

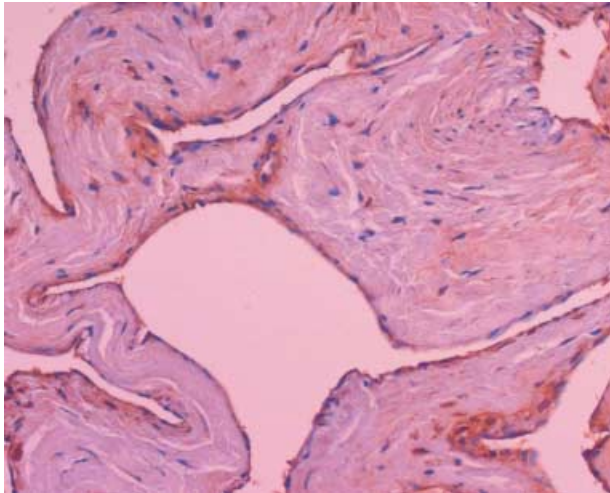


fig. 2 Dilated vascular structures with a thin monolayer lining and filled with amorphous, homogeneous materials on H&E stain.

rather than a proliferative tumoural change of lymphatic structures.¹

A 45-year-old woman presented with a solitary, tender, 7 × 8 mm sized, blue-to-black-coloured, hard nodule on the earlobe (fig. 1) for 3 years. The nodule was excised and reviewed microscopically. On H&E stain, the lesion consisted of dilated vascular structures with a thin monolayer lining and filled with amorphous, homogeneous materials or empty spaces. On immunohistochemical staining of factor VIII antigen, a negative result supported the presence of a lymphatic structure (fig. 2) and a final diagnosis of lymphangioma was made. There have been no local recurrences or other complications after 6 months.

Classic pathological classification of cutaneous lymphangioma is divided into four categories – cavernous haemangioma, cystic hygroma, lymphangioma circumscriptum and benign lymphangioendothelioma.¹ Cutaneous lymphangioma commonly develops as a soft subcutaneous nodule or large mass, typically present at birth or prior to 2 years of age. Cavernous haemangioma and cystic hygroma are commonly seen in the head and neck area, while lymphangioma circumscriptum and benign lymphangioendothelioma are preferentially involved in the limbs. Management involves various modalities but surgical treatment is required in most cases. The prognosis of cutaneous lymphatic malformation after excision depends on its location and its affect on nearby structures.

To the best of our knowledge there has been no report of cutaneous lymphatic malformation that developed in the earlobe as a solitary nodule in an adult. Lesions with various degrees of lymphangiectasia rarely occur in association with congenital or acquired lymphoedema² or after radiotherapy³, and are indistinguishable from classic

lymphangioma circumscriptum or cavernous lymphangioma. Our case presented unique clinical and pathological findings with adult-onset, and did not satisfy any kind of classic categories. Also, there was no history of lymphoedema or previous radiotherapy. This suggests that there are unclassified forms of cutaneous lymphatic malformation with a more diverse clinical and pathological presentation. Enjolras¹ classified lymphatic malformations into two broad categories – microcystic or macrocystic, including the malformations developing in the deep portion of the head and neck. This classification is more applicable to our case, which showed a limited lesion. The reason why there was no rapid increase in size or numbers might be explained by its anatomical location, that is to say, the limited space to grow and spread out.

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Unusual psoriasiform lesions in a patient with Hyper-IgE syndrome

Editor

Hyper-immunoglobulin E (IgE) syndromes (HIES) are a very rare primary immunodeficiency characterized by the clinical triad of high serum levels of IgE (> 2000 IU/mL), severe dermatitis, and recurring staphylococcal cutaneous and sinopulmonary infections.¹ We report here a case of hyper-IgE syndrome with unusual psoriasiform lesions. To our knowledge this association has not been reported previously.

A 17-year-old boy presented with erythroderma for 2 months and a history of recurrent dermatitis and furunculosis since 1 month of age. His skin problems began as multiple, exudative, pruritic eruptions, which had always been diagnosed as eczema or atopic dermatitis and treated with corticosteroids, topical moisturizer, antibiotics, and antihistamines. However, dermatitis relapsed quickly and gradually became severe. One year ago, scattered erythematous plaques with white scales occurred on his trunk and arms. Two months ago lesions evolved into erythroderma with very little itching. During his teenage years, he was often afflicted with recurrent skin infections, pneumonia, bronchitis and bronchiectasia. He was also noted to have a propensity to develop cutaneous fungal infections of the nails and elsewhere on his skin. He was born in a consanguineous marriage. His younger sister died of pulmonary infection with severe eczema at 1 year of age, and his parents had no history of asthma or atopic dermatitis.

Physical examination showed marked growth retardation for his age; his height was 140 cm and weight was 30 kg. However, no intellectual or mental abnormality was detected. The coarse facial features were obvious, including increased width of the nose, a full lower lip, and thickening of the nose and ears. Erythroderma with fine, powdery white scales coalesced with each other and involved the trunk and proximal extremities. On the scalp, numerous flaky, branny desquamations were seen on infiltrated erythematous plaques (fig. 1). Lichenified plaques were noted on the groin and the axilla. Multiple



fig. 1 The scalp showing numerous flaky, branny desquamations on infiltrated erythematous plaques.

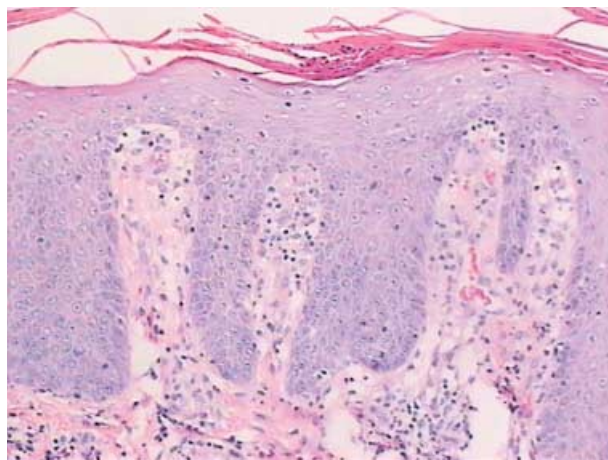


fig. 2 Skin biopsy revealed psoriasiform features (haematoxylin and eosin staining, original magnification $\times 100$).

atrophic scars on the back, which were the result of recurrent furunculosis, were still present.

Laboratory analysis revealed markedly elevated serum IgE level (78.743 IU/mL of 0–100 IU/mL) and eosinophilia (11.7%, $4\text{--}10 \times 10^9/\text{L}$). Testing of IgG and IgM to HSVI and IgG to HSVII were positive. Chest X-rays showed intrapulmonary interstitial hyperplasia, right lower lateral bronchiectasia and right pleural adhesion. Ultrasonography revealed splenomegaly.

Skin biopsy revealed epidermal hyperkeratosis and confluent parakeratosis associated with rete ridges, which were uniformly elongated. Exocytosis of neutrophils and Munro's microabscesses were also seen. In the dermis papillary oedema and dilated capillaries with perivascular lymphocytic infiltration were prominent. All these indicated psoriasiform features (fig. 2).

A diagnosis of hyperimmunoglobulin E syndrome was made based on the clinical features and a high serum IgE level.

So far the underlying causes of HIES are still not known. A half of HIES cases occur in infants; some are inherited.² Linkage analysis of 19 families demonstrated linkage to the proximal arm of chromosome 4q. However, genetic heterogeneity exists.³ In the case presented here, death of a sibling in similar circumstances implied an inherited tendency in this family. More recent studies have focused on T-helper 1 cell (Th1)/Th2 cytokine imbalance.⁴ The latest research has supported and confirmed that secretion of IFN- γ into the medium is defective in HIES patients; this impairs neutrophil chemotaxis, and, as a result, increases susceptibility to infection.⁵

Psoriasiform lesions in HIES patients are rare and we suggest that this is the first case reported in the literature. It implies that a disorder of the immune microenvironment in HIES lesions, particularly imbalance of Th1/Th2

cytokine, could cause additional changes aside from eczema dermatitis. Recurrent infections are another possible cause of psoriaform lesions occurring in this patient. However, whether psoriasiform lesions are independent or related to HIES still requires further study.

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Lyell's syndrome skin lesions treated by Veloderm®

Editor

Lyell's syndrome,¹ also known as toxic epidermal necrolysis (TEN), is a drug-related dermatological disorder characterized by epidermal sloughing, mucous membrane involvement, fever, pain, and systemic symptoms.

In TEN topical treatment minimizes heat and fluid loss, prevents infections, reduces pain, and provides a moist, clean wound environment suitable for re-epithelialization.² Lyell's cutaneous lesions are usually managed as superficial burns or as skin graft donor sites. Biological materials³ such as cadaver allografts can potentially expose the patient to infections, and can be difficult to find. Synthetic temporary wound dressings are often expensive (hyaluronic acid matrix), can cause soreness (silver ione-hydrofibres), or, according to our experience, have not shown remarkable results (alginates, hyaluronic acid ester film dressing). For these reasons we tested a new material

(Veloderm®, BTC srl, Ancona, Italy) consisting of a natural cellulose-microfibre biofilm, the effectiveness and excellent tolerability of which has been described in the treatment of burns and superficial wounds^{4,5} (unpublished report, G Borda, N Venturo, M Negrosanti; Clinic of Dermatology, University of Bologna); it has also recently been evaluated in a controlled clinical study as a dressing for skin graft donor site areas.⁶

A 33-year-old man was admitted to our intensive care unit for burns because of an intense and extended cutaneous rash, accompanied by mucositis involving the conjunctivas, oral cavity and genitalia, with erosions, bleeding and crusts. One month previously the patient had started a lamotrigine treatment, known as a possible cause of Stevens-Johnson/Lyell syndrome. He showed a widespread papulo-erythematous dermatitis with flat atypical target lesions, some of which were covered by bullae. Within 2 days the lesions had spread over almost the whole skin, converging in large macules. The area of sloughing covered over 30% of the total body surface area. Nikolsky's sign was positive. Leucopenia and hepatitis were present. The patient was hospitalized in our Intensive Care Unit for Burns and isolated in a single room, on an air-fluidized bed, in appropriate temperature and humidity conditions and in a controlled microbial environment.⁷ The treatment consisted of fluid rehydrating infusions, enteral and parenteral nutrition, intravenous immunoglobulins (0.5 g/kg) for 5 days,⁸ mild antiseptics and emollients for the mucosal lesions of the oral cavity, and hydrating, lubricating and antibiotic solutions for the conjunctival lesions. No systemic corticosteroids were given.⁹ The very painful and exudating cutaneous lesions were treated with mild antiseptic solution compresses (0.05% clorexidin) until the fourth day. When the sloughing process and the exudative phase had ceased, we applied Veloderm® on the denuded skin areas, covering it with dry sterile gauze. Veloderm® adhered to the lesions and in most cases it was not necessary to renew it (fig. 1) until spontaneous detachment at complete wound re-epithelialization and healing, which was achieved in 1 week (fig. 2). It was also effective in the critical-to-heal areas of decubitus. Besides facilitating the medical staff's task and decreasing hospitalization costs, the low frequency of dressing change reduced the patient's discomfort, pain and risk of bacterial contamination; clinical and the microbiological exams of the treated areas were always negative for infections. Veloderm® also decreased fluid, caloric and evaporative losses, thus preventing electrolyte imbalance. Indeed, besides the complete wound re-epithelialization, within 7 days the general clinical conditions of the patient improved: his fever lowered, and leucocyte and transaminases returned to normal values. The patient was discharged from the Intensive Care Unit for Burns after 12 days. Six months later only ocular