Case 18
A 44 year-old Thai female from Prachinburi

Chief complaint: Multiple asymptomatic erythematous to brownish papules on trunk and extremities for 10 years

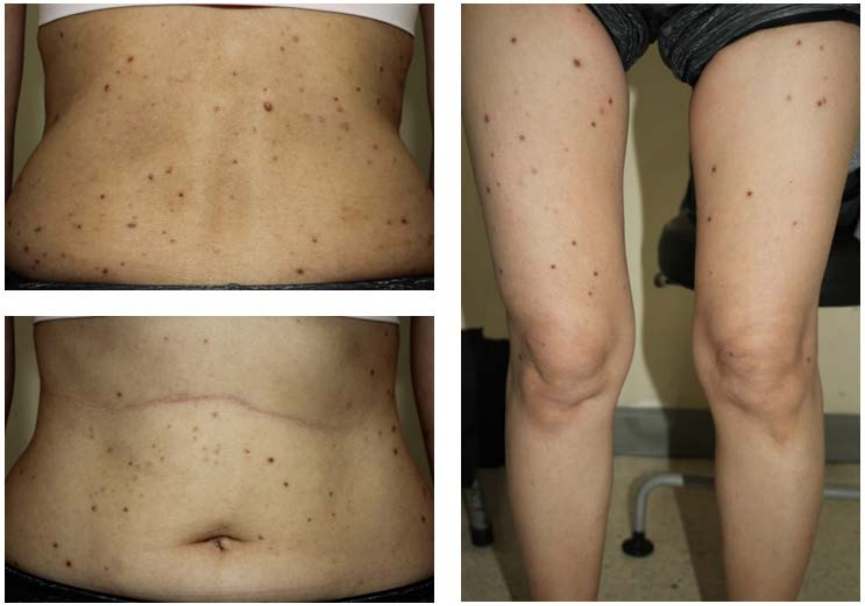

Fig. 18.1

Present illness:

- She presented with a 10-year history of multiple asymptomatic erythematous papules initially appeared on all extremities and trunk, then the lesions turned into erythematous to brownish papules. The lesions slow-progressively increased in number particularly on the lower extremities. They were painless and non-itchy. The oral cavity, conjunctivae and teeth had no lesion.
- She denied any systemic symptom, history of weight loss, fever, or herbal use.

Underlying disease: No underlying disease

Family history: No family history of similar cutaneous lesions

Dermatological examination: (Fig. 18.1)
Multiple discrete firm, erythematous to brownish dome-shaped papules with smooth surface on trunk and extremities.

Physical examination:
Other systemic examinations revealed no abnormality.

Investigations:

- **CBC**
  - WBC 13,190/mm^3 (N 81%, L 14%)
  - Hb 12.9 g/dL, Hct 39.3 %
  - Plt 262,000/mm^3
- **ANA**: negative
- **ESR**: 6 mm/hr

Histopathology: (S18-002094A, skin, left lower back)

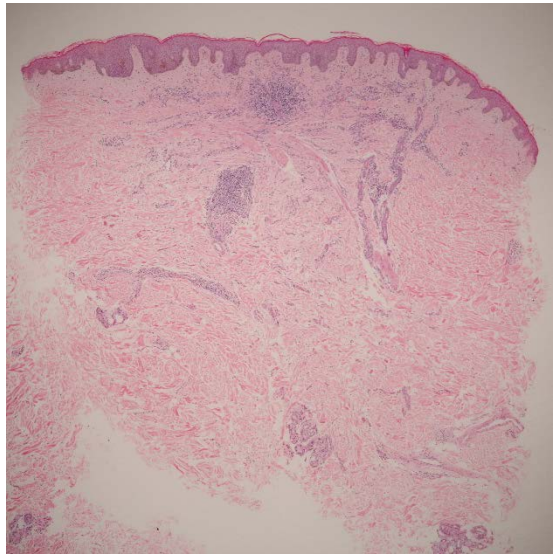

Fig. 18.2

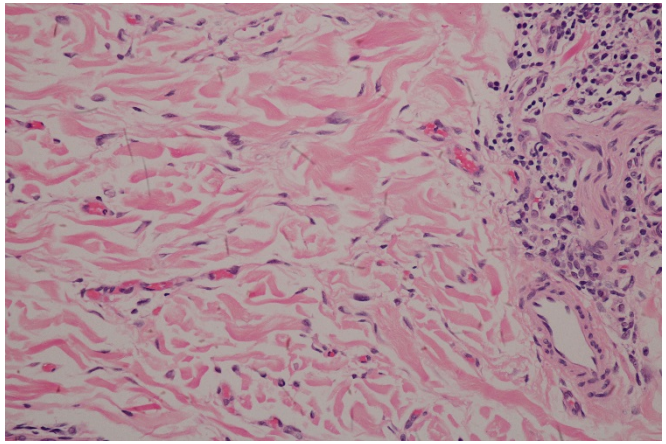

Fig. 18.3

- Epidermal hyperplasia, increased number of dilated capillary (fig. 18.2)
- Perivascular lymphocytic infiltrate and scatter polygonal and spindle cells
- Higher magnification shows multinucleated cells (fig. 18.3)

Immunohistochemistry:

- Positive vimentin of vascular channel, stellate cells, and multinucleated giant cells
- Positive CD34 of vascular channels
- Positive factor XIIIa and CD68 of stellate cells

Diagnosis: Multinucleate cell angiohistiocytoma

Treatment:

- Pulsed dye laser

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Discussion:

Multinucleate cell angiohistiocytoma (MCAH) was firstly described in 1985 by Smith and Wilson Jones.[1] MCAH is a rare benign fibrohistiocytic and vascular proliferation with unknown etiology. Fewer than 150 cases have been reported.[2] MCAH shows absence of extracutaneous involvement and malignant transformation. The majority of cases diagnosed in the 5th to 8th decade is predominantly in women with 3:1 female to male ratio.[3-6] Clinical manifestation of MCAH is multiple sharply demarcated, firm, reddish-brown to violaceous dome-shaped papules with a smooth

surface.[3,5,7] The usual affected distributions are the extremities, including the distal thighs, calves, and dorsal aspect of the hands. Generalized form is rare.[4-8] The lesions are mostly asymptomatic, however, few studies reported mild pruritus.[7,10] None of the patients had any prior history of trauma, insect bites, or scabies.

The diagnosis is based on the histopathological examination which reveals a proliferation of vessels predominantly of capillaries and venules with numerous spindle-shaped mononuclear cells and multinucleated giant cells in the upper to mid dermis. Although not pathognomonic for MCAH, multinucleated giant cells with bizarre shape, angulated cytoplasm and several nuclei arranged at the cell periphery are the most specific histopathological finding.[3,6,9] The dermal collagen bundles can be thickened. A perivascular and perifollicular infiltrate of lymphocytes, histiocytes, mast cells and occasional neutrophils, and plasma cells is often present.[5-7] Immunohistochemically, mononuclear dendritic cells express vimentin, factor XIIIa, CD68, MAC387, and lysozyme, suggesting a fibrohistiocytic in origin (involving both fibrocytes and macrophages). The vascular endothelial cells are positive for antibodies to Factor VIII, vimentin, CD31, and CD34.[3-6] Multinucleate giant cells are strongly positive to vimentin, negative to factor XIIIa and variable reactive to CD68.[3-5] Although there is a lack of monocyte-macrophage markers in multinucleated giant cells, electron-microscopic studies, demonstrating abundant rough endoplasmic reticulum and nuclear membrane reinforcements (Zonula nucleus limitans), lysosomes and pinocytic vesicles, still support their fibrohistiocytic differentiation. These may represent their different maturation or degeneration.[10-11]

Although MCAH should be usually differentiate from dermatofibroma, angiofibroma, Kaposi ‘s sarcoma, bacillary angiomatosis, and pyogenic granuloma, the diagnosis of MCAH can be made by its highly characteristic histopathological and immunohistochemical features.

The pathogenesis of MCAH is still unclear. Most studies suggested a reactive process rather than a neoplasm. Some investigators proposed a possible mechanism whereby an interaction between mast cells and factor XIIIa fibrohistiocytic cells leads to cell proliferation and vascular hyperplasia.[10-11]

MCAH is a benign disease. The lesions usually persist and slowly progress, however, spontaneous regression was occasionally observed.[3,5,12] The treatments is usually not necessary except for cases of generalized lesions, pruritic or cosmetic reasons.[5] The treatment options include surgical excision, cryosurgery, carbon dioxide laser, argon lasers, and intense pulsed light.[3,7,12] In this case, patient was treated with pulsed dye laser and followed up monthly, the lesions were partially resolved with residual hyperpigmentation.

Reference:

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