Koebners phenomenon in Henoch-Schonlein purpura

Sir,
A 36-year-old male presented with red rash over both legs since four days. Since 20 years he has been getting similar lesions. Every year it usually starts in the beginning of summer season and resolves after taking treatment. He had history of abdominal pain and fever in the previous episodes. Now since four days he got lesions on lower limbs, which started as few red raised lesions around ankle and gradually increased in size and number to involve entire limbs and lower part of trunk. There was no history of sore throat or ingestion of any medications. He had history of renal stones.

On examination patient had multiple palpable purpuric papules and plaques on both lower limbs [Figure 1] extending upto the trunk. In the lumbar region, he had palpable purpura arranged in two horizontal and two vertical lines [Figure 2], indicating koebners phenomenon. As with the abdomen, there was no tenderness.

Biopsy from the lesion revealed dense mixed inflammatory cell infiltrate composed of lymphocytes, neutrophils and few eosinophils around the blood vessels. The vessel wall showed neutrophilic infiltrate, fibrinoid necrosis and nuclear dust. The endothelial cells were plump. Direct immunofloresence showed focal weak IgA and C3 deposition on the blood vessel wall [Figure 3]. The features suggested a diagnosis of Henoch Schonlein purpura.

The patient was started on systemic steroids and the lesions resolved completely.

DISCUSSION

Originally described as a tetrad of palpable purpura, arthritis, gastrointestinal involvement and renal glomerular involvement, Henoch-Schonlein Purpura is defined by the Chapel Hill Consensus Conference as a vasculitis affecting small vessels, involving deposition of IgA immune complexes that characteristically involves the skin, gastrointestinal system and glomeruli with or without arthralgia or arthritis.[1]

Here we describe Koebner’s phenomenon in a case of Henoch-Schonlein Purpura, which has been rarely reported in the literature.

Heberden first described Henoch-Schonlein Purpura HSP in 1801, in a young boy with abdominal pain, emesis, bloody stools, arthritis and a purpuric eruption. The eponymous term
HSP commonly occur in childhood. The commonest clinical feature is palpable purpura involving lower limbs and buttock, but may appear on other parts of the body. Gastrointestinal manifestations occur as abdominal pain, nausea, vomiting, diarrhea and constipation and passage of blood in the stools and rarely bowel intussusception. Most patients have polyarthralgia without frank arthritis. The most important manifestation of HSP is renal involvement, which usually produces protienuria, hematuria and red blood cell casts.[5] This condition is precipitated by a variety of agents including viral, bacterial, infections, drugs, certain foods, toxins and insects. Seasonal outbreaks have been described.[11] In our patients, there was classical presentation with childhood onset and seasonal outbreaks.

HSP is a clinical diagnosis, with confirmation by direct immunofluorescence and routine histology. Perivascular IgA deposits are characteristic of HSP and can help to distinguish it from other vasculitides.[1] In our case, Direct immunofluorescence and biopsy findings are very much consistent with the diagnosis of HSP.

Koebner’s or isomorphic phenomenon is occurrence of clinically similar lesions on uninvolved skin along sites of trauma in patients with pre-existing dermatosis. Koebner’s phenomenon has been reported in a variety of dermatoses.[3] There are only two case reports of koebners phenomenon in HSP.[4,5] Nischal et al., reported a case of koebners phenomenon in HSP over the pressure area.[8] Because of the paucity of reports, Boyd and Nelder have classified HSP as a questionable isomorphic phenomenon.[6] Unlike previous reports where koebners phenomenon occurred over pressure area, in this case it occurred along the line of scratching.

The exact pathomechanism of koebners phenomenon in HSP has not been elucidated. Namazi[7] reported that increase of tryptase levels following trauma could be the cause of Koebner phenomenon seen in psoriasis and Naukkarinen et al.[8] also showed that mast cell tryptase and chymase are potential regulators of neurogenic inflammation in psoriatic skin. Therefore, tryptase may play an important role in the pathogenesis of the Koebner phenomenon.[9] Although not extensively studied in vasculitis, recent studies showed that immuno-enzyme-histological staining of tryptase (a marker for mast cell activation) was found in cutaneous allergic vasculitisis[10] or HSP nephritis.[11]

Our case, together with the previous reports[4,5] establishes the presence of koebners phenomenon in Henoch Schonien purpura. Therefore, further studies should be performed to elucidate the relationship between mast cell activation and the Koebner phenomenon in the acute stage of HSP.

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