Multiple clustered myxoid dermatofibroma.

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ABSTRACT:

PRECHANOND P, WITITSUWANNAKUL J. MULTIPLE CLUSTERED MYXOID DERMATOFIBROMA.

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Myxoid dermatofibroma, a rare histological variant of dermatofibroma, is characterized by revealing mucinous material between the collagen bundles admixed with other typical findings of common dermatofibroma. Multiple clustered dermatofibromas are also uncommon clinical presentation, defined by multiple lesions localized on one anatomical region. We report an atypical case of multiple clustered dermatofibromas with myxoid stroma which is a very rare clinical and histologic variant of the common benign tumor. To the best of our knowledge this is the second report of multiple clustered myxoid dermatofibroma.

Key words: myxoid, dermatofibroma, clustered

บทคัดย่อ:

พัชรินทร์ ปรีชานนท์, เจตน์ วิทิตสุวรรณกุล รายงานผู้ป่วยโรค DERMATOFIBROMA ชนิดที่พบได้ไม่บ่อย อาการทางคลินิกพบรอยโรคจำนวนมากกระจายอยู่เฉพาะที่ ร่วมกับผลการตรวจทางพยาธิวิทยาพบ MUCIN ในชั้นหนังแท้ วารสารโรคผิวหนัง 2560; 33: 286-291.

สาขาวิชาตจวิทยา ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

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Myxoid dermatofibroma เป็นชนิดหนึ่งของ dermatofibroma ที่พบได้ไม่บ่อย เมื่อตรวจทางพยาธิวิทยาจะพบ mucin แทรกอยู่ระหว่างมัดของ collagen ร่วมกับการพบลักษณะอื่นๆที่จำเพาะต่อโรค dermatofibroma การพบ dermatofibromas จำนวนมากที่กระจายอยู่บนส่วนใดส่วนหนึ่งของร่างกายนั้น ก็เป็นอาการแสดงทางคลินิกที่พบได้ไม่บ่อยเช่นกัน

รายงานฉบับนี้ได้รายงานผู้ป่วยโรค dermatofibroma ชนิดที่พบได้ไม่บ่อยทั้งอาการแสดงและลักษณะทางพยาธิวิทยาในผู้ป่วยคนเดียว ที่ผ่านมามีเพียง 1 รายงานที่พบอาการแสดง และลักษณะทางพยาธิวิทยาดังกล่าว

คำสำคัญ: พยาธิวิทยา เนื้องอก เมือก ตุ่มเจ็บ ตุ่มที่เท้า

Case synopsis

A 29-year-old woman, who was previously healthy without underlying disease, presented with multiple painful nodules on the left foot that had appeared 6 months ago. She was treated ineffectively by 3 sessions of electrocautery. Multiple new painful nodules have progressively occurred. On examination, nineteen indurated skin colored to pink tender nodules are seen on the left sole, tips of third, fourth and fifth toes. (Figure 1) A skin biopsy was taken.

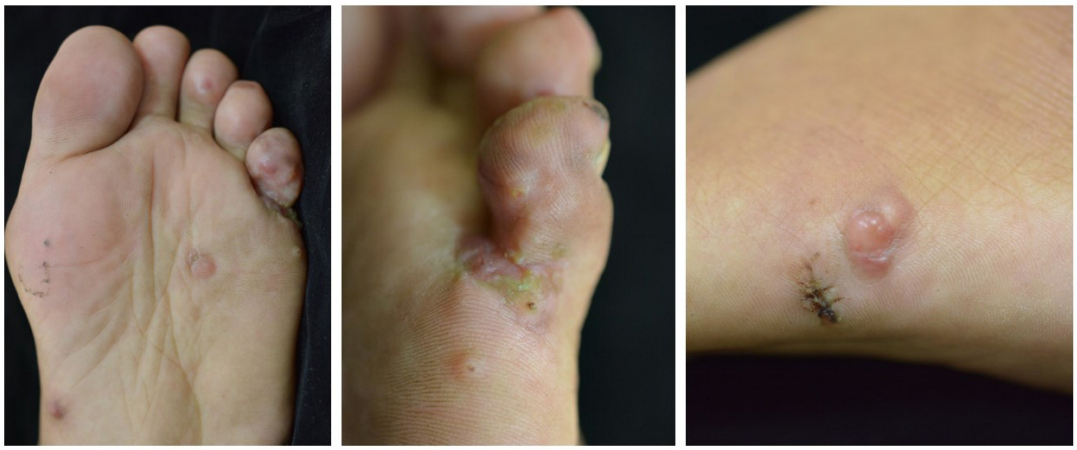

Figure 1 Nineteen indurated skin colored to pink tender nodules are seen on the left sole, tips of third, fourth and fifth toes.

The hematoxylin and eosin sections showed haphazard arrangement of fibrohistiocytic cells among coarse collagen bundles and myxoid stroma within the dermis. Few mast cells are seen. Collagen entrapment is noted at the periphery of lesions. There is overlying epidermal hyperplasia. Alcian blue stained sections are positive for mucin in the myxoid stroma. (Figure 2, 3)

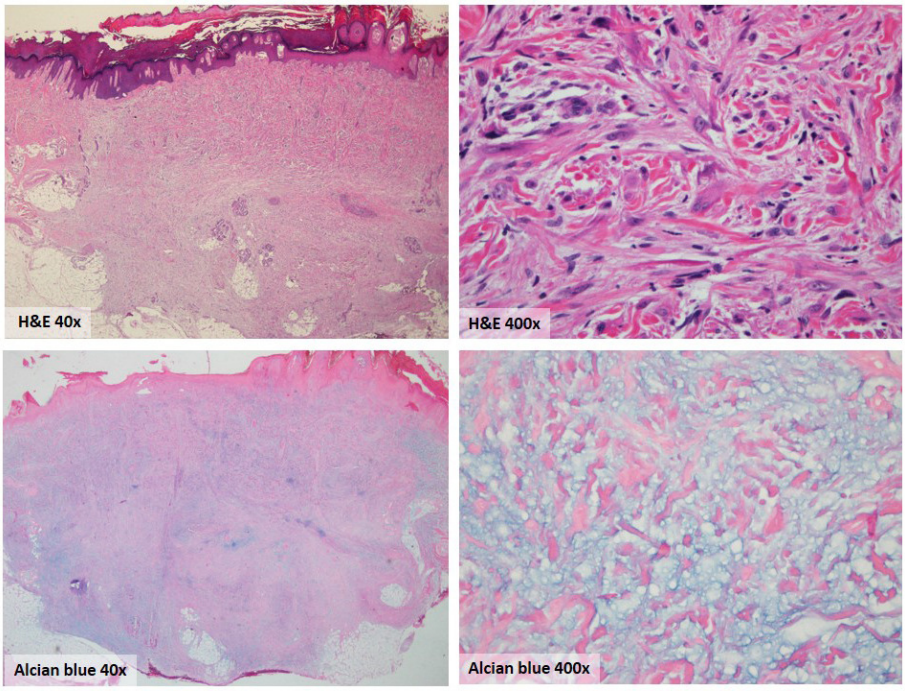

Figure 2 Histopathology showed haphazard arrangement of fibrohistiocytic cells among coarse collagen bundles and myxoid stroma within the dermis. There is overlying epidermal hyperplasia. Alcian blue stained sections are positive for mucin in the myxoid stroma. (H&E: hematoxylin and eosin)

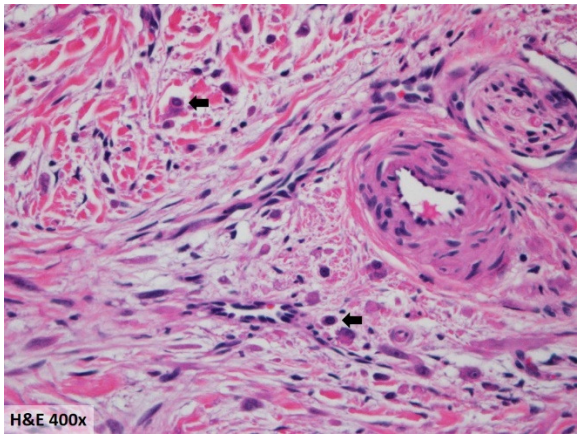

Figure 3 Few mast cells are seen within the dermis.

Immunohistochemical study demonstrated factor-XIIIa-reaction in more than 90% of dermal spindle and stellate shape cells. CD68 is also reactive in dendritic and histiocytic cells. CD34 reactive cells are exclusively confined to blood vessel endothelium. Smooth muscle actin are focally positive. S100 positive cells are greatly decreases in the lesion. (Figure 4)

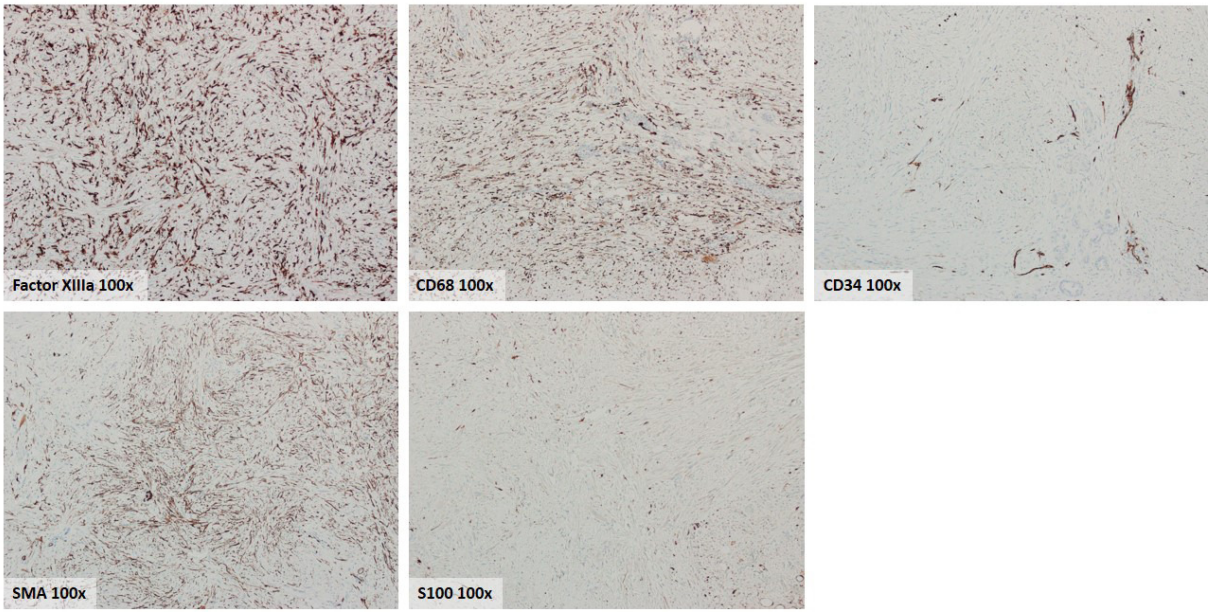

Figure 4 Immunohistochemistry showed positive reaction in Factor XIIIa and CD68 staining and focally positive reaction in SMA staining. CD34 reactive cells are exclusively confined to blood vessel endothelium. S100 positive cells are greatly decreases in the lesion. (SMA: smooth muscle actin)

Complete blood count, human immunodeficiency virus (HIV) antibody and antinuclear antibodies are normal. Five operable lesions were removed by surgical excision without recurrence after 7-month follow-up. Intralesional cryotherapy was used in treating lesions at the fourth and fifth toes without improvement.

Discussion

Dermatofibroma, so called fibrous histiocytoma, is a local cutaneous soft tissue response to minor trauma or inflammation. The solitary or occationally a few lesions are commonly seen in healthy middle-aged women. Lower extremities are usually involved.[1]

Histologic finding of common dermatofibroma shows dermal lesions consisting of interlacing fascicles of fibroblast-like spindle cells and histiocytes in the dermis, occasionally extending to superficial subcutaneous fat, separated from epidermis by grenz zone. Hyalinized collagen bundles, located at the periphery of the lesion, are entrapped by these spindle cells. Epidermal hyperplasia and basal hyperpigmentation are usually seen[1] Almost all cases of ordinary dermatofibroma express factor XIIIa and CD68, but not CD34 and S100.[2,3] However, the diagnosis should be made with clinicopathological correlation.[2]

Dermatofibromas can present with multiple clinical and histological variability. Histopathologic variants include cellular, aneurismal, hemosiderotic, epithelioid, atypical, lipidized, clear cell, palisading, atrophic, keloidal, granular cell, myxoid, lichenoid, balloon cell and signet-ring cell.[4] Myxoid dermatofibroma is an extremely rare variant, representing about 0.4% of all dermatofibromas.[5] This subtype of dermatofibroma is indicated by showing mucin deposition in stroma other than typical finding of classic dermatofibroma.[1] Postulated mechanisms for myxoid changes are mast cell-derived, inducing mucopolysaccharide production[6] and CD44 expression defect resulting in hyaluronate accumulation.[7] Immunohistochemically, reactivity for factor XIIIa are frequently absent, particularly in late lesions.[5] Though coexpression of factor XIIIa and CD34 occasionally seen.[8] CD68, a histiocytic marker, and smooth muscle actin staining are variable result.[1, 5, 8] There are some soft tissue tumors including superficial acral fibromyxoma, cellular digital fibroma, superficial angiomyxoma, myxoid dermatofibrosarcoma protuberans and myxoid neurofibroma, showing fibroblast –like cells in myxoid stroma that should be differentiated from myxoid dermatofibroma. All of these clearly show CD34 reactive cells, not the same as myxoid dermatofibroma, except the latter. Negative S100 staining can be used to differentiate myxoid dermatofibroma from myxoid neurofibroma which present the positive result.[9]

Multiple dermatofibromas, defined by more than 15 lesions, clinically manifested as eruptive, persistent, diffuse and clustered.[10] Multiple lesions are often associated with underlying diseases especially immunological disorders such as SLE, HIV infection, myasthenia grevis, pemphigus vulgaris, ulcerative colitis, Sjögren syndrome or immunosuppressive drug use. Other diseases such as lymphoproliferative disorders (chronic myeloid leukemia, myelodysplastic syndrome, Sézary syndrome, multiple IgA myeloma), metabolic diseases (diabetes mellitus, obesity, hyperlipidemia, hypertension), atopic dermatitis and pregnancy are reported previously[1]

Multiple clustered dermatofibromas, an atypical clinical variant, present with multiple dermatofibromas confined to one anatomical location. The predilection sites are the proximal legs in both sexes.[1, 10] Acral involvement is rarely seen.[11] Most cases of this distinct entity were not associated with any underlying diseases.[10] Only one case of multiple clustered myxoid dermatofibroma was reported previously.[8] In conclusion, this patient manifested atypical clinical and histological variant of dermatofibroma localized on atypical location. Clinical and histopathological data included

immunohistochemistry are necessary for definite diagnosis.

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