QUIZ SECTION

MJS PUBLISHING

ActaDV Acta Dermato-Venereologica ActaDV Advances in dermatology and venereology

An Asymptomatic Brown Plaque on the Face: A Quiz

Daoning ZHANG[1–4], Pingping LIN[1–4] and Hang LI[1–4]*
*[1]Department of Dermatology and Venerology, Peking University First Hospital, No. 8 Xishiku Street, Xicheng District, Beijing, [2]National Clinical Research Center for Skin and Immune Diseases, [3]Beijing Key Laboratory of Molecular Diagnosis on Dermatoses, and [4]NMPA Key Laboratory for Quality Control and Evaluation of Cosmetics, Beijing, China. *E-mail: drlihang@126.com*

A 53-year-old woman was referred to our hospital with a 9-month history of an asymptomatic grain-sized brown lesion on her left cheek. It gradually enlarged and became indurated. She denied history of trauma and systemic symptoms or relevant family history. The physical examination revealed a 10 x 6 mm depressed brown indurated plaque on her left cheek, with ill-defined border (Fig. 1a). Dermoscopy revealed pigmented network and linear streak (Fig. 1b). A biopsy specimen was taken and the histopathology showed large amounts of spindle-shaped cells parallel to the surface, interlaced and proliferated in the middle and lower portion of the dermis, as well as the upper portion of the subcutaneous septae, and some with large nuclei (Fig. 2a, b). The immunohistochemical staining of spindle-shaped cells was positive for smooth muscle actin (SMA) and factor XIII, and negative for CD34, S-100 and Ki-67 (Fig. 2c, d).

What is your diagnosis?

Differential diagnosis 1: Dermatomyofibroma

Differential diagnosis 2: Dermatofibrosarcoma protuberans

Differential diagnosis 3: Dermatofibroma

Differential diagnosis 4: Fibroblastic connective tissue nevus

See next page for answer.

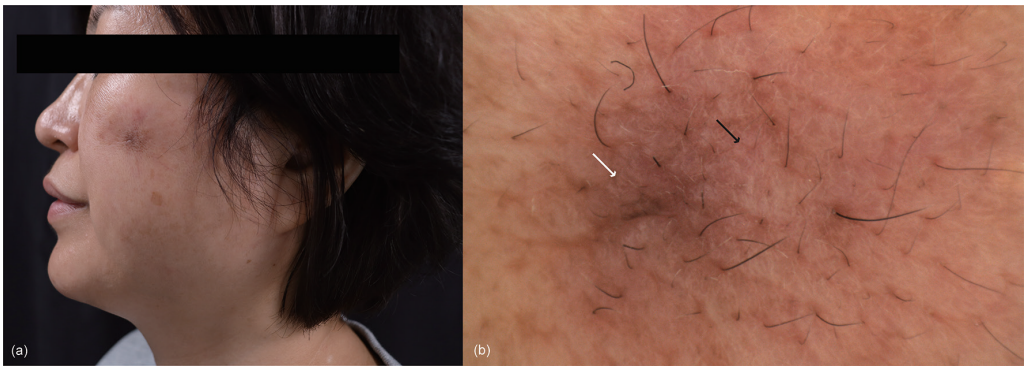

Fig. 1. Clinical appearance and dermoscopy. (a) A 10 x 6 mm flat brown indurated plaque on the left cheek, with ill-defined border. (b) Dermoscopy revealed pigmented network (white arrow) and linear streak (black arrow).

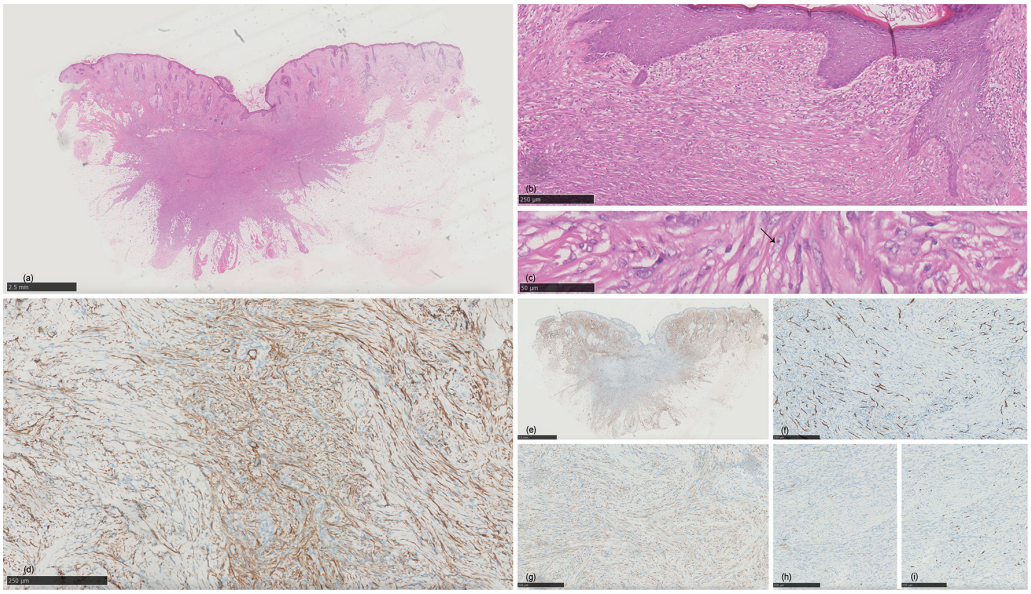

Fig. 2. Histology and immunohistochemistry findings. (a) Large amounts of spindle-shaped cells interlaced and proliferated in the middle and lower fat lobules of the dermis (haematoxylin-eosin; scale bar=2.5mm). (b) Spindle-shaped cells parallel to the surface.(haematoxylin-eosin; scale bar=250 μm) (c) Spindle-shaped cells with large nuclei (black arrow; scale bar=50 μm). (d) Immunohistochemistry showed SMA-positive cells (anti-SMA; scale bar=250μm) (e–f) Immunohistochemistry showed CD34 negative cells (anti-CD34; (e) scale bar=2.5mm and (f) scale bar=250 μm). (g) Immunohistochemistry showed Factor XIII-positive cells (anti-factor XIII; scale bar=250 μm). (h) Immunohistochemistry showed S-100-negative cells (anti-S100; scale bar=250 μm). (i) Immunohistochemistry showed Ki-67-negative cells (anti-Ki67; scale bar=250 μm).

ANSWERS TO QUIZ

An Asymptomatic Brown Plaque on the Face: A Commentary

Acta Derm Venereol 2024; 104: adv40377
DOI: 10.2340/actadv.v104.40377

Diagnosis: Dermatomyofibroma

Dermatomyofibroma is a rare benign cutaneous tumour of fibroblasts and myofibroblasts. It predominantly affects young females, and commonly involves the shoulder, upper arm, axillary region, neck, and upper trunk (1). In a review of 56 dermatomyofibroma cases, the shoulder (13/56) and upper arm (7/56) were most frequently affected. Other locations including the neck, thigh, chest wall, and back were also mentioned (2). Facial dermatomyofibroma is extremely rare, with only 1 reported case affecting the nasion (3).

In our case, as clinical presentation was not typical or apparent, a careful inspection was needed to achieve a precise diagnosis. While initial visual inspection revealed only a non-specific asymptomatic brown plaque that mimics many common dermatologic conditions including seborrheic keratosis or melasma, careful palpation was imperative to augment the assessment of the lesion, whose visual changes are unimpressive or nonspecific. Palpation revealed induration and depth, prompting further examinations including pathology and immunohistochemistry examinations, where dermatofibroma, dermatofibrosarcoma protuberans (DFSP), and fibroblastic connective tissue nevus (FCTN) should be differentiated as they share a similar pathological appearance. The arrangement of spindle-shaped cells parallel to the skin surface and involvement of the upper subcutaneous septae, with positive staining for SMA and negative staining for CD34, collectively support the diagnosis of dermatomyofibroma and distinguish it from dermatofibroma, DFSP, and FCTN, which exhibit varying histological patterns or immunohistochemical features.

Histologically, dermatomyofibroma exhibits long fascicles of spindle-shaped cells parallel to the skin surface. The spindle-shaped cells are positive for vimentin and smooth muscle actin (SMA), while negative for S-100 and desmin (2). It commonly infiltrates the reticular dermis and the upper portion of the subcutaneous septae, presenting as long fascicles of spindle-shaped cells parallel to the skin surface with elongated nuclei. Diagnosis is made based on immunohistochemical results. The spindle-shaped cells are positive for vimentin and SMA, and negative for desmin, factor XIII, and CD34. However, focal expression of CD34 and factor XIII was seen in some cases (2,4). In our case, the spindle-shaped cells were positive for SMA and factor XIII, and negative for CD34, S-100, and Ki-67. Verhoeff–van Gieson stain for elastic fibres may also assist in diagnosis.

Dermatofibroma is a common benign dermal nodule that frequently affects the lower extremities. It can be divided into 2 types, including the fibrous type and cellular type (5). Positive immunohistochemical reactions for vimentin and factor XIIIa, as well as negative reaction for CD34, support the diagnosis of dermatofibroma. In our case, distinguishing between dermatomyofibroma and dermatofibroma through pathological features could have been challenging, but the clinical appearance and the distinctive pattern of the subcutaneous septae involvement provided valuable assistance in achieving a differential diagnosis.

DFSP is an uncommon fibroblastic cutaneous sarcoma of intermediate malignancy. It often presents as an asymptomatic and indurated raised plaque or nodule. Immunohistochemically, distinguishing between dermatomyofibroma and DFSP relies on specific markers. DFSP presents as strongly positive for CD34 and negative for factor XIIIa. In contrast, dermatomyofibroma demonstrates positive staining for vimentin and SMA as well as negative staining for desmin and CD34. Though focal expression of CD34 was seen in some cases (2,4), it is still the most significant distinguishing feature. Verhoeff–van Gieson stain for elastic fibres may also assist in the diagnosis of dermatomyofibroma. Molecular examinations such as the detection of COL1A1-PDGFB fusion gene are also helpful, with a high detection rate of DFSP (6). In this case, the involvement of the subcutaneous septae and the positive CD34 staining for endothelial cells may lead to potential confusion with atrophic DFSP. Cautious immunohistochemical analysis is necessary for precise diagnosis.

Other differential diagnoses including fibroblastic connective tissue nevus should also be considered. This is a rare variant of connective tissue nevi that presents as solitary, slowly growing asymptomatic firm plaque or nodule. Typically, FCTN usually presents at a much younger age and exhibits a predilection for the trunk, head, and neck. Histologically, spindle cells show a variable and disorderly orientation in FCTN compared with dermatomyofibroma. Immunohistochemically, FCTN is typically positive for CD34 with only focal SMA positivity observed in some cases, while dermatomyofibroma is usually positive for SMA (7).

Surgical excision is recommended for the treatment of dermatomyofibroma. Even with incomplete excision, no progression or recurrence has been reported (2). Therefore, punch biopsy and clinical follow-up could be an alternative approach. In our case, as it affected the cheek where a good cosmetic appearance should be considered, complete local excision was adopted. This underscores the critical correlation between the lesion's location and the selection of appropriate treatment.

In conclusion, this case indicated a rare location and non-specific clinical appearance of dermatomyofibroma that affected the cheek. A comprehensive analysis of the clinical characteristics and pathological features is helpful for precise diagnosis and the choice of appropriate treatment.

ACKNOWLEDGEMENT

The authors would like to thank the patients for granting permission to publish this information. They also thank Menglong Ran, MD, from Peking University First Hospital, for his assistance in dermoscopy, and Jingru Sun, MD, Ping Tu, MD, Xixue Chen, MD, and Yang Wang, MD, all from Peking University First Hospital, for their assistance in pathology.

REFERENCES

1. Gomez-Moyano E, Vera-Casano A, Martinez-Garcia S, Sanz-Trelles A, Crespo-Erchiga V. Two cases of dermatomyofibroma (plaque-like dermal fibromatosis). Int J Dermatol 2010; 49: 914–917.
2. Mentzel T, Kutzner H. Dermatomyofibroma: clinicopathologic and immunohistochemical analysis of 56 cases and reappraisal of a rare and distinct cutaneous neoplasm. Am J Dermatopathol 2009; 31: 44–49.
3. Fu S, Zhang J, Zhan Y, Zhang H, Herman EI, Lu Q, et al. A rare case report for dermatomyofibroma in nasion. Zhong Nan Da Xue Xue Bao Yi Xue Ban 2018; 43: 1037–1040.
4. Campagnolo OA, Morais CF, Ughini MO, Aoki CM, Menegazzo M. Dermatomyofibroma. An Bras Dermatol 2017; 92: 101–103.
5. Yamamoto T. Dermatofibroma: a possible model of local fibrosis with epithelial/mesenchymal cell interaction. J Eur Acad Dermatol Venereol 2009; 23: 371–375.
6. Lim SX, Ramaiya A, Levell NJ, Venables ZC. Review of dermatofibrosarcoma protuberans. Clin Exp Dermatol 2023; 48: 297–302.
7. de Feraudy S, Fletcher CD. Fibroblastic connective tissue nevus: a rare cutaneous lesion analyzed in a series of 25 cases. Am J Surg Pathol 2012; 36: 1509–1515.