CASE REPORT

Linear dermatomyofibroma over the nape of neck: a report of an unusual case and literature review

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Abstract
Dermatomyofibroma is a rare cutaneous mesenchymal tumor of benign fibroblastic and myofibroblastic derivations. It predominantly affects young women, and it usually presents as a reddish-brown plaque or nodule, which is commonly located over the upper trunk. We report the case of a 41-year-old female patient who presented with progressive linear dermatomyofibroma over the nape of her neck. This case report expands the knowledge about the clinical and histopathological features of this rare, benign and cutaneous tumor.

INTRODUCTION
Dermatomyofibroma is a rare cutaneous mesenchymal tumor of benign fibroblastic and myofibroblastic derivations. It predominantly affects young women with a median age of 30 years. It has also been reported in prepubertal male children. Clinically, patients present with a long history of a slowly growing asymptomatic plaque or nodule associated with reddish-brown hyperpigmentation. The clinical presentation is indefinite and vague, and for this reason, histological and immunohistochemical (IHC) assessments are necessary to reach a final diagnosis. We report the case of a posterior cervical dermatomyofibroma in a female patient. The clinical, histopathologic and IHC features of this unusual presentation are described.

CASE REPORT
A 41-year-old female patient presented to a dermatology clinic with an asymptomatic lesion over the nape of her neck (Fig. 1). The lesion had been slowly progressing over 5 years. The patient denied any history of trauma. Her medical, surgical and medication histories were unremarkable. On examination, there was an ill-defined linear reddish-brown indurated plaque measuring approximately 7 × 3 cm over the nape of her neck. Based on the clinical presentation, the differential diagnosis included dermatofibrosarcoma protubersans (DFSP), dermatofibroma, morphea and hypertrophic scar. A skin punch biopsy measuring 3 mm at the maximum diameter was obtained under local anesthesia. The specimen was fixed in 10% formal saline and sent for histopathological examination.

Histopathological features
Sections showed an ill-defined dermal lesion consisting of bland-looking oval and spindle cells. The cells were arranged in vague interlacing fascicles, which extended to the superficial part of the subcutaneous fat and entrapped some adipocytes and blood vessels (Fig. 2A and 2B).
There was no evidence of necrosis, pleomorphism or increased mitoses. IHC staining was performed. The tumor cells were strongly positive for h-caldesmon and vimentin (Fig. 3A and 3B) and focally positive for desmin. Stains for factor 13A, CD34, S100 protein and CD56 were negative. A final diagnosis of dermatomyofibroma was made.

DISCUSSION

Dermatomyofibroma is a relatively rare benign mesenchymal tumor, which usually presents as a cutaneous plaque-like proliferation of fibroblasts and myofibroblasts. A similar condition was first reported in the German literature by Hügel in 1991 in a series of 25 cases under the name of ‘plaqueförmige dermale fibromatose’ (plaque-like dermal fibromatosis) [1]. Later in 1991 and 1992, Kamino et al. [2] reported a series of nine patients with the same condition, and they were the first to describe it under the name of dermatomyofibroma. Dermatomyofibroma has been reported in both males and females, but it predominately affects young females with a mean age of 30.8 years [3]. The lesion has a wide anatomical distribution [3], but it is most frequently located on the upper trunk such as the shoulder, which is the most common anatomic site for this lesion [2,3]. The upper arm, posterior aspect of the neck and axillary region are other possible but less frequent sites. The lesion is usually solitary, but there were two reported cases of multiple dermatomyofibromas in the literature [4,5]. The lesion is characterized by the presence of an ill-defined indurated plaque showing a variable and slightly erythematous discoloration. Dermatomyofibroma is usually small and on average measures between 1 and 2 cm at the maximum diameter [2]. However, larger lesions of 13 cm in diameter have been reported [6,7]. The differential diagnosis of the tumor includes DFSP, dermatofibroma, morphea and hypertrophic scar. The clinical diagnosis of dermatomyofibroma constitutes a challenge for clinicians because of its close resemblance to other similar lesions. Therefore, histologic examination and immunohistochemistry markers are necessary to reach a conclusive diagnosis. Histologically, the tumor is characterized by a non-encapsulated, ill-defined and plaque-like proliferation of uniform, slender and bland-looking spindle-shaped tumor cells that are located mainly in the reticular dermis. The tumor cells are arranged in elongated intersecting fascicles that are parallel to the overlying epidermis [2,3,8]. On high-power microscopic view, the uniform spindle cells are separated by thin collagen fibers, and these tumor cells have spindle- and oval-shaped nuclei without atypical features. Additionally, in some cases of dermatomyofibroma, there is an extension of tumor cells into the superficial subcutis, which is similar to the changes seen in our patient’s biopsy [3,8,9]. Moreover, adnexal structures in dermatomyofibroma are preserved, while elastic fibers are thickened, are increased and appear scattered [2,3]. The latter is an important microscopic finding for differentiating dermatomyofibroma from dermatofibroma and hypertrophic scar, where elastic fibers are often decreased [2,3,10]. Immunohistochemically, these cells stain negatively for CD34, factor 13A and S100 protein. This finding helps to exclude other lesions such as dermatofibroma, which stains positive for factor 13A and DFSP, where elastic fibers are often decreased [2,3,10].
Figure 3: (A) IHC stain for h-caldesmon is strongly positive. This finding is in favor of dermatomyofibroma. The tumor was, however, negative for CD34, factor 13A and S100 protein. IHC stain ×200. (B) IHC stain for C-34 is negative. Note the presence of positive internal control in some blood vessels. IHC stain ×400.

regression of the lesion [10]. Gomez-Moyano et al. reported the longest follow-up period of a non-excised lesion, which was present for 2 years and still remained stable during this time. Moreover, Mentzel et al. [3] reported a considerable number of cases that were managed by marginal and incomplete excision, in spite of that, none of these cases recurred or showed evidence of progression. Hence, the prognosis of dermatomyofibroma is excellent [5,7].

Our initial plan was to do a complete excision for both diagnostic and therapeutic purposes. However, due to the site in the nape of the neck, the extensive length of the lesion and its deep induration, the treating physician decided to do a biopsy first to confirm the diagnosis before proceeding to excision. After the biopsy result came out as benign, the physician explained to the patient the nature of the lesion and his plan to excise it, but the patient refused excision due to the benign nature of the lesion and some cosmetic concerns. The lesion remained stable with no changes after 6 months of careful follow-up.

To conclude, our patient's lesion is unusual in its location, size, shape and presentation, and to the best of our knowledge, only one case has been reported to date with a similar linear configuration in the upper neck [8]. Recognition of such conditions is important in order to distinguish it from other aggressive cutaneous neoplasms.

FUNDING
None.

CONFLICT OF INTEREST STATEMENT
None declared.

ETHICAL APPROVAL
The approval of the Institutional Review Board of King Saud University is not required for case reports.

CONSENT
A written informed consent was obtained from the patient.

GUARANTOR
The guarantors of the manuscript are Prof. Dr Fahad M. AlSaif and Prof. Dr Ammar C. AlRikabi.

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