Non-episodic angioedema associated with eosinophilia

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DESCRIPTION

A 26-year-old woman presented with a 1-week history of peripheral oedema, fever and polyarthralgia. Physical examination revealed symmetrical non-pitting oedema of her hands and legs (figure 1). Her medical history was unremarkable. Laboratory findings showed blood eosinophilia (3800/ μ L). The serum IgM level was within normal. Skin histopathology revealed slight interstitial oedema, dilated venous vessels with endothelial swelling and inflammatory cells extending into the subcutaneous tissue (figure 2). Her condition was diagnosed as non-episodic angioedema with eosinophilia (NEAE). Her symptoms quickly improved with prednisolone (15 mg/day). After tapering off of prednisolone, she has been recurrence-free for 2 years.

Angioedema with eosinophilia is classified as episodic angioedema with eosinophilia (EAE) or NEAE. EAE is characterised by recurrent episodes of angioedema, arthralgia and fever with a markedly high peripheral blood eosinophil count and high serum IgM level. NEAE is characterised by a single episode of persistent oedema of the extremities, peripheral eosinophilia, lack of an increased serum IgM level and a less severe clinical course than EAE.¹ NEAE generally affects young Asian women.¹ NEAE is also characterised by the effectiveness of low-dose prednisone or spontaneous remission.² The differential diagnosis includes Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) syndrome consisting of a severe rash, fever, lymphadenopathy, elevated liver enzymes and/or eosinophilia.³

Figure 2 Histopathology of the skin showing slight interstitial oedema, dilated venous vessels with endothelial swelling and inflammatory cells extending into the subcutaneous tissue.

Learning points

- ▶ Angioedema with eosinophilia is classified as episodic angioedema with eosinophilia or non-episodic angioedema with eosinophilia (NEAE).
- ▶ NEAE is characterised by a single episode of persistent oedema of the extremities, peripheral eosinophilia, lack of an increased serum IgM level.
- ▶ NEAE is also characterised by the effectiveness of low-dose prednisone or spontaneous remission.

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Contributors KS, YH, SN and MI were involved in managing the patient. KS wrote the first draft. SN made the pathology specimen. MI edited the manuscript.

Competing interests None declared.

Patient consent Obtained.

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Figure 1 Symmetrical non-pitting oedema of her legs.

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