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REVIEW ARTICLE

Multinucleate Cell Angiohistiocytoma

A Review and Report of Four Cases

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Multinucleate cell angiohistiocytoma (MCA) was first characterized by Smith & Wilson-Jones. Although only a few cases have been published, this very characteristic benign tumor is probably not rare. The clinical pictures are firm circumscribed papules, mainly of the hands, which progress slowly over the years. The histologic features show an increase of capillaries and venules in the reticular dermis, overlaid by epidermal hyperplasia. Bizarre-shaped multinucleate cells are present between the vessels, and factor XIIIa-positive interstitial cells are increased in number. MCA can be easily recognised if the pathologist is aware of the diagnosis. We present here the clinical, histologic and immunopathologic features of 4 new cases and review the literature. ***Key word: benign tumor.***

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OWN OBSERVATIONS

Clinical findings

The patients were 3 females and one male of 35 to 72 years of age. They all had a few papules (1 to 7) located on the dorsum of the hand or wrist for 1 to 9 years. Despite the fact that all papules were distributed on the hands, none of our patients remembered local trauma. The lesions were well circumscribed papules of 4 to 8 mm diameter (Fig. 1), and they had a red to violaceous coloration. In 2 cases, the lesions were firm and the suggested diagnosis was fibroma. In one patient, we observed papules having an annular distribution which was very similar to the features observed in granuloma annulare. One described a slow increase in the number of papules over 9 years. None of our patients experienced spontaneous regression of the lesions. Patients 3 and 4 complained of pruritus, and the diagnosis of lichen planus was proposed in one of these patients. All had a good general condition and there were no associated cutaneous diseases.

Histopathological examination and immunohistochemical analysis

The cutaneous biopsies were fixed in Bouin solution, included in paraffin and stained with hematoxylin-eosin-safran. Immunohistochemical examination was performed using the following antibodies: factor XIIIa 1/100 (Behring Diagnostic), leucocyte common antigen 1/100 (Dako), B-cell CD 45 R 1/40 (Dako), T cell UCHL1 1/80 (Dako), MAC 387 1/100 (Dako), factor VIII (Ortho Diagnostics Systems).

In each of the 4 patients, one or two distinct papules were totally excised and analyzed by conventional microscopy. Similar histopathological changes were observed in the six papules that were examined. The main changes were located in the superficial and mid-dermis. On both sides, the lesions were poorly demarcated from the normal-appearing dermis. In almost all lesions, the dermal changes were overlaid by acanthotic epidermis. The rete ridges were enlarged and increased in length, showing in some cases a psoriasis-like pattern. These epidermal changes were not only due to the localization of the lesions on the hands, because the observed acanthosis was well limited on both sides of the lesions. The basal layer was not hyperpigmented, as it is in dermatofibroma. The very superficial papillary dermis had a normal appearance. In the reticular dermis, there was an increase in the number of vessels which were often surrounded by an infiltrate of inflammatory cells, mainly lymphocytes (Fig. 2). The lumina of these enlarged capillaries and venules were moderately dilated, and the endothelial cells showed enlarged nuclei and abundant cytoplasm. Some of the endothelial cells were protruding in the vessel lumina. There were neither extravasated erythrocytes nor hemosiderin deposition. Between these vessels, there were many elongated cells which were very similar to the fibroblasts and dendrocytes usually observed within the normal dermis, but they were increased in number in most of our cases. Some had very little cytoplasm and an elongated nucleus, and others were shorter and had a round nucleus. These cells were intermingled with large and bizarre-shaped cells (Fig. 3). Some were triangular; others had a geometric shape with thin cytoplasmic prolongation. They often had multiple nuclei, usually two to five, and abundant dense cytoplasm. In one of the six papules, there were multinucleate cells with nuclei arranged in a ring-like fashion. Thickened collagen bundles with a horizontal distribution were observed in 3 cases in the central part of the lesions. In patient 2, some of the collagen bundles were hyaline. The underlying hypodermis was normal in all cases. Orcein straining did not show significant changes of the elastic tissue.

The inflammatory cells located around the dermal vessels expressed common leucocyte antigen and were positive with UCH L1 antibody. None were CD 45 R-positive. Elongated cells from the interstitial cell population stained strongly positive with anti-factor XIIIa antibody. A minority of the interstitial cells were factor XIIIa-negative. The large multinucleate cells were factor XIIIa- and MAC 387-negative. Only the endothelial cells were labelled with anti-factor VIII.

REVIEW OF THE CASES PUBLISHED

Between 1985 and 1995, 20 cases of MCA have been reported (1–5). MCA occurred in 18 cases in women and only in 2 cases in men. The age range of these patients was 37–74, and the mean age was 56. The duration of the disease was 4 months to 17 years and there was a slow increase in the number of papules over the years, but spontaneous disappearance of some lesions was observed. The lesions were often located on the limbs, especially on the dorsal part of the hands or wrist in 9 of the 20 reported patients and in all cases of the present series. The legs and thighs were also frequently affected, and lesions of the face were reported in 2 patients (3, 4). In one patient, multiple papules of the chest were observed (3). The elementary lesions were dome-shaped papules from 2 to 15 mm diameter and of erythematous or violaceous colour. The consistence was firm and the papules were well demarcated from the normal skin. In one case described by Wilson-Jones et al. (3) and in our case No. 2, the papules had an annular shape. Two of our patients com-

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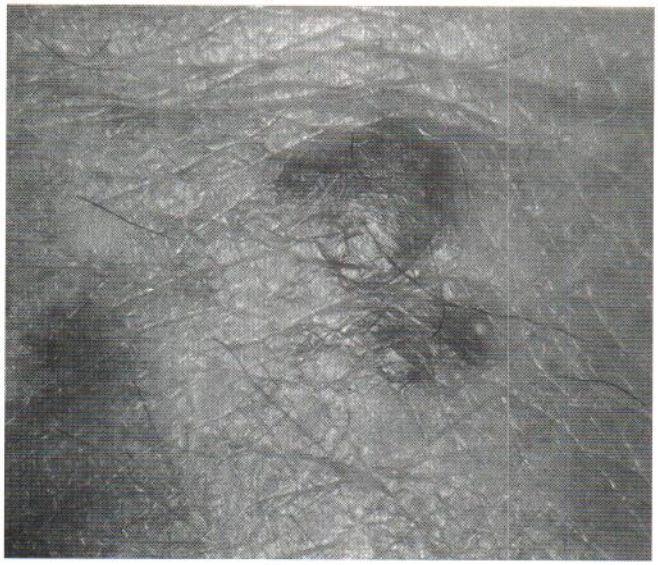

Fig. 1. Isolated erythematous papules of the dorsum of the left hand observed in patient 1.

plained of pruritus, but this symptom was not reported in the previous reports.

On the basis of the 24 reported cases, the main clinical characteristics can be summarized as follows:

- occurrence of multiple small erythematous papules;
- specific site predilection on the limbs, mainly the dorsal part of hands;
- slow progression over years without associated disease.

The histopathologic and immunopathologic features of our cases are very similar to the initial description of Smith & Wilson-Jones and to the other reports. The remarkably constant histopathologic changes confirm that MCA is a distinct and recognizable entity. MCA is characterized by an increase of the

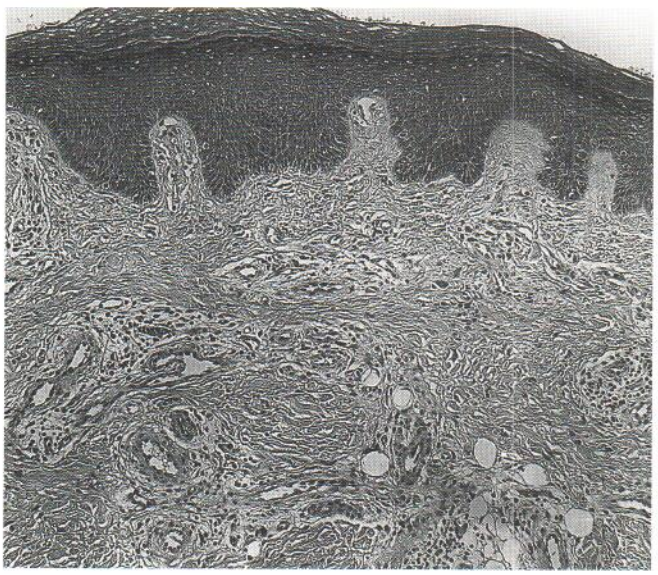

Fig. 2. Epidermal hyperplasia and vessels increased in number in the reticular dermis (hematoxylin-eosin-safran staining × 112).

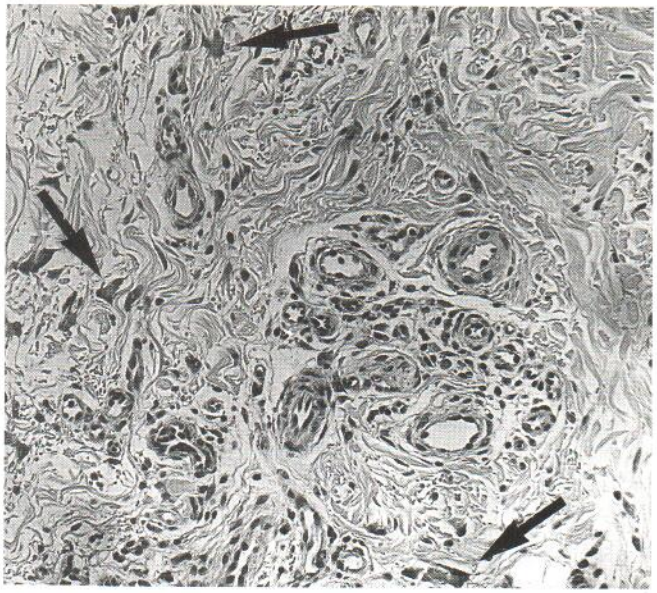

Fig. 3. Multiple multinucleate cells (*arrows*) between capillaries and venules of the reticular dermis (HES × 280).

dermal capillaries and venules, which show prominent endothelial cells and moderately enlarged lumina. The dermal collagen bundles can be thickened. Around the vessels, a sparse infiltrate of T-lymphocytes is often observed. Multinucleate cells of geometrical shape and angulated cytoplasm are located within the dermis. The morphologic features of these cells are characteristic but not specific, because these cells can also be observed in atrophic vascular histiocytomas and in fibrous papule of the nose (3). The normal fixed connective tissue factor XIIIa-positive cells are increased in number, but not as much as in true histiocytomas. Because the majority of cells of histiocytomas are factor XIIIa-positive, the term "angiohistiocytoma" was proposed by Wilson-Jones et al. (1, 3). As previously demonstrated by Smolle et al. (2) and by Wilson-Jones et al. (3), the multinucleate cells present in our cases were factor XIIIa- and MAC 387-negative. The histogenesis of the multinucleate cells remains unclear (2, 3). They could represent degenerate connective tissue cells or macrophages that have lost their functional properties. In addition to these histologic features described by Smith & Wilson-Jones (1), we have noted an epidermal hyperplasia overlying almost all lesions.

The differential diagnosis of MCA was developed in the work by Shapiro et al. (5). Briefly, MCA must be histologically distinguished from dermatofibroma and from angiofibroma. Because Kaposi's sarcoma (KS) shares many clinical features with MCA, differential diagnosis is based on histologic criteria: there are more spindle cells and hemosiderin deposition in KS and there is lack of multinucleate cells (3). As shown by the clinical diagnosis suggested before histologic examination in both our series and in the previously reported patients, MCA can mimic granuloma annulare, lichen planus and even lymphocytoma. The differential diagnosis with these diseases can easily be made by the histopathologist.

MCA has a benign course and treatment is not recommended, except surgical excision in order to differentiate MCA from

other skin diseases which have a similar presentation. Dermatologists and histopathologists should be aware of this characteristic and distinct entity.

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