



Case Report:

Nodular Fasciitis of Neck in Childhood.

Authors

Saurabh Varshney, Professor & Head,

Bhagat S, Associate Professor,

Bist SS, Associate Professor,

Mishra S, Assistant Professor,

Shirazi N, Associate Professor*,

Agarwal V, Senior Resident,

Kabdwal N, Senior Resident,

Department of E.N.T. & *Pathology, Himalayan Institute of Medical Sciences, (HIHT University), Jollygrant, Doiwala, Dehradun - 248140, India.

Address for Correspondence

Dr. Saurabh Varshney,

Professor & Head,

Department of E.N.T.,

Himalayan Institute of Medical Sciences, (HIHT University),

Jollygrant, Doiwala,

Dehradun - 248140, India.

E-mail: drsaurabh68@gmail.com

Citation

Varshney S, Bhagat S, Bist SS, Mishra S, Shirazi N, Agarwal V, Kabdwal N. Nodular Fasciitis of Neck in Childhood. *Online J Health Allied Scs.* 2012;11(4):13. Available at URL: <http://www.ojhas.org/issue44/2012-4-13.html>

Open Access Archives

<http://cogprints.org/view/subjects/OJHAS.html>

<http://openmed.nic.in/view/subjects/ojhas.html>

Submitted: Nov 29, 2012; Accepted: Jan 9, 2013; Published: Jan 25, 2013

Abstract: Nodular fasciitis, is a benign, pseudo sarcomatous proliferative lesion of the soft tissue, which is frequently misinterpreted as sarcoma, both clinically and microscopically. It is a reactive lesion composed of fibroblasts/myofibroblasts and most commonly found in extremities and trunk. NF has been described in the head and neck region in 10-20% of cases. Many pathologists do not consider NF in the differential diagnosis of soft tissue masses arising in the Head neck region. NF that occurs in otherwise healthy individuals usually presents with a history of rapid growth, and is commonly found in the upper extremities and on the chest and trunk. The importance of otolaryngologists being aware of the existence of this entity in this area of the body is stressed. It has a confirmed perfectly benign clinical course, and simple excision, as tissue-sparing as possible, is the treatment of choice. A case of NF over the neck in a 05-year-old female not associated with trauma who presented with a localized mass over her left neck is presented.

Key Words: Benign; Neck; Nodular fasciitis; Paediatric.

Introduction

Nodular fasciitis (NF), a benign proliferation of fibroblasts, commonly presents as a solitary, well-circumscribed, rapidly growing soft tissue mass. It is most commonly located in the upper extremities, particularly on the volar aspect of the elbow, and on the chest and trunk. NF is located in the head and neck only in 10% to 20% of cases. Although benign, it can often be confused with a more malignant process and therefore must be properly diagnosed histologically. The cause of NF is unknown but an association with trauma may be present. Treatment is most commonly by local surgical excision, and recurrence is rare.

Case Report

A 5 years old female child referred by other hospital presented with 2 years history of a progressively growing

swelling over the left side of neck. She denied any history of trauma to the region or any previous radiation to the head or neck.



Figure 1: Left neck mass

Physical examination revealed a 3 x 4 cm firm, nontender, well defined, non fluctuating, non reducible, immobile mass, with pulsations over mass on left side of neck (External carotid artery), along upper 1/3rd of SCM, extending vertically from level of hyoid bone to lower border of thyroid cartilage, horizontally from anterior border of SCM to midline. (Figure 1)



Figure 4a, 4b: Per operative images



Figure 5: Excised mass

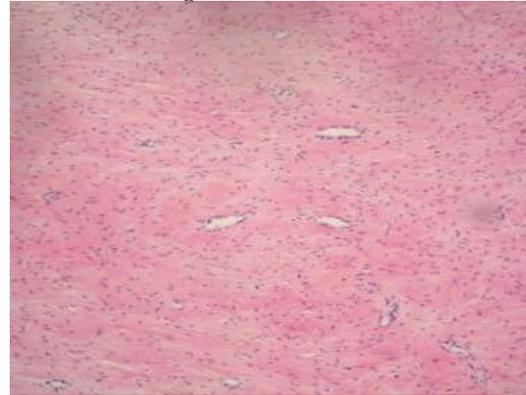


Figure 6: Histopathological Photograph of nodular fasciitis, showing loosely organized spindle cells with scattered extravasated red blood cells and dense keloidal collagen (HE stain, ×400)

The patient's postoperative course was uneventful except for features of Horner's syndrome on left side, and a six-month follow-up showed that her scar healed well. There was no evidence of residual neck contour deformity. (Figure 7).



Figure 7: Post operative image

Gross examination of the pathological specimen showed a well-circumscribed, 7 cm greyish-white mass. (Figure 5) Microscopically there were ill-defined whorls and fascicles of spindle-shaped cells with tapering cytoplasm and elongated nuclei (Figure 6). The latter were vesicular with a fine chromatin pattern and small nucleoli. Nuclear hyperchromatism and pleomorphism were absent. Cellularity was generally moderate as a result of separation of the cells by edema. However, a few foci were more solidly cellular. Scattered mitotic figures were observed but no atypical forms could be found. There was a prominent vascular network in the lesion, some parts of which had a "tissue culture" appearance that resembled granulation tissue.

The presence of an edematous and richly vascular tissue culture pattern combined with the absence of nuclear atypia, abnormal mitotic figures and a well developed fascicular pattern constituted reassuring evidence that the lesion was indeed benign and the features seen best fitted nodular fasciitis—a pseudosarcomatous lesion.

Discussion

Nodular fasciitis was first described in 1955 by Konwaler et al, as Subcutaneous pseudosarcomatous fibromatosis (fasciitis).¹ A variety of names have been applied to these lesions including inflammatory pseudotumor, pseudosarcomatous fibro-myxoid tumor, pseudosarcomatous

fasciitis, pseudosarcomatous fibromatosis, infiltrative fasciitis, postoperative spindle cell nodule and nodular fasciitis.² It is believed to be an exaggerated reparative process of uncertain cause. It is a reactive, self-limited, proliferation of fibroblasts most commonly found in the subcutaneous tissue. NF is a benign lesion but can often be confused with myofibromatosis or a sarcoma due to its rapid rate of growth, rich cellularity and mitotic activity.³ Nodular fasciitis is common in young adults (between 25 and 35 yrs) and less frequently in children. NF can occur almost anywhere on the body; however, the most common location in the adult population is in the upper extremities (39% to 54%), especially on the volar aspect of the elbow. They can also be found on the trunk (15% to 20%) and the lower extremities (16% to 18%). NF is located in the head and neck only in 7% to 20% of cases.³ In children, however, the head and neck region is the most common site. Approximately 10% of all lesions are found in children.⁴ In addition, it is more commonly found in those in the third through fifth decades of life; only 10% to 20% are found in those over 50 years of age.⁵

Exact cause of the lesion is not known but it is considered to be a self-limiting reactive process rather than a true neoplasm.^{6,7} The cause is unknown but has often been linked to local injury or trauma. In the literature, 10% to 15% of cases have been found to be associated with trauma,^{2,5,8} but this was not the case in our patient. Although most commonly located on the extremities and then the trunk, it is estimated that the head and neck region represents only 10 to 20%. The majority of cases arise in the soft tissue, i.e. fascia, muscle, or subcutaneous tissue. Interestingly, cases in the head and neck region often involve dermal tissue. It presents as a solitary painless, rapidly growing nodule over several weeks' duration. It usually ranges from 1 cm to 3 cm in greatest diameter.

In adults, these are commonly located in the upper extremities (flexor surface of the forearm) and the trunk (chest wall and back). In infants and children, nodular fasciitis is present in the head and neck region. The lesion consists of nodular, nonencapsulated mass usually less than 3cm in diameter. The cut surface may show firm and grey white or soft and gelatinous areas.⁶

The subtypes are:

- Fascial type- Poorly circumscribed lesion, extends along the superficial fascia and interlobular septa of subcutaneous fat.
- Subcutaneous type- Well circumscribed lesion, extends into the subcutis.
- Intramuscular type- Well circumscribed lesion, grows into the skeletal muscle.
- Intra-dermal type - Lesion present in the dermis (intra-dermal fasciitis)

The condition is self-limited, and proper diagnosis is essential to avoid unnecessary aggressive treatment. Diagnosis is often a challenge because it may be confused with a malignant tumor due to its aggressive clinical behavior and histological features. Immunohistochemical staining can be a useful tool to aid in the diagnosis.^{7,9}

Proper diagnosis requires histological confirmation. NF typically shows well-circumscribed, unencapsulated nodules composed of spindle cells. Microscopically early cases of Nodular fasciitis display zonation effect with maturation from the centre (hypocellular or hyalinized) to the periphery (hypercellular with inflammatory cell, blood vessels). In between, the loose myxoid area is populated by non-pleomorphic myofibroblasts loosely arranged with a tissue culture look. The background stroma shows variable myxoid change. The proliferating cells are of fibroblastic and myofibroblastic type. The high cellularity and mitotic activity

can mimic sarcoma. It is especially problematic in the pediatric population in which nodular fasciitis is not commonly encountered while mesenchymal malignancies of the head and neck are of fundamental concern. The alternative possibilities are leiomyosarcoma, fibrosarcoma and even sarcomatoid carcinoma. The rapid clinical onset, presence of stromal chronic inflammation and lack of cytological atypia are all critical features that help identify the lesion as benign.^{7,10} On immunohistochemistry NF demonstrates focal smooth muscle and muscle specific actin and calponin, but not usually desmin, h-caldesmon or CD34. CD68 may be positive in some cases. The following features rule out malignant tumour: i) Absence of atypia ii) Absence of atypical mitotic figures iii) Small size iv) Short history v) Superficial location in young adults.⁶

Local excision is the treatment generally advocated for nodular fasciitis but intra-lesional steroids have been tried with varied success.¹¹ Spontaneous regression of these lesions is also known to occur.¹² Owing to its ominous presentation nodular fasciitis has a tendency to be over-treated. Generally, if recurrence does occur after excision, incomplete resection should be suspected but a malignant process should be reconsidered.^{13,14}

Conclusion

NF is an uncommon benign proliferation of fibrous tissue rarely seen in neck. It is important to distinguish it from a malignant process because NF can be successfully treated with complete local excision.

References

1. Konwaler BE, Keasbey L, Kaplan L. Subcutaneous pseudosarcomatous fibromatosis (fasciitis) *Am J Clin Pathol.* 1955;25:241-252.
2. Han W, Hu Q, Yang X, Wang Z, Huang X. Nodular fasciitis in the orofacial region. *Int J Oral Maxillofac Surg.* 2006;35:924-927.
3. Silva P, Bruce IA, Malik T, Homer J, Banerjee S. Nodular fasciitis of the head and neck. *J Laryngol Otol* 2005;119:8-11.
4. Zuber TJ, Finley JL. Nodular fasciitis. *South Med J.* 1994;87:842-844.
5. Martinez-Blanco M, Bagan JV, Alba JR, Basterra J. Maxillofacial nodular fasciitis: A report of 3 cases. *J Oral Maxillofac Surg.* 2002;60:1211-1214.
6. Dahl I, Jarlstedt J. Nodular fasciitis in the head and neck. A clinicopathological study of 18 cases. *Acta Otolaryngol.* 1980;90(1-2):152-159.
7. Weinreb I, Shaw AJ, Perez-Ordoñez B, Goldblum JR, Rubin BP. Nodular fasciitis of the head and neck region: a clinicopathologic description in a series of 30 cases. *J Cutan Pathol.* 2009 Nov;36(11):1168-1173.
8. de Paula SA, Cruz AA, de Alencar VM, Akaishi PM, Chahud F. Nodular fasciitis presenting as a large mass in the upper eyelid. *Ophthalm Plast Reconstr Surg.* 2006;22:494-495.
9. Nishi SPE, Brey NV, Sanchez RL. Dermal nodular fasciitis: three case reports of the head and neck and literature review. *Journal of Cutaneous Pathology.* 2006;33(5):378-382.
10. Nair P, Barrett AW, Theodossy T. Oral nodular fasciitis: Case report. *Br J Oral Maxillofac Surg* 2004;42:360-362.
11. Graham BS, Barrett TL, Goltz RW. Nodular fasciitis: Response to intralesional corticosteroids. *J Am Acad Dermatol* 1999;40:490-492.
12. Fletcher CD. Diagnostic Histopathology of Tumors. vol 2. 2nd ed. Churchill Livingstone: London; 2000. p. 1486-1489.
13. Vyas T, Bullock MJ, Hart RD, Trites JR, Taylor SM. Nodular fasciitis of the zygoma: A case report. *Can J Plast Surg.* 2008 Winter;16(4):241-243.
14. Huang CD, Lin YH, Lee FP. Postauricular nodular fasciitis. *Otolaryngol Head Neck Surg.* 2007;137:164-165.