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DERMATOFIBROMA OF THE GREAT TOE – A RARE CASE REPORT

Plastic Surgery

Surya Rao Rao Venkata Mahipathy* Professor & Head, Dept. of Plastic & Reconstructive Surgery, Saveetha Medical College & Hospital, Thandalam, Kanchipuram *Corresponding Author

Arjunan Babu Lakshmanan Consultant General Surgeon, Jaya Hospital, 3, Car Street, Sriperumbudur, Kanchipuram Dist. - 602 105

Susruthan Murali Consultant Pathologist, Sushrutha Diagnostics, Sri Amman Nagar, MS Nagar, Porur, Chennai - 600125

ABSTRACT

Dermatofibroma (DF), also referred to as cutaneous fibrous histiocytoma (CFH), is a common benign tumor of the skin presenting as a firm nodule located predominantly on the limbs and shoulder and pelvic girdles that often extends into superficial subcutaneous tissue. Dermatofibroma has many clinicopathological variants. This report presents a case of a dermatofibroma on a great toe that had been slowly growing for four years. Histopathologically, the relatively well-circumscribed dermal tumor was separated from the epidermis by a small grenz zone. The tumor tissue consisted of spindle-shaped cells with well-defined cell borders and spindly condensed nuclei. No cytologic atypia or mitotic figures were found. Immunohistochemically, the tumor cells showed negative staining for CD34 and S-100. Histopathological differential diagnoses of dermatofibroma include superficial acral fibromyxoma, cellular digital fibroma, superficial angiomyxoma, myxoid dermatofibrosarcoma protuberans and low-grade fibromyxoid sarcoma. Immunohistochemical staining can be useful in the differential diagnosis of these tumors. Pathologists should keep in mind the diagnosis of dermatofibroma when dealing with circumscribed, firm nodules presenting on the digits.

KEYWORDS

Benign, Dermatofibroma, Rare, Surgical Excision

INTRODUCTION

Dermatofibroma (DF) is a common benign fibrohistiocytic tumor of unknown etiology [1]. Due to varied histopathological aspects, synonymous designations include benign fibrous histiocytoma, histiocytoma cutis, nodular subepidermal fibrosis and fibrous xanthoma [2]. Over the last three decades numerous distinctive clinicopathological variants have been described. DFs are solitary, slowly growing nodules that usually appear during early to mid-adult life and can occur in response to minor trauma or insect bites [3]. They are most commonly found on the extremities, shoulder, and buttocks. DFs are relatively common and account for approximately 3 percent of all specimens submitted to dermatopathology [4]. DFs can be protuberant, plaque-like, or umbilicated with a storiform histopathologic pattern comprised of spindle-shaped and angular collagen synthesizing cells. Although any surface of the skin may be affected, DFs are most common on the lower extremities [5]. We present a case of dermatofibroma arising on a great toe which is rare. Although any surface of the skin may be affected, dermatofibromas are most common on the lower extremities [5]. Presentation of dermatofibroma on the digits is not commonly reported in the literature [6,7]. We describe the clinicopathological findings of digital dermatofibroma and also discuss the immunohistochemical staining results.

Case Report

A 54 year old male presented to our hospital with a swelling over the medial half of the left great toe for the past 4 years. It was spontaneous in onset and gradually progressive. The patient complains of occasional pain initially, but for the past 4 months the pain has worsened. On examination, a 2 x 1 cm firm swelling was present over the medial half of the nail region of the left great toe replacing that half of the nail plate complex. (Fig. 1)

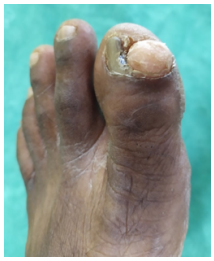

Fig. 1 – Pre-op picture showing the lesion

It was not warm or tender. The swelling not friable and was not fixed to the underlying terminal phalanx and keratinized all around. There was no regional lymphadenopathy. X-ray of the foot showed a soft tissue shadow over the medial aspect of the left great toe with no bony involvement. (Fig. 2)

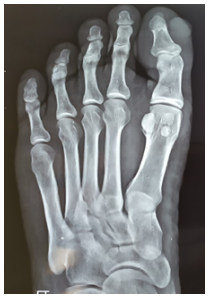

Fig. 2 – X-ray foot showing the soft tissue shadow with no bony involvement

A diagnosis of a benign soft tissue tumour was made and we proceeded for surgical excision. The lesion was excised in toto under local anaesthesia and sent for histopathological examination. (Fig. 3)

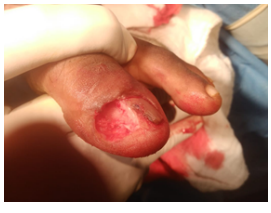

Fig. 3 – Photograph after excision of the lesion in toto

The resultant defect was closed primarily with 2 – 0 nylon sutures and a compression dressing was applied. (Fig. 4)

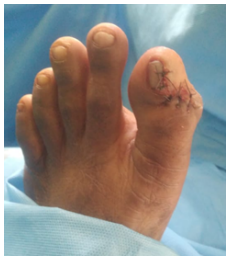

Fig. 4 – Immediate post-operative picture

Histopathology revealed a circumscribed lesion in the dermis composed of spindle shaped cells arranged as fascicles, bundles and focally in storiform pattern, with no evidence of atypia, necrosis or increased mitosis, thereby suggestive of a benign storiform dermatofibroma. (Fig. 5)

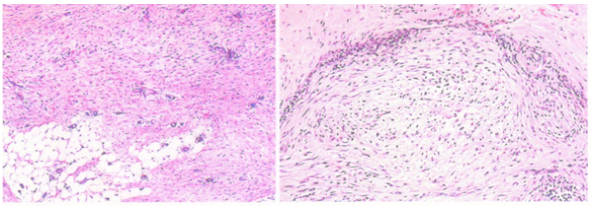

Fig. 5 – Microphotographs showing a circumscribed lesion in the dermis composed of spindle shaped cells arranged as fascicles, bundles and focally in storiform pattern

DISCUSSION

Dermatofibroma is a reactive hyperplastic response of the skin of unknown etiology, which is mostly seen on the extremities of young or middle-aged women [2,8]. Histologically, DFs are unencapsulated, well circumscribed tumors located in the reticular dermis with occasional extension into the superficial subcutis along interlobular septae. DFs are primarily composed of short spindle-shaped and stellate cells, sometimes with a mixture of giant cells, foamy macrophages, siderophages, lymphocytes, and plasma cells [9]. These factor XIIIa positive, benign fibrosing overgrowths of dermal dendrocytes [10] often induce overlying pigmented epidermal hyperplasia [11].

A wide variety of clinicopathological variants of dermatofibromas has been described over the last decade, including fibrocollagenous, vascular, angiomatoid, sclerosing, palisading, epithelioid, atypical, clear cell, myofibroblastic and cellular types. Histopathological differential diagnoses include superficial acral fibromyxoma, cellular digital fibroma, superficial angiomyxoma, cellular myxoma of soft tissue, myxoid neurofibroma, myxoid dermatofibrosarcoma protuberans and low-grade fibromyxoid sarcoma. Most importantly superficial acral fibromyxoma commonly express CD34, which is rarely found in dermatofibroma [2,12]. Even though there may occasionally be an increase of CD34 expression at the periphery of the lesion, which derives from the intrinsic reactivity of the surrounding stromal tissue response, this finding is in sharp contrast to superficial acral fibromyxoma and dermatofibrosarcoma protuberans, which are usually diffusely and strongly positive for CD34. CD68-positive / CD34-negative immunophenotype, together with the presence of histopathologic features of classic dermatofibroma, is useful in the differential diagnosis.

Cellular digital fibroma is a benign tumor that usually develops in the fingers and toes. This tumor is superficially located and consists of monomorphic spindle cells arranged in a storiform pattern and immersed in a stroma with abundant collagen. Unlike myxoid dermatofibroma, cellular digital fibromas display a lower degree of vascularization in comparison with myxoid dermatofibroma, and the stroma is more fibrotic than myxoid [13,14]. Immunohistochemically, the spindle cells of cellular digital fibroma strongly express CD34, but are usually negative for CD99. Superficial angiomyxoma is most common in the head, neck and trunk, and is exceedingly rare in acral sites. They display multilobulated appearance and a prominent vascular component with a neutrophilic infiltrate scattered within the lesion. The tumor cells of superficial angiomyxoma also express CD34, and immunostaining for CD99 is characteristically negative. Cellular myxoma of soft tissue is a CD34-positive subcutaneous soft tissue tumor, developing most times within a muscle, rarely occurring on hands or feet [15]. Myxoid neurofibroma should be also considered in the histopathological differential diagnosis. This variant of neurofibroma reveals consistent immunoreactivity for S-100 and does not show the increased vasculature of myxoid dermatofibroma. Low-grade fibromyxoid sarcoma is a deep soft tissue tumor that only exceptionally arises on the fingers or toes. Although it can display alternating fibrous and myxoid areas, it is characterized by homogeneous whirling growth of spindle cells in heavily collagenized stroma and abrupt transition from fibrous to myxoid areas.

Dermatofibroma reportedly does not does not express CD99 [16,17], although in a recent study, diffuse and strong CD99 immunoreactivity was detected in all dermatofibromas examined [18]. Tumors of fibrohistiocytic origin have been reported to show CD99 positivity [18]. Role of CD99 immunostaining in the differential diagnosis of dermatofibroma remains controversial.

CONCLUSION

DFs can occur on any finger or even a toe, although it is rarely the preoperative diagnosis. DFs are more commonly found on the dorsal or lateral surfaces of the digits but may present on the palmar surface as well. A thorough examination for the presence of typical features of classic dermatofibroma and immunohistochemical staining are helpful in the differential diagnosis. DF should be included in the differential diagnosis of circumscribed lesions on digits to ensure proper histological diagnosis and treatment.

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