Nodular Fasciitis and Myxolipoma of the Larynx

A rare case report with brief literature review

*Ahmad Al Omari,1 Wisam Qarqaz,1 Rasha A. Alrhman,1 Ra’ed Al-Ashqar,1 Samir Al Bashir,2 Mohammed Alorjani2

1Department of Special Surgery, Faculty of Medicine, Jordan University of Science and Technology, Irbid, Jordan; 2Department of Pathology and Laboratory Medicine, Jordan University of Science and Technology, Irbid, Jordan

*Corresponding Author’s e-mail: aialomari@just.edu.jo

Abstract

Nodular fasciitis (NF) is a peculiar, rapid-growing soft tissue lesion, typically appearing in subcutaneous tissue. 20% of NF occur in the head and neck region, where they can involve any anatomic site. Laryngeal involvement, however, is quite rare. On the contrary, Lipoma is recognized as a slow growing, benign mesenchymal tumour. Myxolipoma is a rare variant which has a prominent myxoid background. Laryngeal lipoma is infrequent, accounting for only 0.6% of all benign laryngeal lesions. Here, we report a unique case of adult laryngeal nodular fasciitis coexisting with myxolipoma in a 61-year-old male patient, describing their clinical and histopathological features, the strategies used to treat such conditions along with a brief review of the literature. The purpose is to broaden the differential diagnosis of rapid-growing laryngeal masses that cause airway obstruction and to stress the significance of integrative interdisciplinary collaboration on reaching an accurate diagnosis, thereby allowing proper management for benign pathologies and avoiding any futile aggressive treatment.

Keywords: Nodular fasciitis, Larynx, Stridor, Myxolipoma.

Introduction
Nodular fasciitis, a subtype of benign mesenchymal spindle cell tumor-like lesions, is a noncancerous, reactive fibrous proliferation which makes up almost 11% of all benign soft tissue tumours.\(^1\)

Although Konwaler et al. first described this condition as ‘pseudosarcomatous fasciitis’ in 1955, it was not until 1961 that Shuman used the term ‘nodular fasciitis’ for the very first time, and since then it has been broadly adopted by authors.\(^2\)

It arises mostly in the extremities and trunk,\(^3\) with the larynx being a very rare location for NF, so diagnosis of NF is challenging in this region. In most cases, the patient presents with a painless, rapidly expanding subcutaneous mass. However, unlike other locations, laryngeal NF may cause life-threatening symptoms by obstructing the respiratory tract.

In contrast, lipoma is a common slow developing benign mesenchymal tumour, with roughly 15% of lipomas being found in the head and neck. Due to the presence of scant amounts of adipose tissue in the larynx, laryngeal occurrence is infrequent.\(^4\) Myxolipoma is a rare variant which has prominent myxoid areas.

**Case Report**

A 61-year-old male, who is a heavy smoker, presented to the ENT outpatient clinic in King Abdullah University Hospital, October 2020, complaining of a change in his voice, which had started few months earlier, associated with dysphagia, intermittent dyspnoea and choking. His surgical history was unremarkable except for laparoscopic cholecystectomy 1 year ago. There was no history of trauma. Head and neck examination was unremarkable. A large right supraglottic mass was identified by flexible nasolaryngoscopy. Radiological findings were suggestive of a locally advanced laryngeal cancer (Figure 1). Biopsy was taken by suspension microlaryngoscopy for precise diagnosis and results were suggesting a benign/borderline spindle cell neoplasm.

Since it has a benign nature, lesion was debulked trans-orally by laser and histologic diagnosis was in favour of laryngeal nodular fasciitis. Shortly after surgery, the mass rapidly enlarged,
Tracheostomy was performed and he underwent complete mass excision with right partial laryngectomy through an open surgical approach (Figure 2). Repeated pathological examination of microscopic sections and immunohistochemistry study revealed same findings in accord of the aforementioned diagnosis of laryngeal nodular fasciitis, along with incidental myxolipoma (Figure 3 & 4). Postoperatively, the patient recovered uneventfully with no evidence of recurrence. He is currently much better and under regular follow up. Consent was obtained from the patient to publish this case report.

Discussion

Clinical features

Nodular fasciitis is a rare, but distinct, benign self limited mesenchymal neoplasm of fibroblastic/myofibroblastic derivation that resembles soft tissue sarcoma. The etiology is still not fully understood. Although not documented in the vast majority of patients, many believed that traumatic insult is the trigger for this reactive inflammatory response on most occasions. While occurring at all ages, this condition is most often diagnosed in adults 20 to 40 years of age, with no gender predilection.

Clinically, when appearing in the larynx, the main presenting symptoms are related to the degree of laryngeal obstruction. It can include hoarseness of voice, foreign body sensation, dysphagia, as well as dyspnoea and stridor that may require urgent medical attention. Despite most of the lesions being less than 2 cm in greatest dimension at the time of diagnosis, the size may vary from 0.5 to 10 cm.

Equivalently, lipoma is a benign tumor consisting of adipose tissue. It is considered the most common mesenchymal tumor, constituting 16% of soft tissue tumors. Lipoma of larynx can be subdivided according to the site of origin into Intrinsic (Endolaryngeal) type, or more commonly Extrinsic type. It has a male predominance and occurs over a wide age range (mean age 40 years) with a supraglottic predisposition.

The etiology of laryngeal lipomas is unclear. Unlike other locations, laryngeal lipomas may occasionally cause fatal airway obstruction with dyspnea and dysphonia being the most frequent
presenting symptoms. Grossly, it can be sessile or pedunculated, usually appearing as a smooth, well-encapsulated mass. The size typically ranges from 1 to 3 cm, but sometimes it may exceed 10 cm.\(^7\)

**Radiological findings**

On CT scan most of NF lesions appear as a well-defined homogeneous mass with low or isodensity and show moderate to strong enhancement. On MRI lesions will exhibit hypointense or isointense signals on T1-weighted sequences and show heterogeneous intermediate-to-high signal on T2-weighted sequences.\(^8,9\)

For lipomas, imaging serves a vital role in diagnosis. On CT scan it mostly simulates fatty tissue characteristics, thus lipomatous lesions appear as a homogenous mass with low attenuation. At MRI, lipoma has predominantly low signal intensity on T1-weighted images and markedly high signal intensity on T2-weighted images.\(^10\)

**Histopathology findings**

Lesions of NF are often composed of undulating short, intersecting fascicles of haphazardly arranged plump, immature fibroblasts and myofibroblasts in a loose myxoid and/or fibrous stroma which resembles the feathery or tissue culture-like appearance.

The cells have uniform, elongated non-pleomorphic nuclei with pale, fine chromatin and small but prominent nucleoli. Some typical mitotic activity is commonly seen. Numerous extravasated red blood cells, scattered lymphocytes, chronic inflammatory cells and multinucleated osteoclast-like giant cells are also present within the background.\(^11\)

Likewise, lipomas have some distinctive histological features, being generally composed of mature adipocytes bound by thin fibrous capsules. Myxolipoma demonstrates similar features, but with abundant extracellular mucoid matrix.\(^12\)

**Immunohistochemistry findings**
Fibroblasts often stain for Smooth Muscle Actin, muscle-specific actin and Vimentin. 
Meanwhile, none of lesional cells express S100 protein, β-catenin, CD34, keratin, caldesmon and desmin. The proliferation index with Ki67 can be high in reactive lesions such as NF. It should be noted that Immunohistochemistry is not of much help in diagnosing myxolipoma.

**Molecular and cytogenetic findings**
In regard to the post-genomic era and cytogenetic tests, few studies established the molecular and cytogenetic abnormalities and proved the neoplastic nature of NF. For example, Erikson-Johnson MR et al. described the USP6 rearrangement with the formation of the fusion gene MYH9-USP6 which is commonly observed in the lesions, and they referred to nodular fasciitis as ‘transient neoplasia’ in tribute to its self-limiting nature.

**Treatment and prognosis**
Given the paucity of cases of NF involving the larynx, their natural course is not fully understood. However, assuming that their behavior would be as with NF of other anatomic sites, as this lesion had neither a high local recurrence propensity nor metastatic potential, adequate surgical excision of the lesion with negative margins could be sufficient.

Yet, due to anatomic factors and critical structures, laryngeal NF lesions can be unamenable to simple complete laryngoscopic local excision. Hence, Partial laryngectomy, whether endoscopic or open (based on the lesion, surgeon’s skills and patient factors) could be an appropriate choice in favour of vocal function preservation. Total laryngectomy should be used in selected cases with advanced diseases or reserved as salvage surgery.

Other controversial conservative methods like intralesional corticosteroid injection can be considered when there are no substantial symptoms and spontaneous regression is expected. Surgical excision is recommended in the case of laryngeal lipoma. Depending on the size, endoscopic approach is preferred in small lipomas, whereas open surgical approach must be used if the lesions dimension is beyond 2 cm. As for benign lesions, the prognosis for laryngeal lipoma is very good. Recurrence is rare and is mostly due to hidden malignancy or inadequate excision. Because of this, long-term follow-up is recommended.
Conclusion

Nodular fasciitis of the larynx can mimic malignant tumors, thus reaching an exact diagnosis is very challenging. Although it has a favorable prognosis as compared to other aggressive laryngeal lesions, simple lesional resection in this unique location might be difficult to obtain and laryngectomy is inevitable. Laryngeal myxolipoma is rare but must be considered in the differential diagnosis of laryngeal masses. To the best of our knowledge, only four cases of laryngeal myxolipomas, one of them being juvenile, have been reported in the English literature.

Statement of Ethics:

The patient has given his written informed consent to publish his case (including publication of images).

Author’s Contribution:

AO and RAA performed the literature review. All authors contributed to the drafting of the manuscript. All authors approved the final version of the manuscript.

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Figure 1: (A) Axial, (B) sagittal CT scan showed a heterogenous laryngeal mass with thick-enhancing rim, measuring about 5.1 x 3.8 x 3.7 cm starts at the level of the hyoid bone and extends caudally reaching the right cricoid cartilage. (C) Coronal view on T1W MRI showed the same mass causing compression effect on the adjacent vessels with significant stenosis on supra-
glottic region and minimal invasion of the right thyroid cartilage. (D) PET scan revealed a hypermetabolic laryngeal mass.

**Figure 2**: (A+B) Intraoperatively, cystic lesion with extra-laryngeal extension incidentally found, mimicking laryngocele (white arrow). (C) Another smooth, well circumscribed, firm mass which seemed to involve the right thyroid cartilage and reach to the false vocal cords was identified (yellow arrow). (D) Excised specimen which includes part of larynx with cystic structure attached to a well-defined mass.
Figure 3: H&E image shows (A) Spindled and plump cells exhibiting fascicular arrangement in myxofibrotic background. Lesional cells demonstrate storiform architecture. (B) Benign looking, uniform, stellate fibroblasts resembling tissue culture-like appearance. (C+D) Myxolipoma consists of mature-appearing adipocytes with prominent myxoid areas. No evidence of atypia, plexiform vascular network or lipoblasts. (A: 20 x / B,C: 40x / D: 10 x magnification).
Figure 4: H&E image; 40 x magnification, shows (A) Numerous extravasated red blood cells and scattered chronic inflammatory cells. (B) Lesional cells of nodular fasciitis typically have ovoid nuclei and prominent nucleoli. Immunohistochemistry study revealed diffuse expression of SMA Immunohistochemical marker in the lesional cells (C) and CD10 positive immunostaining of nodular fasciitis (D).