# Midfacial degloving approach to myxoid type nodular fasciitis of the maxilla in a 16-month-old female

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#### Abstract

Nodular fasciitis is a rare, benign tumor of mesenchymal fibroblasts arising from the deep fascia or bony periostium and presents as a mass in the subcutaneous tissues. Due to its rapid growth, it may mimic a malignant lesion. In children, the myxoid type of nodular fasciitis (also termed infiltrative fasciitis, pseudosarcomatous fasciitis, or pseudosarcomatous fibromatosis) is most common with involvement of the head and neck. To date, four cases of nodular fasciitis specifically involving the maxilla have been reported, only one of which includes a description of the surgical approach. This case report describes a 16-month-old female with nodular fasciitis of the nasolabial groove arising from the maxilla. The objective is to discuss and illustrate the imaging appearance of this rare maxillary location of myxoid fasciitis and to describe the successful resection of the mass utilizing the midfacial degloving approach.

## INTRODUCTION

Nodular fasciitis is a rare, benign tumor of fibroblastic growth that most often presents in patients between 20 and 40 years of age but may also affect children [ $_{123}$ ]. While the specific cause for nodular fasciitis is unknown, it is thought to be part of a spectrum of "quasi-neoplastic" lesions [ $_4$ ]. Interestingly, but of unknown significance, is the fact that a history of trauma has been reported to precede the occurrence of the lesion in some cases [ $_5$ ]. In addition to the relationship with trauma, chromosomal abnormalities have been described and may suggest a neoplastic origin [ $_6$ ]. The misdiagnosis as a sarcoma stems from the rapid growth, mitotic activity, and numerous spindle cells [ $_1$ ]. The exact rate of occurrence is unknown because of the common misdiagnosis as a sarcoma [ $_4$ ].

## CASE REPORT

A 16-month-old girl presented to our children's tertiary referral center with a rapidly enlarging mass that had been present for 2 weeks. The patient had no pain, fever or epiphora. The parents did not recall any specific trauma (major or minor) recently involving the child's face. The past medical history was unremarkable and development was age appropriate.

Physical exam revealed an overall healthy and active 16month-old child with a firm 2.0 cm mass obscuring the left nasolabial fold. Anterior rhinoscopy revealed the mass nearly obstructing the left nasal cavity. The mucosa of the nasal cavity and overlying facial skin were uninvolved. The remainder of the physical exam, including the eye exam, was unremarkable.

Since the lesion was initially thought to involve the lacrimal sac, the young girl was first seen by ophthalmology. The nasolacrimal duct was probed and flushed intra-operatively, revealing no obstruction. The solid intranasal mass was biopsied, but not resected. The pathology suggested nodular fasciitis (Figure 1).

#### Figure 1

Figure 1: Histopathology of nodular fasciitis in a 16-monthold female. (a) Histological evaluation reveals spindle cells in a loose, myxoid background (low power H&E stain). (b) At the periphery of the lesion, compact stroma (star) is seen at an interface with native partially eroded bone (arrowhead).



CT imaging was then performed and demonstrated a large,

solid mass arising from in the maxilla, following the nasolacrimal fold. Bony remodeling suggested a chronic, non-aggressive process. No bone destruction was seen and the lacrimal gland was uninvolved (Figure 2).

#### Figure 2

Figure 2: Nodular fasciitis in a 16-month-old female with a left side face mass for 1 week. CT images after contrast administration in the (a) coronal and (b) axial planes reveal a low attenuation soft tissue mass with mild enhancement (arrows) that closely follows the course of the left nasolacrimal duct. There was no central contrast enhancement. (c) Axial CT image demonstrates bony remodeling without destruction suggesting chronic mass effect (arrows).



The child was referred to the otorhinolaryngology service and the mass was surgically removed via a midfacial degloving approach (Figure 3).

#### Figure 3

Figure 3: Intraoperative photo illustrates surgical access to the maxillary lesion via the midfacial degloving approach.



The gray, friable tumor measured 2.7 X 2.0 X 1.7 cm (craniocaudal x transverse x anteroposterior, respectively) and pathological evaluation of the entire resected mass confirmed the diagnosis of nodular fasciitis specifying the myxoid type. The surgical and post-operative periods were uncomplicated. The patient was discharged home on post-operative day two. There was no recurrence to date by exam or CT imaging (more than 3 years later).

### DISCUSSION

Nodular fasciitis is rare, but well described in the literature  $[_{789}]$ . It typically presents with a painless, firm, soft tissue mass, however, pain or tenderness is also possible upon presentation  $[_{510}]$ . In adults, the upper extremities are most often involved compared to children who tend to have lesions of the head and neck  $[_{11}]$ . The distribution is as follows: upper extremities (particularly the volar aspect of the forearm), 46%; trunk, 20%; head and neck, 18%; lower extremities, 16%  $[_{123}]$ . It is estimated that up to 20% of myxoid type cases occur in the pediatric head and neck  $[_{12}]$ . Only four cases of nodular fasciitis are reported as specifically arising in the maxilla; the other entries generally refer to the "face"  $[_{8913}]$ .

Upon histologic examination, nodular fasciitis can be further categorized as myxoid, cellular, or fibrous [ $_{310}$ ]. The myxoid type of nodular fasciitis is more common in children with an average age of presentation of 3 years. Of the four cases previously reported in the maxilla, only one describes the surgical approach, which was a Weber-Ferguson incision [ $_{14}$ ].

A variety of lesions may be included in the differential diagnosis of pediatric soft tissue facial masses, such as rhabdomyosarcoma, fibrosarcoma, myxoid liposarcoma, myxofibrosarcoma, fibromatosis, neurolimoma, neurofibroma and fibrous histiocytoma [ $_{815}$ ].

Nodular fasciitis is treated by surgical excision with a 1% chance of recurrent disease after total or near total resection  $[_{271617}]$ . Rarely, chemotherapy is also utilized. Spontaneous degeneration and regression of nodular fasciitis of the cheek over a 6-month duration has been described  $[_{18}]$ . Despite the importance of surgical removal, few descriptions of the surgical approach to head and neck nodular fasciitis are described in the literature. Indeed, of the four cases described as involving the maxilla, only one discusses the surgical intervention  $[_{13}]$ . Cotter, et al described a similar maxillary nodular fasciitis lesion of a 21-month-old child. Cotter's report describes a Weber-Ferguson incision for access to the high maxillary lesion  $[_{13}]$ . The child had a good cosmetic outcome and no recurrence of tumor.

Compared to adults, the pediatric skeleton has several distinct differences that allow a sublabial or midfacial degloving approach to lesions of the upper maxilla. First, the craniofacial ratio is three to one in newborns, compared to two to one in adults [19]. Second, one of the more striking

features of the pediatric facial skeleton is the relationship of the orbital floor, which is nearly even with the nasal floor [19]. These features of the pediatric facial skeleton combine to offer adequate access to the entire maxilla and premaxillary region via a mid-facial degloving approach.

In conclusion, this report of a 16-month-old female with myxoid fasciitis, adds to the sparse literature available on this rare lesion. This report is unique in that it describes the midfacial degloving approach, which can be used in the resection of pediatric maxillary lesions without the need for an external incision, allowing for excellent aesthetic results.

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