Letters to the Editor

Lipophagic granulomatous panniculitis misdiagnosed as purpura in an 8-year old girl

Sirs,

In 1989, Winkelmann et al. initially described in childhood a rare skin condition that they termed lipophagic granulomatous lipoatrophy (LGL). LGL is an idopathic self-limiting form of lobular granulomatous panniculitis with mild systemic features, a favourable outcome, and typical histologic findings. The presenting manifestations are large nodules and plaques in the lower extremities slowly evolving into subcutaneous atrophy, in the absence of internal organ involvement. Different skin conditions mimick LGL, in particular the lobular non-suppurative panniculitides associated with connective tissue diseases such as dermatomyositis (JDM) and systemic lupus erythematous (SLE), and those secondary to trauma or infection. The differential diagnosis also includes the idioopathic panniculitides associated with systemic symptoms, i.e. Weber-Christian disease and Rothmann-Makai syndrome (3,4), and cytopathic histiocytic panniculitis (5,6).

In April 1999, a previously healthy 8-year-old white girl developed multiple cutaneous violaceous patches resembling bruises on the anterior aspect of the legs and on the calves. After the exclusion of hematologic disorders, cutaneous vasculitis related to Group A Streptococcus was diagnosed. Despite a normal ASO titer and a negative throat culture, oral penicillin was unsuccessful prescribed. Due to the suspicion of food allergy, milk was excluded from the diet, but the skin alterations continued to spread in number and size.

In October 1999 the girl was seen in our Unit; her physical examination was unremarkable except for the presence of multiple cold, violaceous, non-tender nodules over the tibiae and patches of subcutaneous atrophy on the lateral sides of both legs (Fig. 1). Laboratory tests including the erythrocite sedimentation rate, complete blood count, transaminases, lactic dehydrogenase, creatine kinase, C-reactive protein, total protein and albumin, creatinine, fibrinogen, complement, amyrase, immunoglobulins, angiotensin converting enzyme, and anti-streptolisin titer were normal. Viral and bacterial infections, including Borrelia burgdorferi, were excluded by normal serological tests. Antinuclear antibodies, c-ANCA, p-ANCA, and anti-endomyosum antibodies were absent. X-rays of the legs excluded bone abnormalities; magnetic resonance imaging with gadolinium showed a slight symmetrical mass reduction of the calf muscles; the subcutaneous fat was normal, and no signal alterations were observed in the bones. Angio-MRI of popliteal vessels did not reveal arterial obstruction. As the skin alterations progressively involved the arms, trunk and face, a nodule was excised; histology showed xantomatous infiltration of the subcutaneous fat by foamy histiocytes, atrophy of fat nodules and lymphoid inflammatory infiltrate consistent with the diagnosis of lipophagic granulomatous panniculitis.

Treatment with prednisone (0.5 mg/Kg/day) was followed by a progressive improvement of skin lesions; large patches of skin atrophy developed on both legs. After 6 months prednisone was tapered and withdrawn. After 2 years of follow-up the girl has not developed systemic features, large patches of atrophy in both calves are the only persistent abnormality, and no recurrences have been observed.

In conclusion, this pediatric case underlines the difficulty in diagnosing lipophagic panniculitis in childhood, as recently reported by Melchiorre et al. (7). The most common panniculitis in childhood is erythema nodosum, while other rarer conditions are inflammatory and involutional lipoatrophy (8,9). Before the report by Winkelmann et al., some children with LGL were diagnosed as having Weber-Christian disease or Rothmann-Makai syndrome. LGL is considered a distinctive subtype of lipoatrophic panniculitis in children that presents with erythematous nodules of short duration, a recurrence of skin alterations and typical replacement of fat by lipohagic giant cells leading to residual large patches of skin atrophy. In our patient an underlying diagnosis of SLE or JDM was excluded on the basis of clinical manifestations and laboratory tests. No history of trauma, infection, or vaccine administration before the onset of symptoms was reported. The suspicion of LGL might have reduced the delay between the appearance of the first skin alterations and the diagnosis. Despite its rarity, LGL should be suspected in children presenting with recurrent violaceous, non-tender nodules in the lower extremities that evolve into patches of subcutaneous atrophy, in the absence of systemic features.

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References

Fig. 1. Legs showing marked regional subcutaneous atrophy 4 months after the acute episode.