Dermatomyofibroma in a young Irish female

Amy Ridge, Grainne Heuston, Darren Kilmartin, Pauline Marren, Birgit Tietz, Mary Laing

ABSTRACT

**Introduction:** Dermatomyofibroma represents a rare, benign mesenchymal neoplasm of fibroblast and myofibroblast differentiation. They are most often found in young adult women and male children. **Case Report:** We report a case of dermatomyofibroma, which to our knowledge is the first reported case in Ireland. Histologically, dermatomyofibroma typically demonstrates proliferation of uniform spindle cells arranged as well-defined intersecting fascicles, parallel to the epidermal surface. The tumor respects skin appendages and the epidermis remains unchanged. Elastic fibers are preserved. With regard to immunohistochemistry, the spindle cells are variably positive when stained with antibodies directed against actin and negative for desmin, S100, and CD34. There are no reports of metastases or recurrence of this benign tumor in the literature. **Conclusion:** Dermatomyofibroma typically occurs in young females and represents a benign tumor with no reported potential to recur or metastasize. It should be considered in the differential by the dermatologist when evaluating lesions resembling large dermatofibroma.

**Keywords:** Dermatomyofibroma, Fibroblasts, Myofibroblasts

INTRODUCTION

Dermatomyofibroma represents a rare, benign mesenchymal neoplasm of fibroblast and myofibroblast differentiation. They are most often found in young adult women and male children.

CASE REPORT

We report a case of dermatomyofibroma, which to our knowledge is the first reported case in Ireland. A 31-year-old female was referred to Dermatology by her general practitioner (GP) with regard to an unusual thickening of the skin on the left side of her neck (Figure 1). The lesion was present for approximately six months and gradually enlarging. The lesion was asymptomatic. She had no personal or family history of skin cancer. She had a significant sun exposure history, having lived in Australia for four years and had a blistering sunburn. She had no past medical history and took no regular medications. She had previously attended Dermatology for review of two nevi on her back which were both benign clinically.

On examination she had a 1 × 1 cm flesh colored plaque on her left neck with milia like cysts within it. There was no epidermal change. Histopathological examination of a 4 mm punch biopsy revealed a plaque like proliferation of fascicles of bland spindle cell with pale eosinophilic cytoplasm and elongated vesicular nuclei.
in the dermis (Figure 2). The epidermis was normal and there was no destruction of adnexal structures. On immunohistochemistry, the spindle cells expressed h-caldesmon (Figure 3) and were weakly positive for actin (Figure 4). There was also some weak nuclear positivity for β-catenin (Figure 5). Calponin, S100, and CD34 were negative. The proliferation was cytologically bland so there was no concern for malignancy. Histologically, the differential was superficial extra-abdominal desmoids fibromatosis, however, it is usually found in deeper soft tissue and not a superficial skin biopsy. Following discussion at the dermatopathology meeting and taking into consideration clinical features, histology, and immunohistochemistry, the consensus was that this skin lesion was consistent with dermatomyofibroma.

DISCUSSION

Dermatomyofibroma is a rare benign dermal neoplasm composed of fibroblasts and myofibroblasts.
It was first described in 1991 by Hügel as a “plaque like fibromatosis” [1]. It presents as an erythematous or brown asymptomatic 1–2 cm plaque. It is most common in females with a mean age of 28 and mostly occurs on the shoulder. Most often it is a single lesion but multiple lesions have been described [2]. In pediatric populations it has been reported to most commonly affect males, whereas it mostly affects females in the adult population. The reason for this is not fully understood, but is possibly related to female hormones [3]. Histologically dermatomyofibroma typically demonstrates proliferation of uniform spindle cells arranged as well-defined intersecting fascicles, parallel to the epidermal surface [4]. The tumor respects skin appendages and the epidermis remains unchanged [5]. Elastic fibers are preserved. With regard to immunohistochemistry, the spindle cells are variably positive when stained with antibodies directed against actin and negative for desmin, S100, and CD34. There are no reports of metastases or recurrence of this benign tumor in the literature. Clinically the differentials include dermatofibrosarcoma protuberans or a dermatofibroma but the clinical and histological features were not consistent in this case. The main histopathological differential is a superficial extra-abdominal desmoid fibromatosis, however, this is usually found in deeper soft tissue than in this case.

CONCLUSION
Dermatomyofibroma typically occurs in young females and represents a benign tumor with no reported potential to recur or metastasize. It should be considered in the differential by the dermatologist when evaluating lesions resembling large dermatofibroma.

REFERENCES
1. Hügel H. Plaque-like dermal fibromatosis/dermatomyofibroma. J Cutan Pathol 1993;20(1):94.
2. Viglizzo G, Occella C, Calonje E, Nozza P, Rongioletti F. A unique case of multiple dermatomyofibromas. Clin Exp Dermatol 2008;33(5):622–4.
3. Wollina U, Schönlebe J. Dermatomyofibroma—a rare mesenchymal tumor with maintained horripilation. Dermatol Ther 2019;32(4):e12967.
4. Rose C, Bröcker EB. Dermatomyofibroma: Case report and review. Pediatr Dermatol 1999;16(6):456–9.
5. Mortimore RJ, Whitehead KJ. Dermatomyofibroma: A report of two cases, one occurring in a child. Australas J Dermatol 2001;42(1):22–5.

**********
Author Contributions
Amy Ridge – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Grainne Heuston – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Darren Kilartin – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Pauline Marren – Acquisition of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Birgit Tietz – Acquisition of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mary Laing – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

Copyright
© 2020 Amy Ridge et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.
