Casos Clínicos

Nodular fasciitis: a benign mimicker of soft tissue malignancies

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Resumen

La fascitis nodular es una masa subcutánea, benigna, autolimitada, que puede simular un sarcoma de tejidos blandos en su presentación clínica e histopatológica. Debido a que la mayoría de las descripciones de esta condición provienen de pacientes caucásicos, es necesario enfatizar su existencia en poblaciones poco representadas en la literatura. Se presenta el caso de un paciente mexicano, masculino, de 49 años, quien fue inicialmente mal diagnosticado y tratado como quiste epidérmico. Tras la recurrencia de la lesión, se realizó una adecuada técnica de biopsia y estudios de imagen, confirmando el diagnóstico de fascitis nodular. El presente reporte resalta la importancia de abordar con cautela las masas subcutáneas que puedan simular tanto tumores benignos como malignos para mejorar el pronóstico de los pacientes.

Palabras claves: Fascitis nodular, sarcoma, biopsia, tumor, subcutáneo

Abstract

Nodular fasciitis is a benign, self-limited, subcutaneous tumor that may resemble a soft tissue sarcoma both clinically and histopathologically. Since most descriptions derive from Caucasian patients, further awareness is necessary in underrepresented populations. Here, we present a case of nodular fasciitis in a 49-year-old Mexican male who had been misdiagnosed and mistreated as an epidermal cyst. After recurrence, an adequate biopsy and imaging studies confirmed the diagnosis of nodular fasciitis. This case illustrates the importance of properly studying subcutaneous masses that can mimic a wide array of benign and malignant tumors to improve overall patient prognosis.

Key words: Nodular fasciitis, diagnostic errors, sarcoma, rare diseases, subcutaneous

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Nodular fasciitis (NF) is a benign, fast-growing, pseudosarcomatous, myofibroblastic tumor that is clinically undistinguishable from true sarcomatous lesions.[1] Although NF tends to self-resolve, its similarity with other malignancies demands a careful workup and treatment strategy. The diagnosis is confirmed with a set of immunohistochemistry markers on histopathology. Excision surgery is considered the first line of treatment,[2] albeit other less invasive alternatives have been proposed.[3] Since most clinical, histopathological, and genomic descriptions arise from Caucasian patients, awareness of this relatively rare entity is warranted in other populations.

Clinical Case

An otherwise healthy 49-year-old Mexican male presented with a 7-month history of an enlarging, tender lesion on his face. On physical examination, there was a 2.5x3x1.7 cm subcutaneous, firm, sharply defined tumor, covered by healthy-appearing skin and a linear scar caused by a failed excision attempt of a suspected epidermal cyst 3 months prior (Figure 1). The lesion ultrasound showed a thick-walled mass with mixed echogenicity (Figure 2A) and no bone damage was identified on computed tomography (Figure 2B). The histopathology demonstrated a dense proliferation of spindle-shaped cells admixed within a fibromyxoid

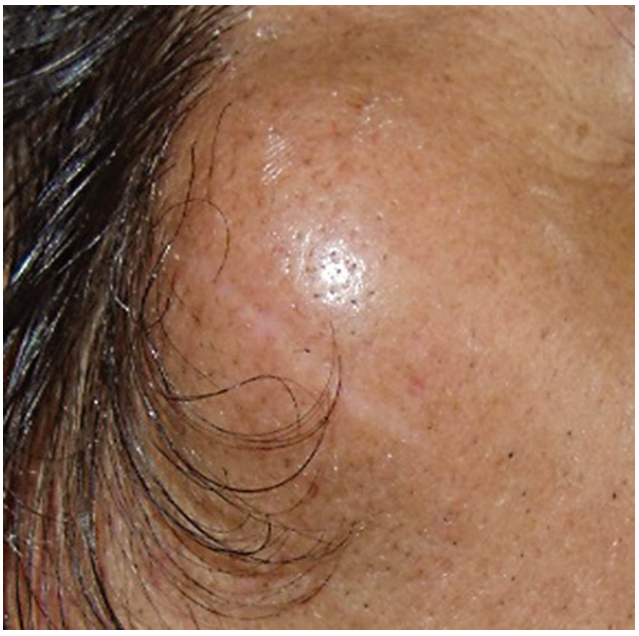

Figure 1
Clinical appearance.

stroma extending to the adipose tissue (Figure 3A). Immunohistochemistry was positive for vimentin (Figure 3B) and negative for CD34, desmin, and S-100. Therefore, the diagnosis of nodular fasciitis was established. The lesion was fully excised and has not recurred after 3 years of follow up.

Discussion

NF was first described by Konwaler et al. in 1955 in a series of 8 cases.[4] Since then, there have been various studies aimed at characterizing this entity at the clinical, genomic, and histopathological level, mostly in Caucasian patients.

An analysis of 250 patients from Japan showed a male to female ratio of 1.03, a mean age of 39 years, and the upper extremity as the most frequently involved site (44%). No face lesions were reported.[1]

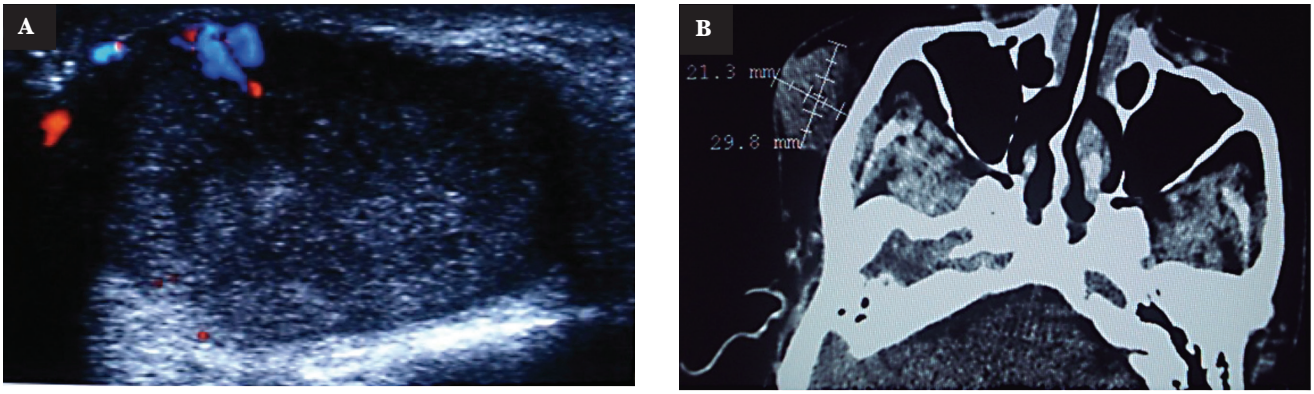

Figure 2
The mass showed mixed echogenicity on ultrasonography (a) no damage to cortical bone structures (b).

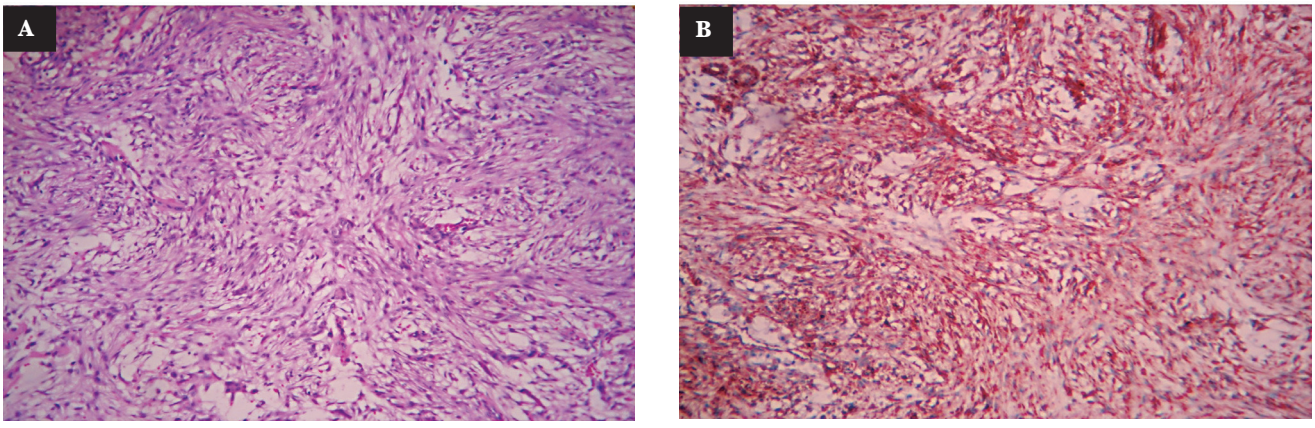

Figure 3
The histopathology of the lesion demonstrated a dense proliferation of spindle-shaped cells admixed within a fibromyxoid stroma (a) and positive vimentin staining on immunohistochemistry (b).

Nonetheless, a report from South Korea exposed 16 cases of NF in the face area with a similar mean age and sex distribution.[3] In the pediatric population, a North American series documented 15 cases of NF of the head and neck regions, with a mean age of 9.3 years.[5]

At the genomic level, NF tissues have shown to harbor a recurrent translocation t(17;22) which conditions the MYH9-USP6 gene fusion, similar to rapidly-growing, benign, aneurismal bone cysts.[6] Given the spontaneous regression of NF, further studies are required to fully understand the transient nature of this mutation.

The diagnosis is established by histopathology and immunohistochemistry (IHC). The accuracy of fine-needle (FNA) aspiration is low. In a study of 33 cases of NF confirmed on excision histopathology, only 11.8% were initially diagnosed as NF by FNA.[7] On histopathology, NF appears as a dense sheet of spindle-shaped cells within a myxoid, cellular, fibrous, or mixed stroma.[1,2] Although mitoses are abundant, atypia is rare. Muscle-specific actin, smooth-muscle actin, and vimentin are usually positive on IHC, whilst S-100, desmin, trypsin, factor VIII, macrophage-specific antigen, HLA-DR1, and CD34 are negative. The main differential diagnoses include desmin-positive leiomyoma and leiomyosarcoma, CD34-positive dermatofibrosarcoma, S-100-positive spindle cell melanoma, and other sarcomas.[8]

Imaging investigations are routinely performed to examine depth and invasion to adjacent structures. Sonographic findings often show mixed echogenicity.[9] Computed tomography (CT) and magnetic resonance (RM) scans may display homogeneous hypodensity and isointensity on T1-weighted images relative to the muscle, respectively.[10] Therefore, most lesions appear superficial and resemble benign cysts on imaging studies.

Treatment of NF often involves surgical excision and shows low recurrence rates (<10%).[2] Although, the use intralesional triamcinolone (ILT) and carbon dioxide laser (CO2) have also demonstrated favorable outcomes in a small case series,[3] the cost and time to resolution must be carefully weighed against any potential cosmetic benefit.

In conclusion, the unremarkable clinical presentation of NF demands a thorough workup, mainly due to its similarity with potentially lethal malignancies. In our case, we chose to omit FNA, ILT and CO2 laser interventions to reduce the cost of care. Yet, appropriate histopathology, immunohistochemistry, imaging studies, complete surgical excision, and follow-up evaluations were performed for a comprehensive approach in a resource-limited hospital setting. Further descriptions and genomic investigations of NF in Latin-American patients are required.

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