of mononeuritis multiplex.

The inflammatory response caused by the cutaneous polyarteritis nodosa should be reduced initially with oral prednisolone for three to four weeks or with three doses of pulsed intravenous methylprednisolone if it is severe.

Treating the mononeuritis multiplex might require other steroid sparing immunosuppressants:

- With advice from neurology specialists, intravenous cyclophosphamide can be given initially
- Oral azathioprine or cyclophosphamide can be given as maintenance treatment for one to two years.

Patients should be followed up in dermatology and/or neurology for at least 12 months after discontinuing treatment and then can be followed up by their general practitioner if they remain asymptomatic.

If remission persists, treatment should be stopped. If there are any signs of relapse—either livedo or neurological signs—they should be dealt with using the same therapeutic approach as above.

Patient outcome

The patient’s rash and paraesthesia resolved after starting prednisolone and azathioprine. Her symptoms improved with this treatment, therefore she was managed as an outpatient in dermatology without referral to neurology. Treatment with azathioprine will be maintained for one to two years and if remission persists it will be stopped. Any signs of relapse, either livedo or neurological signs, will be dealt with using the same therapeutic approach.

We have read and understood BMJ policy on declaration of interests and declare that we have no competing interests.

Patient consent obtained.

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