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

Z Boyd, N Krad, P Nicklaus, L Lowe

Citation

Z Boyd, N Krad, P Nicklaus, L Lowe. *Midfacial degloving approach to myxoid type nodular fasciitis of the maxilla in a 16-month-old female*. The Internet Journal of Pediatrics and Neonatology. 2008 Volume 10 Number 1.

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

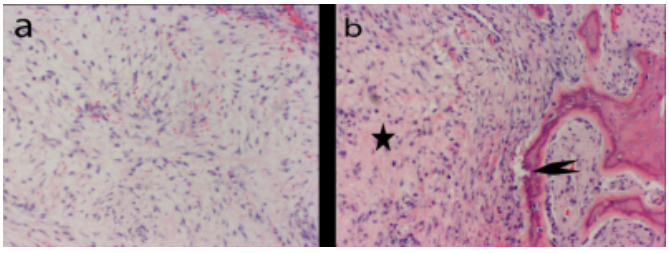
A 16-month-old girl presented to our children’s tertiary referral center with a rapidly enlarging mass that had been present for 2 weeks. The patient had no pain, fever or epiphora. The parents did not recall any specific trauma (major or minor) recently involving the child’s face. The past medical history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

Physical exam revealed an overall healthy and active 16-month-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-month-old 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).

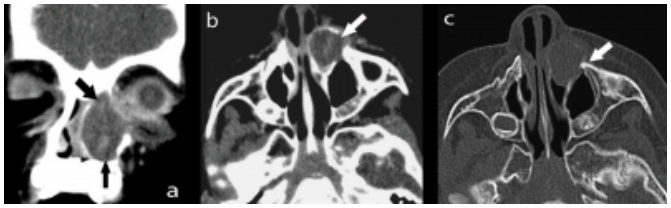


CT imaging was then performed and demonstrated a large,

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

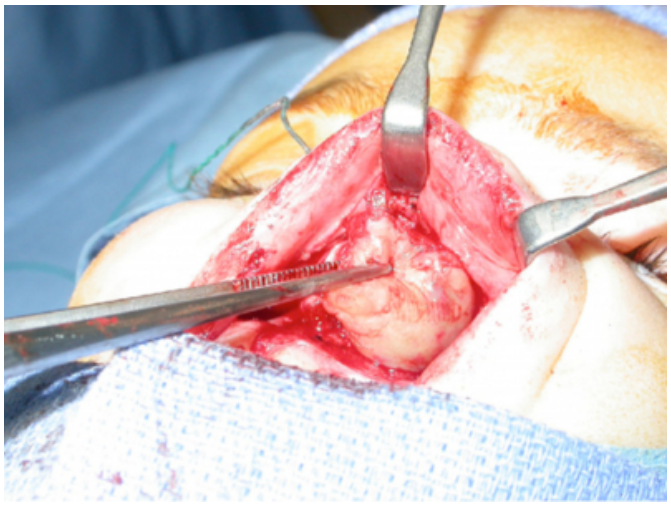
Figure 2: Nodular fasciitis in a 16-month-old female with a left side face mass for 1 week. CT images after contrast administration in the (a) coronal and (b) axial planes reveal a low attenuation soft tissue mass with mild enhancement (arrows) that closely follows the course of the left nasolacrimal 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 pre-maxillary 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.

CORRESPONDENCE TO

Zachary T Boyd University of Missouri-Kansas City School of Medicine 2411 Holmes St. Kansas City, MO 64108 (573) 216-0118 zachary.boyd@gmail.com

References

1. Weiss SW, Goldblum JR, Enzinger FM. Enzinger and Weiss's soft tissue tumors. 4th ed. St. Louis: Mosby; 2001.
2. Meister P, Bückmann F, Konrad E. Nodular fasciitis (analysis of 100 cases and review of the literature). Pathol Res Pract1978 Jun;162(2):133-65.
3. Shimizu S, Hashimoto H, Enjoji M. Nodular fasciitis: an analysis of 250 patients. Pathology1984 Apr;16(2):161-6.
4. Kim S, Kim H, Park S, Baek C, Baek J, Byun H, Kim Y. Nodular fasciitis in the head and neck: CT and MR imaging findings. AJNR Am J Neuroradiol2005 2005 Nov-Dec;26(10):2617-23.
5. Leung L, Shu S, Chan A, Chan M, Chan C. Nodular fasciitis: MRI appearance and literature review. Skeletal Radiol2002 Jan;31(1):9-13.
6. Donner L, Silva T, Dobin S. Clonal rearrangement of 15p11.2, 16p11.2, and 16p13.3 in a case of nodular fasciitis: additional evidence favoring nodular fasciitis as a benign neoplasm and not a reactive tumefaction. Cancer Genet Cytogenet2002 Dec;139(2):138-40.
7. Bernstein KE, Lattes R. Nodular (pseudosarcomatous) fasciitis, a nonrecurrent lesion: clinicopathologic study of 134 cases. Cancer1982 Apr 15;49(8):1668-78.
8. DiNardo LJ, Wetmore RF, Potsic WP. Nodular fasciitis of the head and neck in children. A deceptive lesion. Arch Otolaryngol Head Neck Surg1991 Sep;117(9):1001-2.
9. Werning JT. Nodular fasciitis of the orofacial region. Oral Surg Oral Med Oral Pathol1979 Nov;48(5):441-6.
10. Wang X, De Schepper A, Vanhoenacker F, De Raeve H, Gielen J, Aparisi F, Rausin L, Somville J. Nodular fasciitis: correlation of MRI findings and histopathology. Skeletal Radiol2002 Mar;31(3):155-61.
11. Dahl I, Jarlstedt J. Nodular fasciitis in the head and neck. A clinicopathological study of 18 cases. Acta Otolaryngol1980;90(1-2):152-9.
12. Allen PW. Nodular fasciitis. Pathology1972 Jan;4(1):9-26.
13. Cotter CJ, Finn S, Ryan P, Sleeman D. Nodular fascitis of the maxilla in a child. J Oral Maxillofac Surg2000 Dec;58(12):1447-9.
14. Cotter C, Finn S, Ryan P, Sleeman D. Nodular fascitis of the maxilla in a child. J Oral Maxillofac Surg2000 Dec;58(12):1447-9.
15. Toledo AS, Rodriguez J, Cuasay NS, Koch DB, Ray CG, 3rd, Langer BG, Skoly S. Nodular fasciitis of the facial region: CT characteristics. J Comput Assist Tomogr1988 Sep-Oct;12(5):898-9.
16. Meister P, Buckmann FW, Konrad E. Extent and level of fascial involvement in 100 cases with nodular fasciitis. Virchows Arch A Pathol Anat Histol1978 Oct 26;380(2):177-85.
17. Dinauer P, Brixey C, Moncur J, Fanburg-Smith J, Murphey M. Pathologic and MR imaging features of benign fibrous soft-tissue tumors in adults. Radiographics2007 2007 Jan-Feb;27(1):173-87.
18. Yanagisawa A, Okada H. Nodular fasciitis with degeneration and regression. J Craniofac Surg2008 Jul;19(4):1167-70.
19. Bluestone CD. Pediatric Otolaryngology. 3 ed: Sanders Co.; 1996.

Author Information

Zachary T. Boyd
University of Missouri-Kansas City School of Medicine

Nuha Krad, MD
Department of Radiology, University of Illinois College of Medicine-Peoria

Pamela J. Nicklaus, MD
Department of Surgery, University of Kansas Medical Center

Lisa H. Lowe, MD
Department of Radiology, Children's Mercy Hospitals and Clinics