· CASE ANALYSIS · · 临床病例讨论 ·

DOI:10.11817/j.issn.1672-7347.2018.09.017
www.csumed.org/xbwk/fileup/PDF/2018091037.pdf

A rare case report for dermatomyofibroma in nasion

FU Siqi[1], ZHANG Jing[1], ZHAN Yi[1], ZHANG Huiming[1], Edward I. Herman[2], LU Qianjin[1], ZHOU Ying[1], ZHANG Guiying[1]

(1. Department of Dermatology, Second Xiangya Hospital, Central South University, Changsha 410011, China; 2. St. Vincent's Medical Center, Bridgeport CT 06606, USA)

ABSTRACT Dermatomyofibroma is a benign and rare proliferation of myofibroblasts and fibroblasts of the skin. Dermatomyofibroma commonly locates at the shoulder and neck of young adults and adolescents. Other frequently affected anatomic sites are upper arms, thigh, chest wall, back, axillary region and abdomen. Herein, we reported a case of dermatomyofibroma occurred in the nasion. The asymptomatic firm nodule and histopathological features were consistent with dermatomyofibroma. Immunohistochemically, the tumor cells expressed vimentin, HHF35 and α-smooth muscle actin (α-SMA). The patient was followed up for 2 years after excision of the tumors and recurrences were not observed.

KEY WORDS dermatomyofibroma; nose; myofibroblastic tumor

罕见鼻部皮肤肌纤维瘤1例

付思祺[1]，张静[1]，湛意[1]，张慧明[1]，Edward I. Herman[2]，陆前进[1]，周英[1]，张桂英[1]

(1. 中南大学湘雅二医院皮肤科，长沙 410011；2. 圣文森特医学中心, 美国 康乃狄克州 布里奇波特 06606)

[摘要] 皮肤肌纤维瘤是一种少见的良性皮肤软组织肿瘤，病变为成纤维细胞和肌成纤维细胞增殖，常见于年轻成年人和青少年的肩部及颈部，其次好发于上臂、大腿、胸壁、背部、腋窝和腹部。本文报道1例罕见发生于鼻部的皮肤肌纤维瘤，此例肿物表现为无症状的坚硬的皮肤结节，病理上证实与皮肤肌纤维瘤相符。免疫组织化学结果显示，肿瘤细胞表达波形蛋白(vimentin)、肌特异性肌动蛋白(HHF35)和平滑肌肌动蛋白(α-smooth muscle actin，α-SMA)。手术切除后随访2年无复发。

[关键词] 皮肤肌纤维瘤；鼻部；肌纤维母细胞瘤

Date of reception: 2018-01-05

First author: FU Siqi, Email: fusiqi@csu.edu.cn, ORCID: 0000-0003-1060-8039

Corresponding author: ZHOU Ying, Email: zhouying001@csu.edu.cn, ORCID: 0000-0002-9051-1423; ZHANG Guiying, Email: lindazgy@csu.edu.cn, ORCID: 0000-0001-7617-0215

Foundation item: This work was supported by the Natural Science Foundation of Hunan Province, China (2018JJ3756).

Dermatomyofibromas was first described as a distinct benign mesenchymal cutaneous neoplasia of fibroblastic and myofibroblastic lineage by Kamino et al[1] in 1992. Clinically it is predominant in young female patients at a mean age of 28 years old and only a few cases have been reported in childhood[2]. It commonly locates on the shoulder of adult patients and on the neck of child patients[3]. Up to now, no case reported on the face to our knowledge. We describe a case of dermatomyofibroma arising in the nasion, which adds to the medical literature one more case on the face.

1 Case report

A 44-year-old female patient presented a solitary firm, ill-defined nodule in her nasion. The lesion had increased in size gradually since 8 months ago and had no symptoms. Examination revealed an 1.5 cm in diameter, ill-defined, skin-colored indurated nodule (Figure 1). There was no reported history of trauma. A deep incisional biopsy was performed. Histologic examination of the section showed poorly demarcated proliferation of spindle cells involved the dermis and the subcutaneous tissue. These spindle cells demonstrated a pale eosinophilic cytoplasm and elongated bland nuclei (Figure 2). Immunohistiochemical study was positive for vimentin (Figure 3A), HHF35 (Figure 3B) and α-smooth muscle actin (α-SMA) (Figure 3C), but negative for CD34, CD117, S-100, melan-A, desmin and P53. Based on the histologic and immunohistologic features, the diagnosis of dermatomyofibroma was made. The nodule was completely excision. There was no recurrence of the lesion during 24-month follow-up.

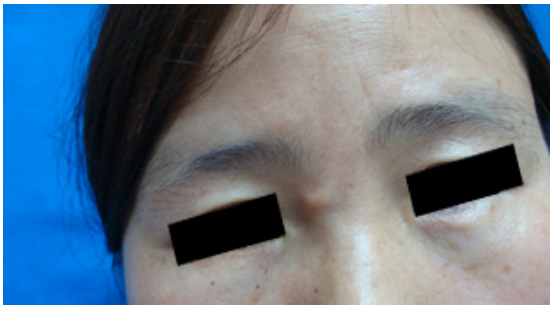

Figure 1 A solitary skin-colored indurated nodule (1.5 cm in diameter) in the nasion

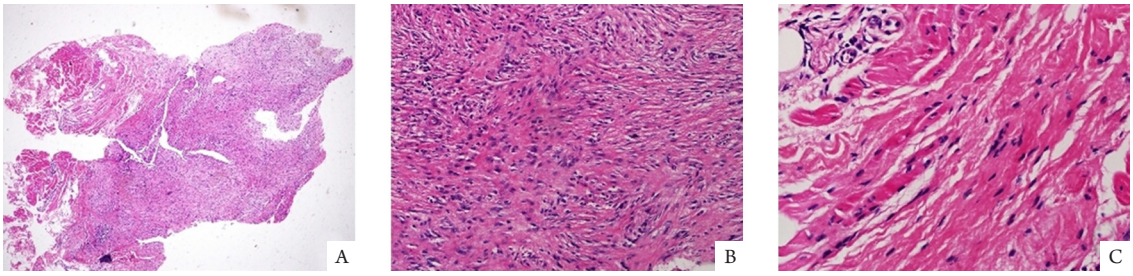

Figure 2 Spindle cell proliferation of intersecting bundles (HE staining)
A: ×40; B: ×200; C: ×400

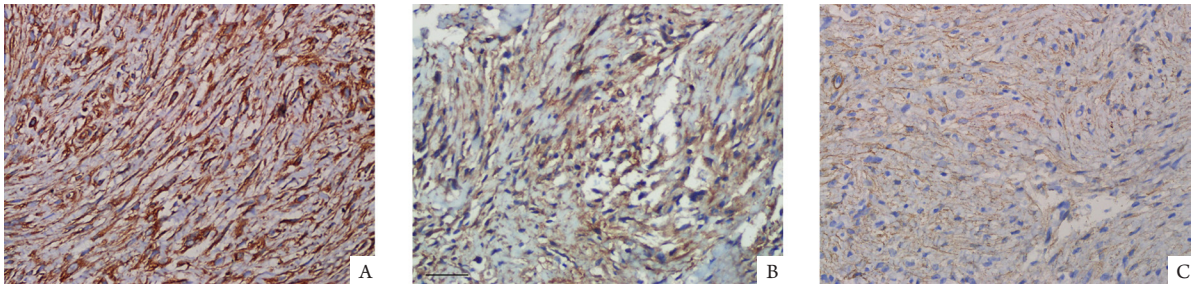

Figure 3 Immunohistochemical staining showing that the tumor cells are positive for vimentin (A), HHF35 (B), and α-SMA (C) (×400)

2 Discussion

Dermatomyofibroma is a benign and rare proliferation of myofibroblastsand fibroblasts of the skin, first described by Hügel[4] in 1991 and under the name of "plaque-form dermalfibromatosis". Kamino et al[1] in 1992 proposed the name of dermatomyofibroma. Clinically, it is a slow-growing, asymptomatic skin-colored to red-brown nodule or plaque. Dermatomyofibroma usually presents on the shoulder of young females, occasionally on the upper arm, axilla, chest, back, abdomen, neck and thigh. However, it also occurs in pediatric patients from 4-month-old infant to 16-year-old adolescent with the neck being the predominant location[5]. Generally, dermatomyofibroma is a solitary lesion below 2 cm in diameter, however, multiple[6] or extensive[7] lesions, even giant annular lesion with 15 cm diameter[8] have already been reported.

Histologically, dermatomyofibroma is a well-circumscribed plaque composed of long fascicles of neoplastic cells involving the reticular dermis and extending into the subcutaneous tissue. The cells which are spindle-shaped with pale eosinophilic cytoplasm and elongated nuclei are parallel to the skin surface. The adnexal structures are spared[9-11].

Dermatomyofibroma is one of the myofibroblastic and fibroblastic cutaneous tumor, the characteristic of immunohistochemistry confirm to myofibroblasts' feature. Mentzel et al[12] summarized the clinicopathologic and immunohistochemical features of 56 cases of dermatomyofibroma, tumor cells in 11 of 48 cases were positive for HHF35, a focal expression in 20 cases, whereas negative in 17 cases. In addition, 10 of 45 cases expressed CD34 focally. Another study[13] showed positive for vimentin and actin in most cases and negative or slightly positive for HHF35. They are negative for desmin, factor XIIIa, CD4, ALK1, and S-100. In our case, the tumor cells were positive for vimentin, HHF35, and α-SMA while negative for CD34, CD117, S-100, melan-A, desmin, and P53.

Dermatomyofibroma can be easily confused with with other spindle cell tumors, such as dermatofibroma, dermatofibrosarcoma protuberance (DFSP) and hypertrophic scar. Dermatofibroma are firm dermal nodules, usually less than 1 cm in diameter. The tumor is composed of prominant collagenous stroma and interstitial spindle cells which is positive for factor XIIIa, vimentin, but usually negative for HHF35 and CD34. DFSP is an uncommon slow-growing neoplasm of the dermis with tendency to invade the subcutis. DFSP is more cellular composed of densely packed monomorphous spindle cells arranged in a storiform pattern. Immunohistochemical staining of DFSP shows diffusely and strongly positive for CD34 and vimentin, but negative for HHF35. Hypertrophic scar shows whorled proliferation of fibroblasts and abundant collagenous stroma. Its immunohistochemical stains also show vimentin positive and HHF35 negative. In dermatomyofibroma, most tumor cells are immunohistochemically positive for HHF35 and vimentin.

As the location of dermatomyofibroma, the shoulder (23%) was the anatomic site most commonly affected, followed by the upper arm (12.5%), the neck (10.7%), the thigh (10.7%), the chest wall (7%), the back (5%), the axillary fold (3.5%), the abdominal wall (3.5%), the forearm (1.7%), the buttock (1.7%), and the popliteal fossa(1.7%)[11]. The tumor of our case report is located in the nasion of 44-years-old female patient, to our knowledge, this is the first case of dermatomyofibroma reported occurring on the nasion. Biopsy for histological and immunohistochemical study is recommended for further diagnosis. Excision and clinical follow-up are needed. To date, no instance of recurrence after excision has been reported, so simple excision seems to be sufficient treatment[5].

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(Edited by GUO Zheng)

本文引用： 付思祺, 张静, 湛意, 张慧明, Edward I. Herman, 陆前进, 周英, 张桂英. 罕见鼻部皮肤肌纤维瘤1例[J]. 中南大学学报(医学版), 2018, 43(9): 1037-1040. DOI:10.11817/j.issn.1672-7347.2018.09.017

Cite this article as: FU Siqi, ZHANG Jing, ZHAN Yi, ZHANG Huiming, Edward I. Herman, LU Qianjin, ZHOU Ying, ZHANG Guiying. A rare case report for dermatomyofibroma in nasion[J]. Journal of Central South University. Medical Science, 2018, 43(9): 1037-1040. DOI:10.11817/j.issn.1672-7347.2018.09.017