of jellyfish that causes the sting, it can be useful to apply vinegar, a 1:1 aqueous solution of sodium bicarbonate, or a saturated solution of magnesium sulfate in a solution of sodium chloride.

Jellyfish sting reactions are very common on the Spanish coast in summer. Therefore, although these reactions tend to be local and short-lasting, we consider it important to know how to deal with them and to be aware of the less common reactions in order to diagnose them correctly and provide early treatment.

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Follicular Lymphoma With Paraneoplastic Autoimmune Multiorgan Syndrome☆

Síndrome multiorgánico autoinmune paraneoplásico asociado a linfoma folicular

To the Editor:

Introduction

Paraneoplastic pemphigus is a rare entity first described by Anhalt et al. in 1990.[1] It is an autoimmune disease of the skin and mucosa, associated with neoplasm, which is generally lymphoid in origin. Because several organs are involved as well as the skin and because the physiopathologic mechanisms associated with the mucosal, skin and internal-organ lesions are not limited to the presence of antibodies specific to adhesion molecules, in 2001, Nguyen et al.[2] proposed the term paraneoplastic autoimmune multiorgan syndrome (PAMS).

We describe a patient with PAMS associated with non-Hodgkin lymphoma.

Case Description

A 69-year-old man with a personal history of cerebrovascular accident and type 2 diabetes mellitus visited our department with oral erosions and ulcers that had appeared 6 months earlier.

Physical examination revealed erosive glossitis, cheilitis, pseudomembranous conjunctivitis, and ulcerative keratitis. The patient also presented erythematous scaly lesions on the scalp, maculopapular lesions and reddish-purple plaques on the torso and legs, hyperkeratosis with fissures on the palms and soles, and erosive lesions on the glans and scrotum (Figs. 1 and 2); all these signs appeared 2 months before the patient visited our department.

A skin biopsy showed lichenoid interface dermatitis with necrotic keratinocytes (Fig. 3). Direct immunofluorescence showed intercellular deposits of immunoglobulin (Ig) G and IgC3 in the epidermis. Indirect immunofluorescence showed intercellular deposits when monkey esophagus was used as a substrate and was negative when rat bladder was used. Immunoblotting identified envoplakin (210 kDa) and periplakin (190 kDa) antibodies.

A study was performed to search for an occult neoplasm. Computed tomography of the chest and abdomen revealed

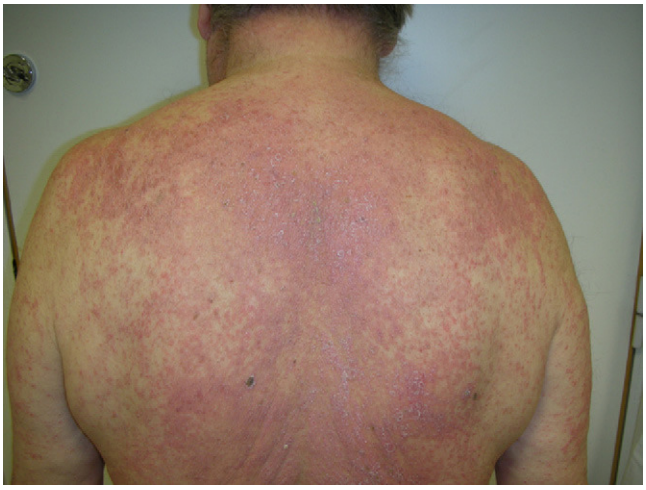

Figure 1 Reddish-purple maculopapular lesions grouped in plaques on the back.

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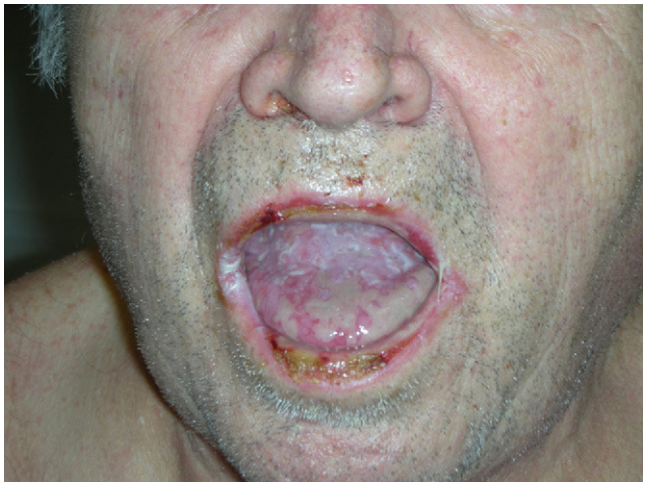

Figure 2 Tongue erosions and erosive cheilitis with crusting.

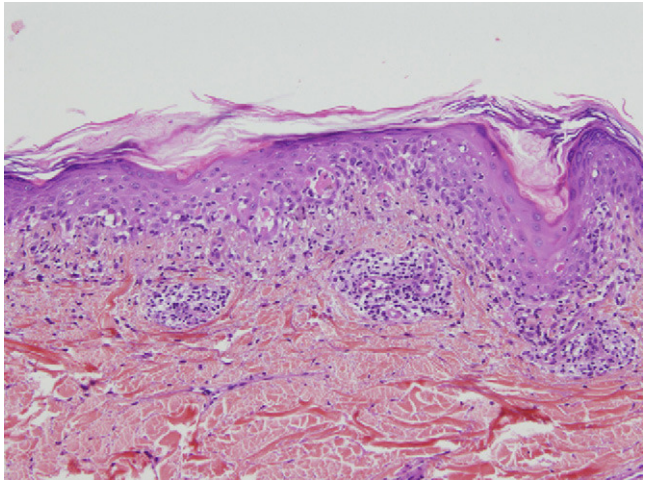

Figure 3 Lichenoid interface dermatitis with necrotic keratinocytes (hematoxylin–eosin, ×100).

enlarged mesenteric, mediastinal, and axillary lymph nodes, and a mass in the retroperitoneal soft tissue.

Lymph-node and bone-marrow biopsies were compatible with non-Hodgkin follicular lymphoma and a diagnosis of PAMS associated with non-Hodgkin follicular lymphoma was reached. Treatment of the underlying neoplasm was instated with 8 cycles of chemotherapy using vincristine, rituximab, cyclophosphamide, and prednisone; 5 mg/kg/day of ciclosporin and 1 mg/kg/day of methylprednisone were also administered at gradually decreasing doses to treat the cutaneous manifestations. Complete remission of the lymphoma was achieved with no recurrences to date, and the skin lesions resolved completely in 2 weeks. After 3 years of follow-up, the patient presents only mild stomatitis, which is being treated with 100 mg/day of ciclosporin.

PAMS is a heterogeneous autoimmune syndrome that involves several internal organs; it is associated with a neoplasm and presents clearly defined clinical, histologic, and immunologic characteristics. The clinical findings of paraneoplastic pemphigus are varied and may resemble pemphigus, lichen planus, erythema multiforme, or graft-versus-host disease. Stomatitis is usually present and often an early sign of the disease—so much so that its absence should call a diagnosis of paraneoplastic pemphigus into question. It is characterized by very painful erosive lesions.[3,4]

Anatomic pathology may show acantholysis but, unlike in common pemphigus, this is less marked and may be accompanied by an intense lichenoid mononuclear infiltrate in the dermal–epidermal junction, with vacuolar degeneration, suprabasal desquamation, and keratinocyte necrosis (lichenoid dermatitis).[4]

Direct-immunofluorescence findings show intercellular immunoreactants (IgG and complement), as in pemphigus, although IgG and/or complement are also commonly found in the basement membrane; this fact is useful in the differential diagnosis of common pemphigus with paraneoplastic pemphigus or PAMS. It should be remembered that, in some cases, direct immunofluorescence tests may be negative, possibly because of the predominance of lichenoid lesions or the presence of necrotic tissue in the biopsies.[5]

Serum antibodies against plakins (envoplakin, periplakin, desmoplakin 1 and 2, plectin, and an uncharacterized 170 kDa protein) have been detected in all epithelia. However, in a particular subgroup of patients, antidesmoplakin antibodies may be absent and antienvoplakin and antiperiplakin antibodies are therefore considered to be more specific in PAMS.[5,6]

Paraneoplastic pemphigus is a mucocutaneous manifestation of a severe PAMS and precedes diagnosis of the neoplasm in approximately a third of patients. Clinical suspicion of paraneoplastic pemphigus is essential for early diagnosis and treatment of both PAMS and the neoplasm, and thus for preventing fatal complications, such as bronchiolitis obliterans.[7,8]

Our patient presented with a case of PAMS associated with lymphoma. Diagnosis was made early and, unlike most cases associated with malignant tumors reported in the literature, the outcome was excellent, with the patient alive more than 2 years after diagnosis.

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Abortive or Minimal-Growth Hemangiomas. A Review of 14 Cases☆

Hemangiomas abortivos o mínimamente proliferativos. Revisión de 14 casos

To the Editor:

Infantile hemangiomas are frequent, benign endothelial tumors that express the marker GLUT1.[1] The diagnosis is generally clinical, based on morphology and a clinical course characterized by a phase of rapid postnatal proliferation followed by a slow spontaneous involution.[2,3] In some cases, when growth is minimal, these tumors are referred to as minimal growth hemangiomas, arrested growth hemangiomas, or abortive hemangiomas (AHs).[4–7]

Our aim was to retrospectively evaluate the clinical and histologic characteristics of AH. We examined the database and photo archives of the dermatology department at our hospital for the period of January 2006 to June 2010. Cases of infantile hemangioma in which the proliferative component of the tumor accounted for less than 25% of the total surface were selected for the review.

Eighteen cases of AH were identified, but 4 were eliminated because of lack of data. The remaining 14 cases affected 13 patients with a female to male ratio of 3:1 (10 girls and 3 boys). All infants were born full term (time of gestation >38 weeks) with a weight greater than 2.760 g (mean weight 3.260 g). During gestation, 2 mothers presented with urinary infection and 1 showed signs of preeclampsia. In 3 cases, a history of classic infantile hemangioma in a sibling was reported. The age of the infants at first consultation ranged between 1 day and 6 months (mean: 2.5 months, median: 3 months). Follow-up was carried out until 7 to 48 months of age. AH was present at birth in 71% of cases and appeared in the first 2 weeks of life in 29% of cases. Sixty-four percent of the tumors were located on the lower half of the body (face: 0; scalp: 1; upper limbs: 4; trunk: 4; lower limbs: 8). Cases were classified as focal AH (42%), partially segmental AH (29%), and segmental AH (29%). The common characteristic in 100% of cases was telangiectasias on normal or reddish skin (with a reticular distribution in 75% of the segmental tumors). Areas of pallor were seen in 50% of AHs and congenital bruise-like lesions were observed in 14% of cases. Proliferation was minimal and took the form of predominantly peripheral reddish papules in 64% of AHs and a few red spots in 22%; no growth was observed in 14% of cases (Figs. 1–3). Fading of the lesions was evident in 86% of cases, starting between 8 and 12 months of age (Figs. 1 and 2). In 2 cases in which the lesion showed no signs of involution, the final follow-up visit was made at 7 months of age. One segmental AH located in the groin ulcerated (7%). One girl presented with 2 AHs simultaneously. Another infant developed 1 classic infantile hemangioma and 1 AH. The 5 arrested growth tumors that were biopsied were positive for the marker GLUT1 (Fig. 2D). Minor developmental anomalies were detected in 50% of patients (syndactyly, sacral dimple [2 cases], nevus sebaceus on the scalp [Fig. 2C], preauricular tags, conjunctival abnormalities, hyperpigmented nevus, cleft lip, and transient pseudocoarctation of the aorta).

Classic infantile hemangioma is a GLUT1-positive tumor characterized by a rapid postnatal growth phase that is followed by a slow involution phase. Incidence is higher in premature infants and in the female sex, with the ratio of girls to boys ranging from 1.5:1 to 3.5:1. These tumors occur predominantly on the head and neck (60%), with focal hemangiomas appearing more frequently than segmental ones. The risk of ulceration has been estimated at 15%.[3,7] In our case series the female to male ratio (3:1) was within the range reported in the literature and the lesions were predominantly focal or partially segmental. Most of the hemangiomas showed signs of regression. Histologic examinations, when performed, revealed GLUT1 expression. Postnatal growth of AHs was minimal and there were no premature infants in the group studied. Only 1 tumor was located on the head, and only 1 lesion, located in the groin, ulcerated (7%).

Few studies have described this type of arrested growth infantile hemangioma and proposed a differential diagnosis with capillary malformations, noninvoluting congenital hemangiomas, or other vascular anomalies. Corella et al.[5] demonstrated that AH is a true form of hemangioma through histologic findings and GLUT1 expression. It has recently been suggested that dermatoscopy may be useful for diagnosis.[6] In a retrospective study involving 47 patients, Suh and Frieden[7] indicated that AHs have a particular clinical appearance, occur more frequently on the

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