

Case Report

Atypical location of nodular fasciitis in a pediatric patient: a case report and literature review

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ABSTRACT

Nodular fasciitis (NF) is a benign, rapidly growing soft tissue lesion that can mimic malignant neoplasms. First described in 1955, it is commonly seen in adults but remains rare in pediatric populations, particularly in the orbital region. A comprehensive literature review was conducted, analyzing all reported cases of orbital NF between 1990 and 2022. Key parameters included patient demographics, clinical presentation, histopathological findings, treatment approaches, and outcomes. Twenty cases were identified, with an equal distribution of male and female patients. The most common clinical presentation was a rapidly growing, painless orbital mass. Histopathological analysis revealed a spindle cell proliferation with myxoid stroma, positive immunostaining for actin and vimentin, and variable expression of other markers. Surgical excision was the primary treatment, with no reported recurrences or postoperative complications. Although rare, orbital NF should be considered in the differential diagnosis of pediatric soft tissue tumors. Awareness of its benign nature is crucial to avoid unnecessary aggressive treatments. Surgical excision remains the treatment of choice, although spontaneous regression has been reported. Further studies are needed to elucidate its etiology and optimize management strategies.

Keywords: Nodular fasciitis, Pseudosarcomatous fibromatosis, Myofibroblastic proliferation, Epidermoid cystic, Children

INTRODUCTION

Nodular fasciitis (NF) is a benign myofibroblastic proliferation characterized mainly by rapid growth and usually affects subcutaneous tissue, muscle and fascia. Although traditionally considered a reactive lesion rather than a true neoplasm, its clinical and pathological presentation can lead to diagnostic challenges.¹⁻⁴ We present the case of a 10-year-old male patient with a dermatosis localized to the inner canthus of his left eye (Figure 1 A, B). Histopathological and

immunohistochemical analyses confirmed the diagnosis, highlighting the importance of precise clinical and diagnostic approaches for such lesions, particularly in the pediatric population, where limited literature exists.

The need for appropriate management is also discussed, as most cases are self-limiting and may respond to various treatment modalities. A literature review was also conducted regarding reported cases of nodular fasciitis in the orbit.

CASE REPORT

A 10-year-old male with no significant medical history presented to the dermatology clinic with a firm, well-demarcated subcutaneous nodule with an erythematous surface, which was located to the head, on the left inner canthus of the eye, with a two-month evolution. The patient mentioned he had moderate pruritus as well. (Figure 1 A, B). Surgical excision was performed, and the histopathological study revealed a non-encapsulated, poorly defined neoplasm composed of a monomorphic proliferation of spindle cells without atypia, interspersed with vacuolated cells and collagen bundles arranged in various directions. Immunohistochemistry studies showed cytoplasmic positivity for smooth muscle actin within the spindle cells. CD34 was positive only in the blood vessel walls (Figure 2 D). The histological morphology and immunostaining pattern were consistent with NF.

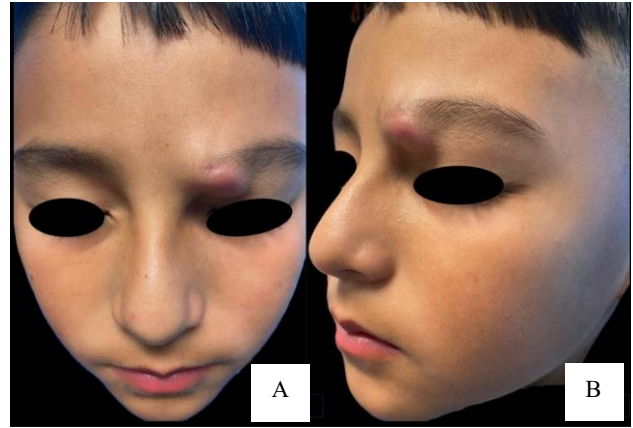


Figure 1: Erythematous-surfaced neof ormation with well-defined margins affecting the inner canthus, displaying regular borders: (A) frontal view and (B) lateral view.

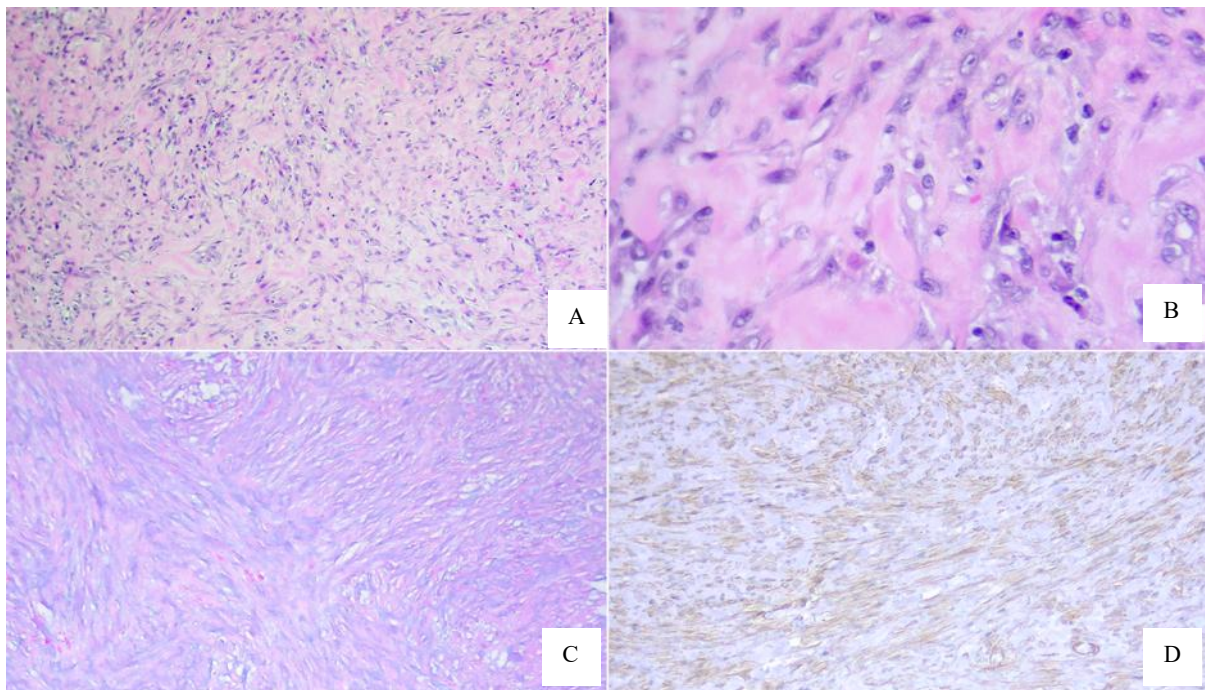


Figure 2: (A) histopathological study showing a spindle cell neoplasm arranged in short bundles and fascicles within a myxoid stroma. He, 10x, (B) higher magnification of figure 2A revealing spindle-shaped to plump fibroblasts and myofibroblasts with ovoid, monomorphic nuclei. He, 40x, (C) immunohistochemical analysis showing positive staining of the mucinous stroma with alcian blue and (D) cells positive for smooth muscle actin.

DISCUSSION

NF was first described in 1955 by Konwaler et al as pseudosarcomatous fibromatosis. In 1961, Stout coined the term "nodular fasciitis", suggesting that the lesions originate in superficial and deep fascial layers. It tends to be more common in adults aged 20-40 years, with approximately 10% of cases occurring in children.¹⁻⁴ However, data on pediatric NF are scarce. Bemrich-Stolz et al reported 18 cases in pediatric patients over 12 years, seven localized to the head and neck.⁵ In contrast Pandian et al conducted a larger review of 26 pediatric

cases in 2013 without reporting lesions on the head or neck, Font et al described 10 cases in the orbit and adnexa in 1966, with pediatric patients accounting for 12-17% of all NF cases.⁶⁻⁸ In 2001, Shields et al. reported an 8-month-old infant with a rapidly growing, firm, painless subcutaneous mass in the right upper eyelid, causing visual impairment.⁹ Surgical excision through a superolateral orbitotomy revealed a spindle cell proliferation interspersed with thick connective tissue trabeculae, multinucleated giant cells and moderate immunoreactivity for vimentin and smooth muscle actin.⁹ No recurrence was observed after 6 months.⁹

Table 1: Reported cases of nodular fasciitis located on the orbit.

Author	Year	Year (months)	Sex	Past medical history	Topography	Clinical presentation	Imaging studies	Clinical speculation	Histopathological study	Evolution	Complications
Holds¹⁶	1990	36	F	None	Supranasal to the eyeball	Rapidly growing neoplasm (5 weeks)	None	Rhabdo myosarcoma	Nodular fasciitis	No recurrence (9 months follow-up)	None
Vestal¹⁷	1990	144	M	None	Right upper eyelid	Rapidly growing red neoplasm (4 weeks)	None	Epidermoid cyst	Non-encapsulated mass composed of interwoven bundles of spindle cells. Cellular areas with focal myxoid regions.	No recurrence	None
Sakamoto¹⁸	1991	84	F	None	Left epicanthus	Painful, rapidly growing neoplasm (8 weeks)	MRI: solitary nodular mass in the anteromedial part of the left orbit.	Tumor	Proliferation of robust spindle cells disorganized within a mucous matrix, mixed with numerous multinucleated giant cells.	No recurrence	None
Kaw¹⁹	1993	120	F	None	Orbital region	Rapidly growing neoplasm with medial deviation of the eyeball	-	Rhabdo myosarcoma	Nodular fasciitis	No recurrence	None
Lang²⁰	1994	Newborn	M	38 weeks gestation after emergency C-section due to fetal distress following 36-h labor. - low birth weight for gestational age (2.6 kg)	Right eyeball	Right eye proptosis, lateral displacement of the eyeball, clear watery discharge from the eye and right nasal cavity. Normal ophthalmologic evaluation.	CT: large soft tissue mass in the orbit extending to the anterior cranial fossa, ethmoid sinus, nasal cavity and right maxilla.	Tumor	Fascicles of spindle cells arranged in a nodular pattern with abundant inflammatory cells. Positive for vimentin and smooth muscle alpha-actin.	No recurrence	Cerebrospinal fluid leakage due to a defect in the anterior cranial fossa floor post-excision. No ocular motility defect; ptosis resolved in 2 weeks.
Recchia²¹	1997	31	F	Retinopathy of prematurity	Supranasal left orbit	Firm, painless lesion with erythema (2 months). Normal eye exam.	CT: well-defined spherical mass on the supranasal orbital rim. No bone erosion, calcification, or soft tissue invasion.	Epidermoid cyst	Mitotic figures, no atypical mitosis, multinucleated cells, or cellular atypia. Positive for vimentin.	No recurrence	None
Hymas²²	1999	168	M	None	Lower eyelid	Painless, firm, mobile lesion with rapid growth after trauma (1 month).	MRI: spherical preseptal mass contacting the left orbital rim, no bone destruction.	Chronic vascular lesion	Pseudo capsule, densely cellular with benign proliferation.	None	None

Continued.

Author	Year	Year (months)	Sex	Past medical history	Topography	Clinical presentation	Imaging studies	Clinical speculation	Histopathological study	Evolution	Complications
Shields²³	2001	8	M	None	Super temporal right orbit	Rapidly growing (5 weeks), firm, painless lesion.	Irregular bundles and interlaced fascicles of large spindle-shaped cells with abundant eosinophilic cytoplasm. Immature fibroblasts, irregularly arranged cells, prominent tumor cell nuclei. Positive for vimentin and smooth muscle actin; negative for desmin and myoglobin.	Epidermoid cyst	Irregular bundles and interwoven fascicles of large spindle cells with abundant eosinophilic cytoplasm. Consisting of immature and active fibroblasts, irregularly arranged cells and prominent nuclei of tumor cells with irregular size and shape. Positive for vimentin and smooth muscle actin. Markers such as desmin and myoglobin were negative.	No recurrence (6 months)	None
Ruoppi²⁴	2004	18	F	None	Left orbital rim	Lesion extending and compressing the left nasal cavity.	MRI: well-defined tumor with necrotic areas. No invasive growth or bone erosion.	Rhabdo myosarcoma	Spindle cells in myxoid stroma with randomly dispersed fibroblasts. No necrosis. Positive for vimentin and actin; negative for desmin. Low proliferation index.	No recurrence (36 months)	None
Kim²⁵	2005	16	M	none	Deep periorbital area (temporal muscle)	Rapidly growing lesion (2 months).	-	Neurogenic tumor	Nodular fasciitis	No recurrence	None
De paula²⁶	2006	36	M	None	Right upper eyelid	Round, firm, painless lesion with ptosis.	CT: well-defined mass in the superolateral orbital wall. No bone involvement.	Myo fibromatosis	Well-defined proliferation of spindle cells with fibroblasts arranged in fascicles, nuclear prominence, focal myxoid areas, and mature-appearing lymphocytes. Positive for vimentin and smooth muscle actin; negative for S-100, desmin, CD34, and cytokeratin's.	No recurrence	None

Continued.

Author	Year	Year (months)	Sex	Past medical history	Topography	Clinical presentation	Imaging studies	Clinical speculation	Histopathological study	Evolution	Complications
Damasceno²⁷	2009	3	M	None	Inner right upper eyelid	Reddish, solid, nodular lesion.	MRI: subcutaneous process with mild extension into the anterior medial orbit.	-	Spindle fibroblastic cells, some robust, in a myxoid stroma. Rounded to oval fibroblastic nuclei, no atypia. Myxoid stroma with numerous inflammatory cells. Positive for smooth muscle actin and vimentin.	No recurrence (60 months)	None
Mukherjee²⁸	2011	108	F	None	Anterior supertemporal left orbit	Gradual enlargement (8 months), painless, no vision impairment.	CT: poorly defined soft tissue mass.	-	Interwoven bundles of fibroblastic cells in loosely arranged myxoid stroma with scattered blood vessels and chronic inflammatory cells.	No recurrence	None
Gupta²⁹	2014	192	F	None	Right orbital apex	3-month progressive vision loss, headache, pain with eye movement.	CT: enhancing mass at orbital apex, local erosion of lesser sphenoid wing. MRI: gadolinium-enhanced tumor extending along optic nerve into intracranial space.	Atypical orbital tumor	Reactive lesion of spindle cells with eosinophilic cytoplasm in myxoid and collagenous stroma. No inflammatory cell infiltration. KI-67 positive, atypical mitotic figures. Positive for CD45, CD68, smooth muscle actin (1A4, HHF35), vimentin.	No recurrence (30 months)	Optic neuropathy
Richards³⁰	2016	144	M	None	Right supraorbital region	Rapidly progressive, painless, unilateral mass with diplopia and proptosis.	CT: frontal bone expansion, cortical erosion, extension through orbital roof. MRI: multilobulated heterogeneous mass in frontal sinus displacing frontal lobe.	Epidermoid cyst orbital compartment syndrome, mucocele, idiopathic orbital inflammation, post septal cellulitis, leptomeningeal cyst.	Myxoid-type nodular fasciitis. Negative for myogenin, ALK-1, ER, and PR. The cytoplasm of spindle cells was positive for vimentin, beta-catenin, smooth muscle actin, and muscle-specific actin (variably positive). BCL-2 was weakly positive, and CD99 was variably positive. Some occasional cells were positive for desmin, and the vessels within the lesion were positive for CD34.	No recurrence (12 months)	None

Continued.

Author	Year	Year (months)	Sex	Past medical history	Topography	Clinical presentation	Imaging studies	Clinical speculation	Histopathological study	Evolution	Complications
Matsushashi ³¹	2018	26	F	None	Left optic canal	Strabismus, right vision loss.	MRI: mass on left optic canal roof. CT: mass with local erosion of optic canal roof.	Osteohemangioma, intraosseous hemangioma, intraosseous meningioma, fibrous dysplasia, Langerhans cell histiocytosis.	Hyperplastic spindle cells in fibrotic interstitial tissue, no significant nuclear pleomorphism or malignancy. KI67 proliferation index: 2%. CD68 staining showed rare positive cells.	No recurrence (3 months)	None
Tom liu ³²	2019	9	M	None	Left temporal orbit	Rapidly growing, firm, immobile, painless mass (2 months).	CT: soft tissue mass with calcifications and bone erosion.	Dermoid cyst Rhabdomyosarcoma	Soft spindle cells, atypical mitosis. Positive for smooth muscle actin.	No recurrence (3 months)	None
Shay keren ³³	2020	96	F	History of suprasellar pilocytic astrocytoma.	Left orbital rim	Rapidly growing, firm, painless mass.	CT: soft tissue lesion deep to the fascial layer, mild underlying bone thinning.	Rhabdomyosarcoma	Spindle cells infiltrating skeletal muscle, associated with small nerve fascicles. Immunostaining positive for smooth muscle actin.	No recurrence (24 months)	None
Navarrete ¹⁰	2021	10	F	None	Right upper eyelid	Firm, painless, oval, immobile mass.	CT: well-defined, homogeneous lesion.	Malignant tumor	Well-defined mesenchymal proliferation, non-encapsulated, focally infiltrative with focal myxoid areas and spindle cells. Mitotic activity. Positive for actin, CD68, factor XIIIa.	No recurrence (8 months)	None
Compton ³⁴	2022	204	M	None	Left supraorbital region	Rapidly growing (1 month), painless, fixed mass.	CT: enhancing mass in the superior right orbit with local bone remodeling and erosion. MRI: 2.2 cm solid mass superficial to temporal muscle.		Benign neoplastic growth of spindle cells surrounded by inflammatory cells and extravasated red blood cells in a loose myxoid stroma, consistent with nodular fasciitis.	No recurrence (6 months)	None

Similarly, Navarrete et al described a healthy infant with a rapidly growing, firm, painless nodule in the right upper eyelid.¹⁰ The histopathological result revealed a well-circumscribed, non-encapsulated mesenchymal proliferation with focal myxoid areas and eosinophilic spindle cells on a collagenized background. Immunostains were positive for actin and calponin, with weak positivity for CD31, CD68, and factor XIIIa.¹⁰

The etiology of NF remains unclear; cases have been linked to previous trauma or a "reactive" process triggered by injury. Another theory involves USP6 gene rearrangement, indicating clonal neoplastic proliferation.^{11,12}

Clinically, NF presents as a solitary, mobile, circumscribed, painful subcutaneous nodule with rapid growth, rarely exceeding 2–3 cm within a few weeks.¹¹

Histopathologically, it is characterized by spindle cell proliferation arranged in a disordered manner with a myxoid stroma, accompanied by dilated blood capillaries and extravasated erythrocytes.² Non-atypical mitotic figures can be observed. Smooth muscle actin and vimentin immunomarkers are positive, desmin is only focally positive, while S100 and CD34 are negative.⁶ Subtypes include subcutaneous, intramuscular and fascial forms, with the subcutaneous type being the most common, as in our case.⁷

Differential clinical diagnoses include lipoma, fibromatosis, dermatofibroma, neuroma, epidermal cyst, among other subcutaneous tumors.¹³ On histopathological grounds, NF should be differentiated from benign fibrous histiocytoma, dermatofibrosarcoma protuberans, fibrosarcoma, leiomyosarcoma, and spindle cell carcinoma.¹⁴

Fibrosarcoma is a malignant tumour that should be ruled out because it is also a spindle cell tumour that can have an active mitotic rate. However, it is common to find atypical mitoses and significant nuclear pleomorphism.¹¹ Fibrosarcoma can additionally be differentiated by its growth pattern and cellularity, characterized by densely packed interspersed bundles of spindle cells forming a "herringbone pattern." Fibrosarcoma typically occurs in a deeper location, is larger, and has a longer duration.¹³

The initial clinical diagnosis for the lesion in our patient was a dermoid cyst, given its prevalence in childhood. Typically, a dermoid cyst presents as a soft, mobile, endophytic mass in the zygomatic arch or orbital/craniofacial region.¹¹ Surgical excision is the treatment of choice, although spontaneous remission has been reported. Fine-needle aspiration cytology (FNAC) and conservative management with clinical monitoring may also be effective.^{2,15}

We conducted a comprehensive literature review, analysing all reported cases of orbital NF between 1990

and 2022. Key parameters included patient demographics, clinical presentation, histopathological findings, treatment approaches and outcomes.

We identified 20 cases, with an equal distribution between male and female patients. The most common clinical presentation was a rapidly growing, painless orbital mass. Histopathological analysis revealed spindle cell proliferation with myxoid stroma, positive immunostaining for actin and vimentin and variable expression of other markers. Surgical excision was the primary treatment, with no reported recurrences or postoperative complications (Table 1).

CONCLUSION

NF, or subcutaneous pseudosarcomatous fibromatosis, is a benign, rapidly evolving lesion that can pose diagnostic challenges, particularly in pediatric patients, due to its atypical presentation. Although rare in children, it should be considered in the differential diagnosis of subcutaneous tumours, especially in the head and neck region. While most cases resolve spontaneously, surgical excision remains the preferred treatment. This case highlights the benign nature of the lesion and the importance of familiarity with this condition for accurate clinical and diagnostic approaches.

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