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CASE REPORT

Facial nodular fasciitis in an adult patient: a case report of an uncommon manifestation

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Abstract:

Nodular fasciitis is a pseudosarcomatous, self-limited lesion composed of fibroblasts and myofibroblasts. Craniofacial lesions are more common in pediatric patients. This report presents a rare case of nodular fasciitis involving the face of an adult patient. A 32-year-old woman presented with a painless subcutaneous mass at the right zygomatic region, with one year of duration. The diagnostic hypotheses were epidermoid cyst and neurofibroma. An excisional biopsy was performed, and the microscopic examination exhibited an admixture of spindle-shaped cells and acellular areas associated with prominent collagen bands. No cellular atypia was observed. Immunohistochemical findings demonstrated positivity for alpha-SMA, HHF35, and beta-catenin. Tumor cells were negative for STAT6. The final diagnosis was nodular fasciitis. The patient has not presented any recurrences so far. Clinicians and pathologists must be capable to distinct nodular fasciitis and malignant tumors to avoid misdiagnosis and overtreatment.

Keywords: Nodular fasciitis; Fibroblast; Myofibroblast; Face.

INTRODUCTION

Nodular fasciitis was first described as "subcutaneous pseudosarcomatous fibromatosis" and is a benign neoplasm composed of fibroblasts and myofibroblasts within a variably collagenous stroma[1-3]. Despite its benign nature, this lesion can mimic sarcomas due to its clinical and histological spectrum, including rapid infiltrative growth, high cellularity, and a prominent mitotic rate[4,5].

The etiology of nodular fasciitis remains uncertain, though trauma or infection has been described as a potential factor[6]. The lesion commonly arises from the muscular fascia and extends into the subcutaneous tissues, although it it may also occur in superficial locations[3,6]. Adults are more affected than children. Common anatomical sites for nodular fasciitis in adults include the upper extremities, trunk, and chest; only 7–20% of the cases occur in the head and neck[7]. On the other hand, in pediatric patients, the head and neck region is the most common site, including the maxillofacial area, which is also known as craniofacial fasciitis[4,7,8].

The diagnosis of nodular fasciitis can be challenging, particularly when it is found in atypical locations or with unusual clinical features. Thus, we report a case of nodular fasciitis on the right zygomatic region bone of an adult patient notable for its slow growth and provide a literature review on this condition.

Statement of Clinical Significance
Craniofacial involvement of nodular fasciitis commonly occurs in infants and children. Cases involving the facial region of adults are frequently diagnosed as benign and malignant conditions of neural, fibroblastic, and myofibroblastic origin. Considering the self-limited evolution of nodular fasciitis in most cases, an appropriate diagnosis of uncommon presentations is essential to avoid misdiagnosis of a malignant neoplasms an to avoid improper treatment.

CASE REPORT

A 32-year-old woman presented to a maxillofacial surgery service with a 1-year history of an asymptomatic facial swelling. The patient's medical history was unremarkable. Extraoral examination revealed a well-circumscribed, round, subcutaneous nodule in the right zygomatic region, causing noticeable facial asymmetry (Figure 1A). There were no inflammatory signs. The main diagnostic hypotheses were neurofibroma and epidermoid cyst.

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Under local anesthesia, an excisional biopsy was performed. Gross examination showed a well-demarcated but unencapsulated mass with a smooth surface, fibrous consistency, and yellowish cut surface (Figure 1B). Microscopic examination showed a proliferation of uniform spindle cells with vesicular nuclei and prominent nucleoli in a tissue culture-like architecture growth pattern. Multiple thin-walled vascular spaces were also noted. The cells lacked nuclear atypia, and no atypical mitoses were observed (Figures 2A–B). Prominent collagen bands were observed alongside neoplastic cells and extravasated red blood cells (Figure 2C).

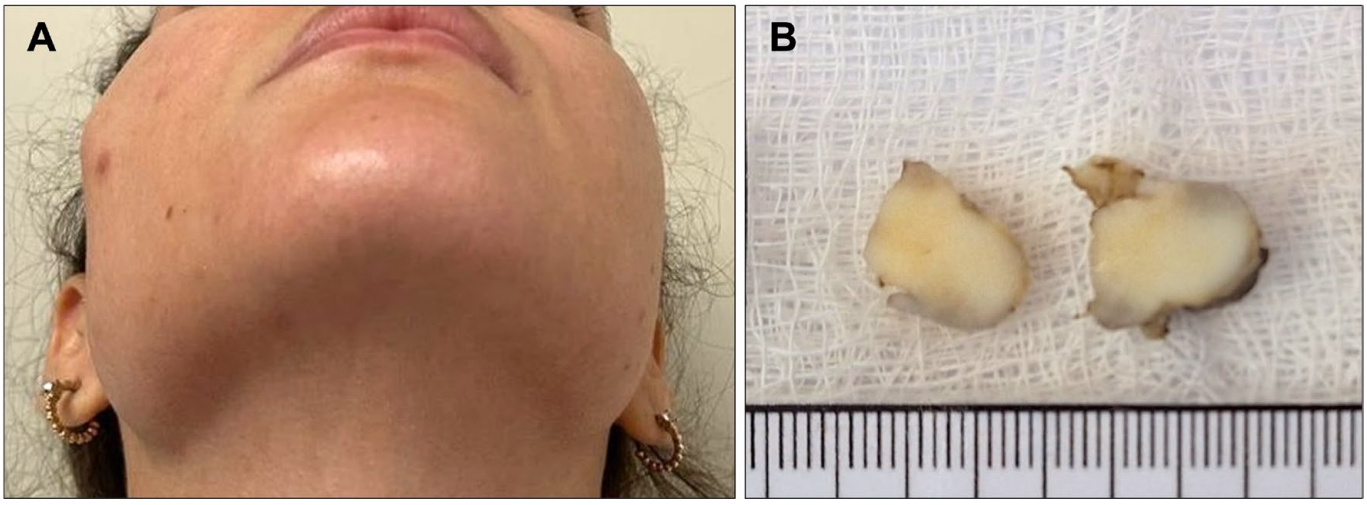

Figure 1. Clinical and macroscopic features of facial Nodular fasciitis. **A)** A round-shaped, nodular lesion in the right malar region. **B)** A unencapsulated soft-tissue fragment with fibrous consistency and yellowish coloration, measuring 1,2 x 1,5 x 0,9 cm.

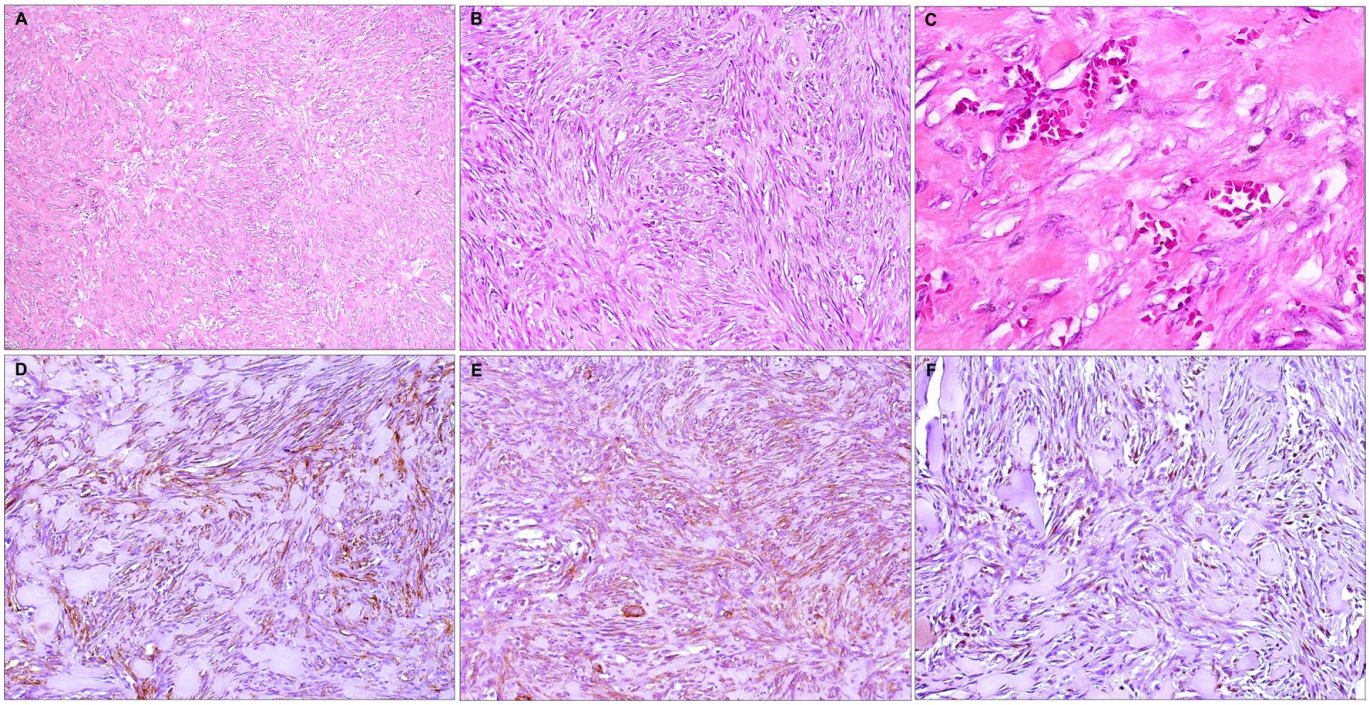

Figure 2. Microscopic and immunohistochemical findings of Nodular fasciitis. **(A)** Diffuse proliferation of spindle-shaped cells lacking atypia in a tissue culture–like architecture (H&E, 200x). **(B)** Multiple vascular spaces alongside neoplastic cells and collagen bands (H&E, 200x). **(C)** Higher power view of the tumor showing hyaline fibrosis with vascular spaces and extravasated red-blood cells (H&E, 400x). **(D)** Positive staining for α-SMA, confirming the myfibroblastic origin of the neoplastic cells (DAB; 200x). **(E)** Diffuse expression of HHF-35 was also observed (DAB, 200x). **(F)** Nuclear and focal positivity for β-catenin (DAB; 200x).

Table 1. Clinicopathological features of facial NF in adults published in the literature.

Clinicopathological variables	n=48	%
Sex		
Female	11	22.9
Male	10	20.8
NA	27	56.2
Age (mean age: 33.7 years)		
<33.7	9	18.8
>33.7	12	25
NA	27	56.2
Site		
Mandibular region	20	41.6
Cheek	10	20.8
Zygomatic region	8	16.6
Face, NOS	6	12.5
Others*	4	8.3
Duration (mean: 3 months)		
<3	7	14.6
>3	8	16.6
NA	33	68.8
Symptoms		
Pain	2	4.1
Asymptomatic	9	18.8
NA	37	77
Immunohistochemistry		
Yes	10	20.8
No	38	79.1
Molecular analysis		
Yes	1	2.1
No	47	97.9
Treatment		
Excision	40	83.3
Incisional biopsy + wait-and-see	2	4.2
NA	6	12.5
Follow-up time (mean: 52 months)		
<52	9	18.8
>52	3	6.2
NA	36	75
Recurrence		
Yes	2	4.2
No	12	25
NA	34	70.8

NA: not available; NOS: not otherwise specified.
*Lip (2), Forehead (1), Left masseter muscle region (1).

Immunohistochemical investigation was performed in 3 μm sections of formalin-fixed, paraffin-embedded sections. DAB chromogen (Sigma-Aldrich, St Louis, MO, USA) with Carazzi hematoxylin counterstain were used. The cells showed diffuse positivity for α-SMA (Figure 2D), and HHF-35 (Figure 2E). There was a focal expression for β-catenin (Figure 2F). Negative markers included desmin, CD34, bcl-2, s-100, and STAT6. These findings confirmed the diagnosis of nodular fasciitis. The patient achieved complete resolution after excision, with no evidence of recurrence after one year.

DISCUSSION

Herein we described a case of nodular fasciitis occurring in the facial region of an adult patient. Our literature review identified 48 reports of facial nodular fasciitis (Table 1). Men and women were equally affected, with a peak incidence in the third and fourth decades of life, in accordance with the literature[4,8,9]. Anatomically, the mandibular region was most frequently affected (20 cases, 41.6%), followed by the cheek (10 cases, 20.8%). In contrast with our case, nodular fasciitis demonstrates a rapid onset, with duration of 2–3 months[8]. Notably, only one previous case involving the orofacial region demonstrated similarly prolonged evolution[10]. Nodular fasciitis typically presents as a solitary mass measuring ≤3 cm, which causes tissue distortion, and may be tender on palpation[7,8,10]. Clinical differential diagnoses encompasses both benign and malignant neoplasms, including lipoma, fibromatosis, dermatofibroma, neurofibroma, fibrosarcoma, and fibrous histiocytoma variants[9,11]. In this case, the lesion's well-circumscribed subcutaneous location also raised consideration of epidermoid cyst.

Microscopic features of nodular fasciitis exhibit myofibroblasts and plump fibroblasts with oval vesicular nuclei and prominent nucleoli arranged in irregular fascicles within a tissue culture-like architecture[4,6]. Cellular areas show a storiform growth pattern associated with a network of capillaries and extravasated, resembling granulation tissue[8,11]. Notably absent are nuclear atypia, tumor necrosis, or atypical mitotic figures, though normal mitoses may be present[8]. The tumor stroma may be myxoid, fibrous, and occasionally present keloidal collagen, as presented in our case[4,6,8]. Depending on the plane of the tissue affected, nodular fasciitis can be classified as subcutaneous, which is usually found in the head and neck region, intramuscular, and fascial[7]. Nodular fasciitis remains a diagnostic challenge due to its histological overlap with several spindle cell neoplasms

including fibrosarcoma, leiomyosarcoma, neurofibroma, and dermatofibrosarcoma protuberans, despite lacking definitive malignant features[9,10].

Although immunohistochemical reactions have been little used in cases of facial nodular fasciitis (Table 1), its application remains crucial for definitive diagnosis given the extensive differential diagnoses. The possibilites include tumors of neural origin — schwannoma, traumatic neuroma; myogenic tumors — leiomyoma and leiomyosarcoma; fibroblastic and myofibroblastic tumors — desmoplastic fibroma, fibromatosis, solitary fibrous tumor, myofibroma, and myofibrosarcoma[6,12]. The characteristic immunohistochemical profile demonstrates consistent positivity for vimentin, α-SMA, and HHF-35, while being negative for S100, CD34, and cytokeratins. Focal desmin reactivity may occasionally occur, whereas STAT6 expression is uniformly absent, as confirmed in our case[8-10]. To rule out the diagnosis of myofibroma, we relied on the expression of α-SMA, HHF-35, and the tissue culture-like architecture of nodular fasciitis, which is absent in myofibroma, presenting a biphasic pattern and the zoning phenomenon[8,9,12]. The latest WHO classification of head and neck tumors states that the expression of β-catenin plays a role in the differential diagnosis of nodular fasciitis; it helps the exclusion of fibromatosis and desmoid tumor, since they exhibit strong nuclear expression[8,13]. Moreover, β-catenin is typically negative in other spindle cell proliferations, like fibrosarcoma and myofibroma[13].

Molecular studies have been useful to comprehend the pathogenesis of nodular fasciitis. The Ubiquitin Specific Peptidase 6 (USP6) is a gene that encodes a deubiquitinating enzyme involved in several cellular processes, such as protein turnover, inflammatory signaling, and cell transformation[2]. USP6 rearrangements have been identified in up to 92% of nodular fasciitis cases, with MYH9::USP6 representing the most frequent fusion[12,14,15]. Additional fusion partners (SERPINH1, PPP6R3, COL3A1, and COL6A2) have been reported in both conventional and cranial fasciitis variants[4,8]. Notably, identical USP6 rearrangements occur in aneurysmal bone cyst, fasciitis ossificans, myositis ossificans, and the cellular variant of fibroma of tendon sheath, suggesting a shared pathogenic mechanism[6,12]. These mutations have indicated that nodular fasciitis represents a temporary, self-resolving neoplasm[4,8]. These molecular findings support the neoplastic yet self-limiting nature of nodular fasciitis. The transient biological behavior may reflect the temporally restricted activity of these genetic alterations.

Surgical excision is the preferred method for treatment of nodular fasciitis, while most cases present spontaneous resolution or regress by scarring after partial excision. In contrast to the trunk and extremities, recurrence is mostly reported on the face[11,15]. Due to aesthetic purposes and recurrences, non-surgical treatments and careful follow-up may be considered for cases involving the face and include triamcinolone intralesional injection and carbon dioxide laser[11,16].

CONCLUSION

Facial nodular fasciitis in adults represents an uncommon benign fibrous proliferation, which requires careful differentiation from malignant soft tissue tumors, providing an accurate diagnosis to prevent overtreatment. In most cases, molecular characterization of *USP6* rearrangements may serve as a valuable ancillary tool for challenging cases, particularly those with atypical clinical or morphological features.

AUTHORS' CONTRIBUTIONS

CIRF: conceptualization, investigation, writing – original draft. **RKBS:** data curation, investigation. **HKFB:** investigation, visualization. **EJAC:** writing – review & editing. **PAV:** investigation. **DECP:** conceptualization, investigation, writing – review & editing.

CONFLICT OF INTEREST STATEMENT

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