Published in final edited form as:

JAMA Otolaryngol Head Neck Surg. 2016 August 1; 142(8): 801–802. doi:10.1001/jamaoto.2015.3775.

Pathology Quiz Case: 22 year old with a supraclavicular mass

Amrita Ray, MPH, Rebecca Hoesli, MD, and Matthew E. Spector, MD

University of Michigan Health System, Department of Otolaryngology- Head and Neck Surgery,
Ann Arbor, Michigan

Amrita Ray: amiray@med.umich.edu; Rebecca Hoesli: rchoesli@med.umich.edu; Matthew E. Spector:
mspector@med.umich.edu

A 22-year-old female presents with a 4-month history of a 2 cm mass in her right supraclavicular region. She states that she had a cough and fatigue at the time, but no other URI symptoms at onset. She attributes her fatigue to staying up late studying. She denies any trauma and has not tried any treatments, including antibiotics.

She denies night sweats, fever, chills, weight loss, generalized pruritus or lymphadenopathy in other areas of her body. She denies any voice change, dysphagia, odynophagia, hoarseness, hemoptysis, hematemesis or weight loss. Her family history is positive for breast cancer in her mother and she is a lifelong nonsmoker.

Physical exam reveals palpable non-tender, freely movable mass in the right supraclavicular region. Palpation of the parotid, submandibular glands and thyroid gland does not reveal any suspicious masses or tenderness. No mucosal lesions, scars or masses were found on intraoral examination, with clear oropharynx. CT performed revealed a 2.0 cm right supraclavicular mass just anterior and superior to the right subclavian vein with central patches of hypoattenuation. [Figure 1]

Initial ultrasound guided FNA performed was non-diagnostic. Subsequent ultrasound guided core needle biopsy on supraclavicular aspirate showed a benign spindle cell proliferation arranged in a tissue-culture like pattern with a myxoid to collagenous stroma. Immunohistochemical stains demonstrate the cells to be positive for actin and negative for cytokeratin and S100 protein. [Figures 2-3]

What is your diagnosis?

Nodular Fasciitis of the Omohyoid Muscle

First described in 1955 by Konwaler et al., nodular fasciitis is a rapidly growing benign soft tissue mass characterized by abundant cellularity, rapid growth, and high mitotic activity.

Corresponding author: Matthew E. Spector, M.D., 1500 E. Medical Center Dr., Ann Arbor, MI 48109-5312, (734)936-3172 Phone, (734)936-9625 Fax, mspector@med.umich.edu.

Author Contributions: Amrita Ray, MPH: Conception and design, acquisition and interpretation of data, drafting of the manuscript
Rebecca Hoesli, MD: Revision of the manuscript

Matthew E Spector, MD: Revision of the manuscript, administrative and technical support

Disclosures: The authors have no conflicts of interest to report

Located in either the deep subcutaneous tissues or superficial fascia, nodular fasciitis is a non-neoplastic proliferation of fibroblasts and myofibroblasts. Although the pathological and clinical presentation resembles malignant soft tissue sarcomas, often leading to misdiagnosis, the condition is benign. The etiology of nodular fasciitis is uncertain, with some proposing it as a reactive process associated with trauma while others describe it as a self-limiting clonal process with neoplastic origins (Botton et al, 2006). Originally titled pseudosarcomatous fibromatosis, other titles such as nodular pseudosarcomatous fasciitis, pseudosarcomatous fasciitis, infiltrative fasciitis or proliferative fasciitis may be used to refer to the same disease process.

Nodular fasciitis preferentially affects adults (80%) usually within the first 2-4 decades of life with a slight male predominance (Allen, 1972). Although rare in children, it can present as a chest wall tumor in children (1.8% of all childhood solid chest wall tumors (Suh *et al*, 2014)) and should be kept in consideration. After surgical excision, local recurrence has been documented but is rare (Bemrich-Stolz *et al*, 2010).

The majority of lesions are subcutaneous and circumscribed, lending them amenable to biopsy or excision. Patients notice a rapidly growing subcutaneous painless mass over the course of 3-6 weeks. Masses are solid, nodular, firm or rubbery and usually plateau at 2-3 cm in size, rarely exceeding 4 centimeters. The upper extremities are the most common area of presentation, especially the flexor aspect of the forearm. They are less frequently found on the trunk (20%), head and neck (20%), and lower extremities (13%) (Allen, 1972), and may have an associated history of trauma (Shimizu *et al*, 1984). In the head and neck region, lesions are often found on the neck or face, and less rarely on the forehead/scalp regions or around the eyes (Allen, 1972).

Histologically, nodular fasciitis demonstrates non-neoplastic proliferation of fibroblasts and myofibroblasts, although there is vast array of presentation pattern. The cellular composition of the mass can further divide nodular fasciitis into three subtypes: type 1 (myxoid), type 2 (cellular) and type 3 (fibrotic). (Shimizu *et al*, 1984). The subtype of nodular fasciitis diagnosed may be dependent on the duration the lesion has been present, as fibrotic type may represent the most mature and longest lasting lesions. Shimizu *et al*, 1984

- Type 1 myxoid variant has fibroblast-like cells within a stroma rich in collagen and/or myxoid ground substance. This type tends to form a large tumor, occurs more frequently in older patients and appears similar to the myxoid variant of malignant fibrous histiocytoma.
- Type 2 cellular nodular fasciitis has significantly less ground substance and stroma, with large plump fibroblast-like spindle cells that have vesicular nuclei. The high cellular proliferation and mitotic activity most closely resembles sarcomatous lesions.
- Type 3 fibrous type most closely resembles other benign lesions and have more spindle-shaped, slender fibroblast-like cells and increased collagen production. (Shimizu *et al*, 1984)

The tumor's rapid presentation and histological profile closely resembles sarcomas, often leading to misdiagnosis. Beyond sarcomas, other pathological processes to consider include neurofibromas, palmer fibromatosis, extra-abdominal desmoid tumors, and early presentation of myositis ossificans if intramuscular.

The diagnosis can be confirmed by fluorescence in situ hybridization using a probe to the USP6 17p13 locus. (Oliveira 2004) USP6 rearrangements in 10% or greater lesional cells are considered positive with a sensitivity and specificity of 90% and 100% respectively. Our patients show a USP6 rearrangement in 44% of lesional cells, confirming the diagnosis.

Imaging with ultrasound may show ill-defined hypoechoic nodules in the deep or superficial fascial layers, while imaging with MRI showed T1 isointense nodules and T2 hyperintense nodules. (Khuu *et al*, 2014). However this utility is limited, and fine needle biopsy or core biopsy may reveal more detailed histological findings. An initial observation period of several weeks may be warranted, especially considering the self-limiting/spontaneous regression course of the disease. Steroid injections have also been found to aid lesion regression. However, surgical excision still remains the mainstay treatment of choice. Lesions will rarely reoccur after excision.

References

- Botton A, Biere A, Galakhoff C. Nodular fasciitis of the face: causal role of piercing. *Presse Med*. 2006; 35(2 Pt 1):237–238. [PubMed: 16493353]
- Allen PW. Nodular fasciitis. *Pathology*. 1972; 4:9–26. [PubMed: 4501523]
- Suh JH, Yoon JS, Park CB. Nodular fasciitis on chest wall in a teenager: a case report and review of the literature. *J Thorac Dis*. 2014; 6(6):E108–E110. DOI: 10.3978/j.issn.2072-1439.2014.05.18 [PubMed: 24977016]
- Bemrich-Stolz CJ, Kelly DR, Muensterer OJ, et al. Single institution series of nodular fasciitis in children. *J Pediatr Hematol Oncol*. 2010; 32:354–7. [PubMed: 20502354]
- Shimizu S, Hashimoto H, Enjoji M. Nodular fasciitis: an analysis of 250 patients. *Pathology*. 1984; 16:161–6. [PubMed: 6462780]
- Oliveira AM, His BL, Weremowicz S, et al. USP6 (Tre2) fusion oncogenes in aneurysmal bone cyst. *Cancer Res*. 2004; 64(6):1920–1923. Oliveira et al. *Cancer Res* 2004 64:1920-3. [PubMed: 15026324]
- Khuu A, Yablon CM, Jacobson JA, Inyang A, Lucas DR, Biermann JS. Nodular fasciitis: characteristic imaging features on sonography and magnetic resonance imaging. *J Ultrasound Med*. 2014; 33(4): 565–73. [PubMed: 24658936]

Figure 1. Ultrasound guided FNA

Figure 2. CT

Figure 3. Immunohistochemical Stain: 10× magnification

Figure 4. Immunohistochemical Stain: 40× magnification