

Recurrent Aggressive Fibromatosis with Laryngeal Involvement: A Case Report and Literature Review

Nathan Lloyd ^{1*}, Nadia Tello ², Daniel Nguyen ², Rahul Varman ², Joehassin Cordero ²

¹Texas Tech University Health Sciences Center School of Medicine

²Texas Tech University Health Sciences Center Otolaryngology Department

Corresponding Author: Nathan Lloyd, Texas Tech University Health Sciences Center, School of Medicine 3601 4th Street, Stop 8312, Lubbock, TX 79340

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Abstract:

Aggressive Fibromatosis (AF) is a rare, benign neoplasm originating from musculoaponeurotic stromal structures characterized by aggressive growth and infiltration of local tissues. To date, there are only six previously reported cases of AF involving the larynx.

Our case involves a 70-year-old female with a 5-year history of hoarseness and an enlarging neck mass consistent with aggressive recurrent fibromatosis. MRI showed a large, solid mass arising from the left anterior cervical space displacing the trachea and upper airway to the right. A year after the initial radical resection, the patient presented with recurrence. A second radical excision was performed not including a laryngectomy. Radiation therapy was consulted for possible local treatment to prevent subsequent recurrence of tumor.

Literature has reported up to 40-70% recurrence rates of AF within 18 months. There is minimal literature to guide treatment. Surgery seems to be the treatment of choice for head and neck AF, but management of recurrence is controversial.

Keyword: Aggressive Fibromatosis; musculoaponeurotic; laryngectomy

Introduction:

Aggressive fibromatosis (AF) is a rare, benign neoplasm with an incidence of 2 to 4 per 1 million people annually. It originates from musculoaponeurotic stromal structures and aggressively grows and infiltrates local tissues.¹ 10% of reported AF cases appear in the cervical region, and only six cases have reported involvement of the larynx.^{2,5} AF of the head and neck region tends to be more locally aggressive, making complete resection difficult.³ We present a case of aggressive recurrent fibromatosis involving the left anterior cervical neck along with the thyroid in a 70-year-old woman who has undergone multiple neck resections.

Case Report:

A 70-year-old woman presented to the ENT clinic with a 5-year history of an enlarging left neck mass. She did not seek treatment because she was afraid of the possible diagnosis. She denied any pain, dysphagia, or dyspnea. Her most noticeable symptom was a higher vocal pitch. On examination, a fixed, firm, greater than 10 cm mass with a superior boundary of the inferior portion of the left mandible and an inferior boundary of the left thyroid cartilage was noted. MRI showed a large, solid mass arising from the left anterior cervical space displacing the trachea by 2-2.5 cm to the right (Figures 1 and 2). The mass measured 10.8 x 7.5 x 9.7 x 7.5 x 6.6 cm. The mass was inferior to the parotid glands, invading medially and posteriorly to the trachea with medial and inferior extension to the left lobe of the thyroid.

The patient consented to surgery and the mass was excised by left modified neck dissection through levels I-VI with preservation of nerves, vessels, and surrounding structures. The mass was carefully removed from the carotid sheath, strap muscles, anterior body of the thyroid, and anterior trachea in succession. The mass was noted to extend into both retro-pharyngeal and retro-laryngeal spaces and was then removed posteriorly from the posterior trachea, thyroid cartilage, and thyroid. The dissection was difficult due to aggressive local invasion; thus, clear margins were not achieved. The mass was at least 12-15 cm in length, 10 cm in width, and 8 cm in height. It was sent for pathology which showed spindle cell proliferation consistent with deep fibromatosis with no clear margins. A post-operative CT scan with contrast was performed 8 months after surgery and showed significant reduction of fibromatosis tissue in the left neck.

Another noteworthy finding included residual fibromatosis tissue between the lower cricoid cartilage and the medial aspect of the upper left thyroid lobe. 15 months after resection the patient returned to clinic with a new chief complaint of left neck paresthesia with involvement of the left ear. She also mentioned that she has not been able to sing as well since the operation. She confirmed hoarseness and dysphagia but denied change in pitch and dyspnea. On physical exam, a noticeable mass of the left neck in the same location as the original mass was present. On follow-up 5 months later, the mass roughly doubled in size. A CT with contrast showed an 8.1 x 8.5 x 10.3 mass with a superior border of the mandibular ramus and inferior border of the left thyroid (Figure 3). Invasion of the left para-pharyngeal space, rightward deviation of the airway, and

narrowing of the glottis was also noted on imaging. The mass invaded deeper into the left lateral margin of the tonsillar soft tissues and vallecula. The mass was resected approximately 21 months after the first dissection (Figures 4, 5, and 6). Left radical neck mass excision included levels II-IV, removing the mass from the left thyroid bed, left carotid sheath, left parotid bed, and left posterior digastric muscle after careful mobilization of the sternocleidomastoid muscle. Erosion of the left thyroid cartilage into the pharynx was noted but the hyoid bone was left intact. After resection, radiation oncology was consulted for possible local radiation treatment to prevent tumor recurrence.

Discussion:

Fibromatosis is a benign, mesenchymal lesion composed of a proliferation of well-differentiated fibroblasts.⁴ The majority (two-thirds) of AF develop in the abdomen while the remainder are found extra-abdominally.³ 19-49% of AF cases are preceded by nonsurgical or surgical trauma.³ It is theorized that the pathogenesis involves an abnormal healing response with proliferation of immature fibroblasts leading to a fibromatosis tumor.³ AF of the head and neck regions are more aggressive variants of fibromatosis; they tend to be locally aggressive and have been known to erode bone, soft tissues, and vital structures.³ Due to this aggressive nature, clinical features such as paresthesia and weakness are seen as a result of pressure effects on local nerves and vascular structures as seen in our patient.³ After treatment, these tumors have a greater tendency to recur (40-70%) with most cases recurring within 18 months of surgical excision which was also observed in our patient.³ There is little literature currently available to guide treatment of wide local invasive AF. Treatment modalities include surgery, chemotherapy, radiotherapy, hormonal therapy, or a combination.¹ The choice of treatment modality depends predominantly on the tumor location, tumor size and age of the patient.⁵ To date, for head and neck AF, the standard treatment is primary surgical excision with clear margins to minimize the chance of recurrence. Unfortunately, numerous studies have reported that recurrence rate of AF is independent of surgical margin status.¹ Positive margins do not affect overall survival rate or 5-year disease-free survival, making standard treatment with clear margins controversial.¹ The few cases of AF that involve the larynx, have performed total or hemi-laryngectomy.² The majority of reported cases (66%) that performed hemi-laryngectomy and other less radical surgical interventions experienced recurrence.² Of those that recurred in the larynx, 75% performed a laryngectomy.² Other treatment modalities such as radiotherapy seem to reduce local recurrence rates of AF in adults but 40% of patients suffered from severe complications including pathologic fracture, pain, contracture, impaired range of motion, and skin cancer.¹ Chemotherapy regimens including Vinblastine and Methotrexate have lowered recurrence rates in pediatric patients.¹ The decision regarding treatment regimens for patients with AF of the head and neck is best made by a multidisciplinary team, consisting of otolaryngologists, radiation oncologists and medical oncologists. These multidisciplinary teams have been shown to improve decision making and reduce waiting times for cancer patients.⁶ Regardless of treatment modality performed, close follow-up and a high suspicion of recurrence is necessary.

Conclusion:

Controversy over treatment and management of AF in the head and neck region still exists. Minimal amount of literature on the topic is currently available, with even less available to guide the cases that involve the larynx. Cases described in the literature agree that surgery is the best treatment option but consensus regarding management of tumor recurrence has not been reached. Treatment and management, due to the lack of evidence in the literature, should be tailored to each individual patient and education of the risks and benefits of treatment modalities should be emphasized since no treatment, even surgery with clear margins, guarantees prevention of recurrence. The use of

multidisciplinary teams may improve decision making for these patients. The treatment of AF in the head and neck region is a topic that requires more attention and research.



Figure 1: T1 Non-contrast MRI demonstrating left neck mass with compression of midline structures

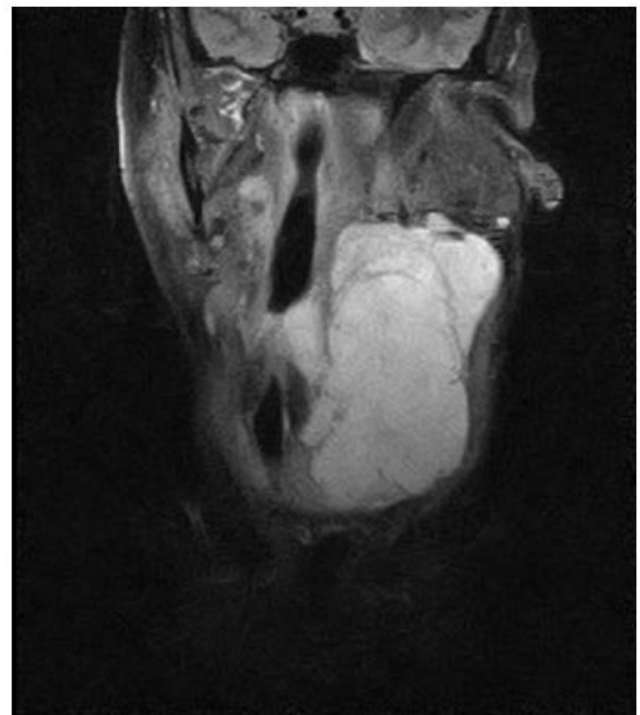


Figure 2: STIR MRI demonstrating left neck mass with compressive symptoms

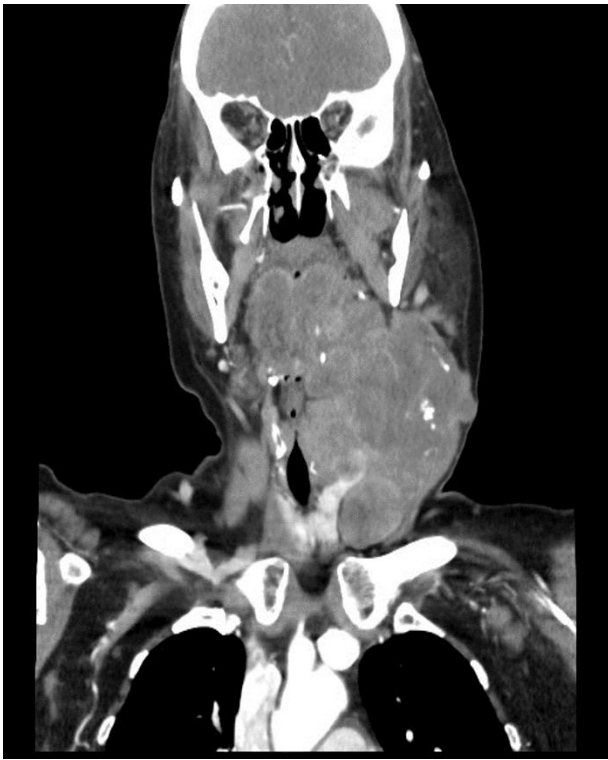


Figure 3: CT Scan with contrast demonstrating Recurrence of Aggressive Fibromatosis

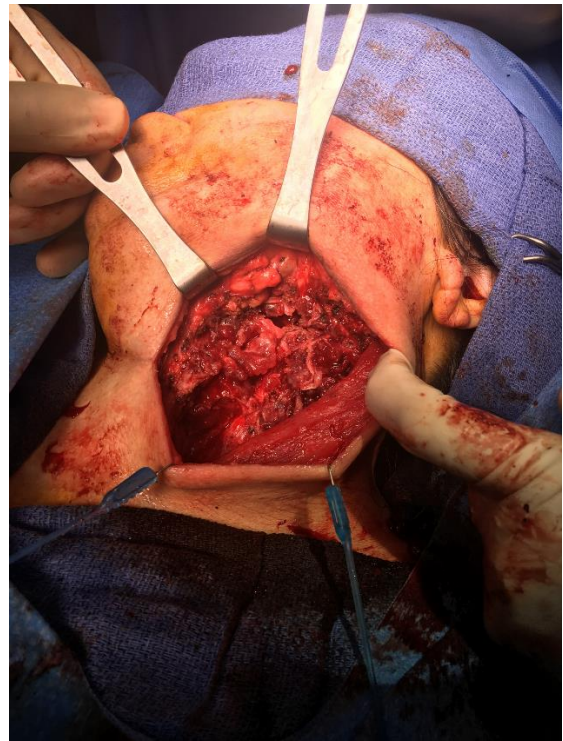


Figure 5: Intraoperative image of the neck after resection of the mass

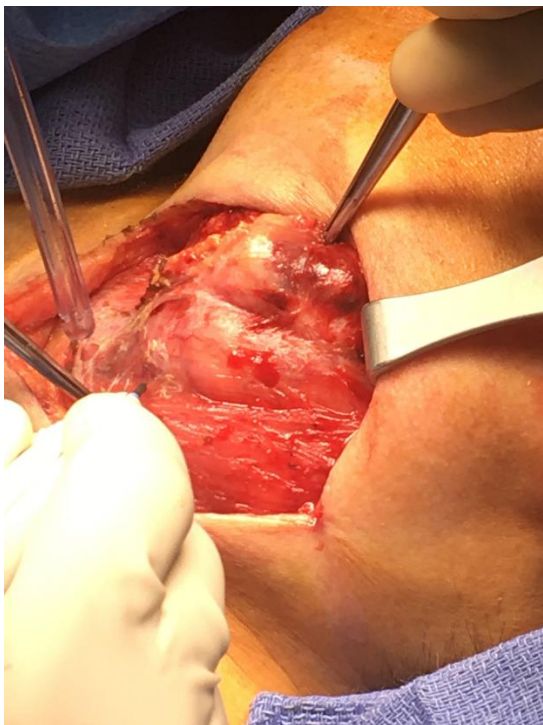


Figure 4: Intraoperative image of the neck mass prior to resection



Figure 6: Image of the resected mass

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