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CASE AND RESEARCH LETTERS

Acral Nodular Lesion Following Trauma☆

Lesión nodular acral tras traumatismo

To the Editor:

We present the case of a 37-year-old man with no personal history of interest who came to our department with a lesion on the dorsum of the distal phalanx of the third finger of his right hand. The lesion had first appeared after an injury a few months earlier. It had deformed the nail and was painful on palpation.

Physical examination revealed a solid subungual tumor that deformed the nail in such a way that the curvature was increased both longitudinally and transversally (Fig. 1).

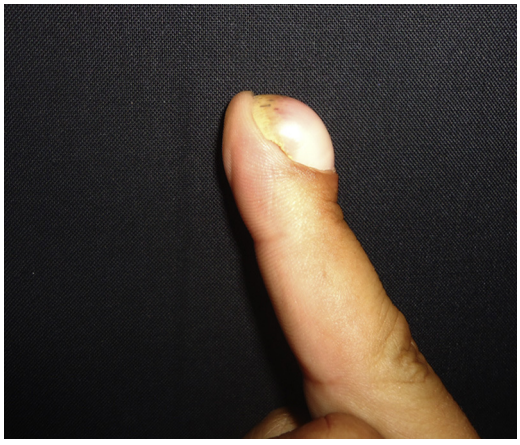

Figure 1 Longitudinal and transverse deformity of the nail plate on the third finger of the right hand.

A punch biopsy of the lesion was performed after avulsion of the nail plate. Staining with hematoxylin-eosin revealed a well-defined nodular tumor in the dermis that spared the Grenz zone. Greater magnification revealed that the tumor was composed of a very dense population of spindle-shaped cells that were stellate in appearance with an eosinophilic cytoplasm and surrounded by a myxoid stroma with abundant vessels and isolated mastocytes. The vessels were small and had fine walls lined by endothelial cells with no atypia. Immunohistochemical staining for CD34, S100, and epithelial membrane antigen was only positive for CD34 (Fig. 2). The correlation between clinical and histology findings confirmed the diagnosis of superficial acral fibromyxoma.

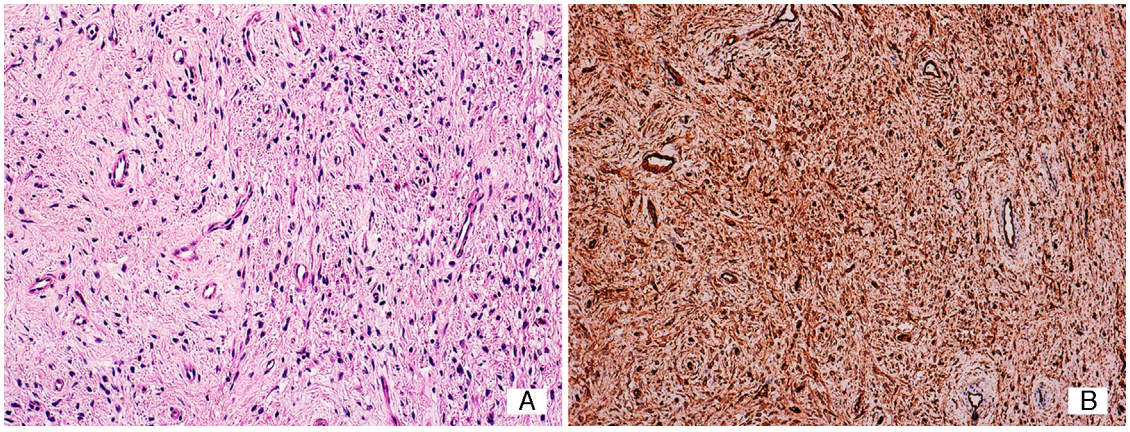

Figure 2 A, Spindle-shaped cells immersed in a myxoid stroma. Small, fine-walled vessels together with occasional mastocytes (hematoxylin-eosin × 200). B, CD34 immunoreactivity in neoplastic cells (× 200).

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A plain radiograph taken before surgery revealed a nodule attached to the soft tissue, with no bone involvement (Fig. 3).

Once the diagnosis was confirmed, the lesion was removed completely. First, the area was anesthetized using digital block with mepivacaine. Second, after application of local ischemia at the base of the finger, a small transverse incision was made in the eponychium in order to better visualize the extent of the lesion. Third, the lesion was dissected carefully and removed. Finally, curettage was performed on the nail bed and the eponychium was sutured. The patient continues to be followed in our department. No recurrence of the tumor has been observed, and the cosmetic outcome is good.

Superficial acral fibromyxoma is a benign tumor that was first described by Fetsch et al. in 2001.[1] It is more common on the fingers and toes of middle-aged men. Clinically, it presents as a solid, well-defined, solitary nodule. The nail is involved in as many as 50% of cases.[2–4] Diagnosis is usually delayed because of the slow growth of this type of lesion.

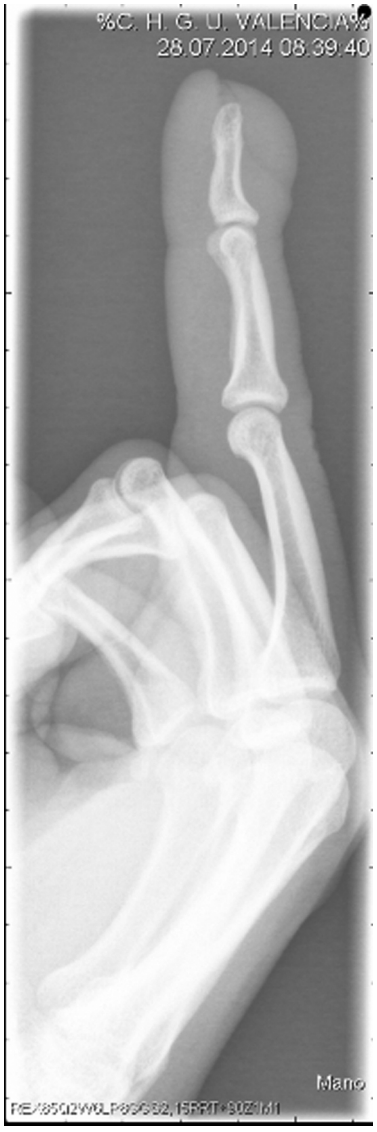

Figure 3 Nodule in contact with soft tissue. No bone involvement is observed on the radiograph.

Table 1 Differential Diagnosis of Superficial Acral Fibromyxoma.

Disease	Clinical Findings	Histology
Superficial acral fibromyxoma	Solitary nodule in the distal area of the extremities	Spindle-shaped cells immersed in a myxoid stroma Positive for CD34, CD99, and EMA Negative for S-100
Cellular digital fibroma	Small nodule on the fingers and toes	More cellular tumor Dense collagen in the dermis Positive for CD34 and factor XIIIa Negative for EMA and S-100
Dermatofibrosarcoma protuberans	Variable appearance depending on time since onset. Acral presentation is extremely rare	Spindle-shaped cells with a storiform pattern Positive for CD34, actin, and vimentin Negative for S100, HMB-45, and factor XIIIa
Superficial angiomyxoma	Flesh-colored papule or nodule found mainly on the trunk and lower extremities	Multilobulated, poorly defined tumor formed by spindle-shaped cells with a myxoid stroma. Inflammatory infiltrate with neutrophils Positive for actin and vimentin Negative for S100 and factor XIIIa
Benign fibrous histiocytoma	Brown nodule or papule, pink on lower limbs	Poorly defined tumor. Bundles of spindle-shaped cells in a myxoid stroma. Collections of hyalinized collagen in the dermis Positive for factor XIIIa Negative for CD34
Onychomatricoma	Longitudinal and transverse thickening of the nail plate. Splinter hemorrhages	Nail plate with fibrovascular projections covered with keratinizing epithelium in the matrix, together with empty cavities

Abbreviation: EMA, epithelial membrane antigen; HMB, human melanoma black.

Histopathology reveals a well-defined, nonencapsulated tumor affecting the dermis and, sometimes, the subcutaneous cellular tissue. In most lesions, a Grenz zone is visible with no involvement of the epidermis. The tumor is composed of spindle-shaped cell bundles with pale eosinophilic cytoplasm and basophilic nuclei. These cells are immersed in areas of dense hyaline collagen and other areas of myxoid stroma. In most cases, we can see mastocytes and small vessels. Cellular necrosis and lymphovascular invasion are not usually present.[1–8] Immunohistochemical staining is positive for CD34, CD99, and epithelial membrane antigen and negative for S-100, muscle-specific actin, and glial fibrillary acidic protein.[2–7]

Histologically, it is important to differentiate this tumor from cellular digital fibroma, a CD34-positive tumor composed of spindle-shaped cells with a less myxoid stroma than in acral fibromyxoma that does not show immunoreactivity for epithelial membrane antigen or CD99. The differential diagnosis also includes myxoid neurofibroma, superficial angiomyxoma, fibrous histiocytoma, dermatofibrosarcoma protuberans, and onychomatricoma, owing to the histological characteristics shared by these tumors (Table 1).

Treatment is based on surgical removal, although the tumor can recur in as many as 20% of cases if the excision is not complete.[2]

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Mixed Connective Tissue Disease in a Patient With Castleman Disease and Hodgkin Lymphoma: Excellent Clinical Response to Rituximab[☆]

Enfermedad mixta del tejido conectivo en el contexto de enfermedad de Castleman y linfoma Hodgkin con excelente respuesta clínica a rituximab

To the Editor:

Castleman disease (CD), or angiofollicular lymphoid hyperplasia, is characterized by a process of reactive hyperplasia affecting the immune system. The condition can, therefore, give rise to a clinical picture similar to that of either a connective tissue disease or an autoimmune disease.[1] We report a case of Castleman disease and Hodgkin lymphoma in a patient who developed a mixed connective tissue disease (MCTD).

Case Description

The patient was a 49-year-old man, who had recently been diagnosed with Hodgkin lymphoma and multicentric Castleman disease and was being treated with adriamycin, bleomycin, vinblastine, and dacarbazine. He was referred to our department when he developed skin lesions and photosensitivity in association with Raynaud's phenomenon and muscle weakness a month after completing chemotherapy.

Physical examination revealed the following: erythematous-violaceous coloration on sun-exposed areas, elbows, and knees; edema of the palms and soles; and erythematous scaly papules on the dorsal surfaces of the interphalangeal and metacarpophalangeal joints of the hands consistent with Gottron papules (Fig. 1). Capillaroscopy demonstrated disorganization of the capillary bed, loss of normal capillary distribution, and a few thrombosed giant capillaries.

Two of the explorations performed yielded significant results: an electroneurogram evidenced sensory-motor polyneuropathy; and autoimmune tests revealed antinuclear antibodies (titers of 1:1280), anti-Smith antibodies, and

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