Primary Cutaneous Angioplasmocellular Hyperplasia

Hiperplasia angioplasmocelular cutánea primaria

To the Editor:

The term primary cutaneous angioplasmocellular hyperplasia was proposed in 1995 to describe skin lesions characterized histologically by a vascular proliferation associated with a polyclonal inflammatory infiltrate of plasma cells; such lesions were first described on the trunk of young adults.\(^1\) We present a case of this rare condition.

A 40-year-old man with no past history of interest and who did not smoke or drink consulted for an asymptomatic lesion that had been present on his chest for 2 months. He did not recall a previous injury, insect bite, or pre-existing lesion. Physical examination revealed an erythematous-violaceous nodule measuring 2.5 × 2 cm on the anterior aspect of the chest (Fig. 1). The nodule was firm, was not adherent to deeper planes and did not blanch on pressure. There was a central ulcer that was covered by fibrin. Nothing remarkable was found on examination of the rest of the skin and the patient was in a good general state of health.

An incisional biopsy that included the border and center of the lesion revealed partial ulceration and epidermal hyperplasia with moderate acanthosis. A vascular proliferation constituted predominantly by thin-walled capillaries was observed in the reticular and papillary dermis (Fig. 2). A mainly perivascular inflammatory infiltrate formed principally of plasma cells with no atypia or mitoses was also observed; immunohistochemistry showed this to be a polyclonal infiltrate. Tissue culture and periodic acid-Schiff, Giemsa, and Warthin-Starry stains were negative. It was decided to excise the complete lesion. Histology confirmed the earlier biopsy findings and revealed the presence of Russell bodies. The vessels were dilated capillaries and venules. Some of the capillaries presented a prominent endothelium and focal endothelial vacuolization (Fig. 3). Additional tests, including complete blood count, biochemistry, coagulation studies, immunoglobulins, protein electrophoresis, antinuclear antibodies, and complement, were normal or negative. Serology was negative for syphilis and Lyme disease. Urinary protein electrophoresis and the chest x-ray presented no abnormal findings.

A diagnosis of primary cutaneous angioplasmocellular hyperplasia was made on the basis of the clinical and histological characteristics of the lesion and the results of the additional tests.

The patient presented no recurrence or systemic involvement after 9 months of follow-up.

The majority of reports of primary cutaneous angioplasmocellular hyperplasia involve single patients,\(^2,3\) with the exception of the series published by Hsiao et al.\(^4\) The condition is characterized clinically by erythematous-violaceous or brownish papules or nodules measuring between a few millimeters and 2 to 3 cm in diameter and that occur most commonly on the head and trunk. The lesions are usually single, though a case in which the patient had 2 simultaneous lesions has been reported.\(^4\) Central ulceration and a peripheral erythematous halo are common findings. No trigger factor has been identified, although a relationship with previous injury or with varicella has been described in some cases.\(^4\) At the time of diagnosis, the lesions may have been present for weeks to years.\(^1,3,4\) Histologically these lesions are characterized by a central ulcer with peripheral epidermal hyperplasia with acanthosis and a variable degree...
of hypergranulosis. In the dermis there is a proliferation of
dilated capillaries and venules and an inflammatory infiltrate
formed predominantly of polyclonal plasma cells with
no atypia or mitoses. Although the infiltrate tends to be
perivascular in early lesions, it acquires an interstitial
distribution in more advanced lesions. Though cell proportions
are variable, plasma cells make up more than 60% of the
infiltrate; lymphocytes, neutrophils, and eosinophils are present
in small numbers. Foci of prominent endothelial cells with
no atypia or mitoses and of endothelial vacuolation are
common.

The clinical and histological characteristics of previously
reported cases and of our patient are presented in Table 1.
The presence of Russell bodies—homogeneous, intensely
eosinophilic structures formed of immunoglobulins secreted
by plasma cells—has not been described in any of the previ-
ously published cases, but they may be observed in lesions
with a marked plasma cell component, whether reactive or
neoplastic.  

Plasma cells are not common in healthy skin, though they
are much more prevalent in mucosas. Their numbers also
increase in areas of infected or injured skin. Infections such
as leprosy, chronic folliculitis, syphilis, leishmaniasis, and
bacillary angiomatosis are associated with a more intense
plasma cell infiltrate; serology and specific stains are use-
ful to reach a diagnosis in such cases. Other conditions to
be considered in the differential diagnosis include the pseudo-
dolymphomas and reactions to arthropod bites, which are
clinically similar but histologically different, presenting a
dense perivascular lymphocytic inflammatory infiltrate with
lymphoid follicles and isolated plasma cells.

Neoplastic infiltrates of plasma cells are observed in
extramedullary cutaneous plasmacytoma (primary or sec-
ondary to myeloma), but the cells are monoclonal and
the vascular component is minimal.

Among the dermatoses with a predominantly vascu-
lar component, the most important disease that must be
included in the differential diagnosis is pyogenic granu-
loma, which has a lobular histologic pattern that is not
present in angioplasmocellular hyperplasia. The vessels in
pyogenic granuloma are of smaller size and the infiltrates
are formed mainly of lymphocytes, although they can con-
tain plasma cells in ulcerated or long-standing lesions. In
the histological differential diagnosis we should also consid-
erated angiolymphoid hyperplasia with eosinophilia,\textsuperscript{4} Kaposi sarcoma,\textsuperscript{7} epithelioid hemangiendothelioma,\textsuperscript{1} and cutaneous epithelioid angiomatous nodule.\textsuperscript{8}

In summary, we have presented a new case of a patient with a lesion on the center of the chest with the clinical and histological characteristics described to date for primary cutaneous angioplasmocellular hyperplasia. Our patient’s lesion was large and contained Russell bodies, a finding not previously described in this condition.

In agreement with other authors, we consider this to be a reactive hyperplasia, although no triggering stimulus can be found in the majority of cases.

References


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