Retropharyngeal Inflammatory Myofibroblastic Tumor: A Case Report

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Abstract

Inflammatory myofibroblastic tumour (IMT) is a rare neoplasm that can mimic malignant disease both clinically and radiologically. Although more than 40 cases have been reported in the head and neck region, involvement of the retropharyngeal space remains uncommon. Preoperative diagnosis is often challenging due to nonspecific imaging features and inconclusive biopsy results. A 43-year-old male presented with intermittent but progressive stridor and neck swelling. A CT scan of the neck demonstrated a large multilobulated heterogeneous mass within the retropharyngeal space. A transcervical surgical excision was performed for removal of the mass. The final histopathology confirmed the diagnosis of an IMT. Within the follow up period of 12 months there was no evidence of disease recurrence. IMT is a rare neoplasm that uncommonly presents in the head and neck region, especially in the retropharyngeal area with two reported cases in literature. The rarity of this tumour, combined with variable histologic features present a challenge for diagnosis, treatment and prognostic considerations. Herein we present a case where surgical resection resulted in successful treatment without recurrence after 1 year.

Keywords: Inflammatory Myofibroblastic Tumour, Retropharyngeal Space, Head and Neck, Case report

Introduction

Inflammatory myofibroblastic tumour (IMT), also referred to as inflammatory pseudotumor, pseudosarcomatous myofibroblastic, fibromyxoid lesion and plasma cell granuloma, is a rare soft tissue neoplasm. Up to 60% of IMT cases are associated with a chromosomal rearrangement at location 2p23, the site of the anaplastic lymphoma kinase (ALK) gene.[2-4] Though most commonly known for its indolent behaviour, IMT is classified as an intermediate malignant neoplasm due to its potential aggressive biological behaviour. The subset of IMTs associated with aggressive behaviour have a 25% risk of local recurrence and 5% risk of metastasis. Recurrence risk is partly dependent on anatomical site and surgical resectability.[2]

As IMTs are rare, the majority of the published papers are case reports or small case series with variable anatomical locations (lung, orbit, peripheral nervous system, abdomen, pelvis, soft tissue, breast and other sites).[2,4]

Overall, they are less common in the head and neck region, with an incidence of 15%.[4] Of these, a small number have been reported involving the parapharyngeal space[5-8] and an even smaller number (2 reported cases in literature) in the retropharyngeal area.[9,10] Tumors in this location can clinically and radiologically mimic sarcoma, lymphoma, metastatic disease, neurogenic tumors, thyroid malignancy, or deep neck space abscess. Herein, we present a case of IMT located in the retropharyngeal space with extension to the retro-esophageal area. The case highlights challenges in clinical diagnosis in deep neck IMTs and the consequent need for definitive surgical excision for diagnosis; and the feasibility of safe en-bloc resection of a large mediastinally extending retropharyngeal IMT.

Case Report

A 43-year-old healthy male presented to the emergency department with a two-month history of intermittent stridor and intermittent dysphagia. There was no history of neck trauma, infection, or surgery. Upon clinical examination, the patient was stable and not in any acute airway distress. Neck examination revealed fullness in the left neck, just anterior to the sternocleidomastoid muscle (SCM), resulting in lateral displacement of the trachea. Flexible fibreoptic examination revealed an incidental finding of a small epiglottic cyst, no airway obstruction, good mobility of vocal cords and anterior displacement of the left arytenoid. Contrast enhanced CT scan demonstrated a large multilobulated heterogeneous mass centred behind the trachea and the thyroid gland, with inferior extension through the thoracic inlet into the superior mediastinum measuring 10.4*6.4*8.1cm. The mass displaced the thyroid gland and the trachea to the right and was spanning T1-T3 (Figure 1). An ultrasound-guided needle biopsy was done, which was non-diagnostic. Further to these initial investigations, he presented after one month later with the progression of his stridor with minimal effort and at rest. The clinical examination showed increased fullness in his neck. Based on imaging and clinical features a decision was made for a definitive surgical excision.

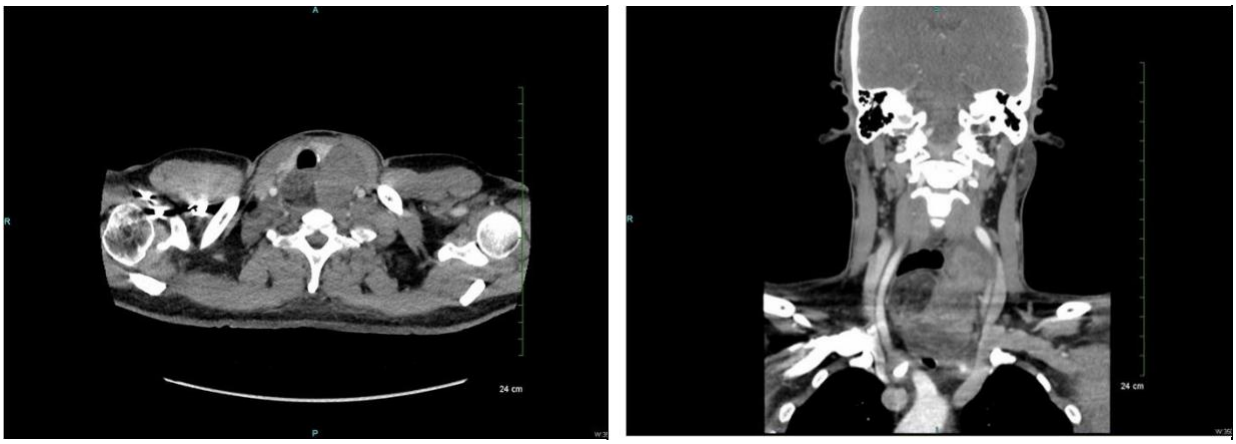
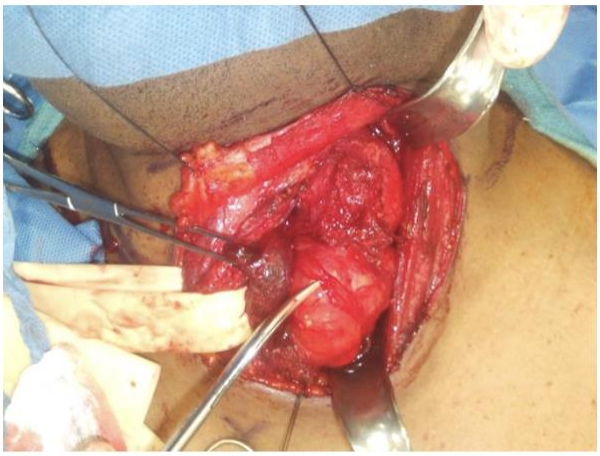


Figure 1. Pre-operative axial and coronal section of CT scan showing a multilobulated mass occupying retropharyngeal space and left neck with mediastinal extension and displacement of the trachea to the right side.

The patient was booked for an urgent excision of the mass to decompress his airway. Intraoperative neuromonitoring (NIM) of the recurrent laryngeal nerve was used. Intraoperatively the mass was firm and multilobulated and occupying neck space from level II to thoracic inlet. It was separate from the thyroid gland; however, the recurrent laryngeal nerve was displaced anteriorly by the mass, resulting in some tension on the nerve. Despite this, the nerve was responsive to the NIM throughout the procedure. The tumour extended inferiorly into the thoracic inlet and was pulled upward after freeing it from the recurrent laryngeal nerve. The tumour extended posteriorly and superiorly into the retropharyngeal space without the involvement of the prevertebral facia. The tumour was delivered in en-bloc with dimensions of 10.9*8.1*6.7cm (Figure 2). Following the removal of the mass, a laceration was noted in the trachea, which was repaired primarily. Bronchoscopy and esophagoscopy were done at the end of the procedure to ensure there were no other abnormalities.

a)



b)

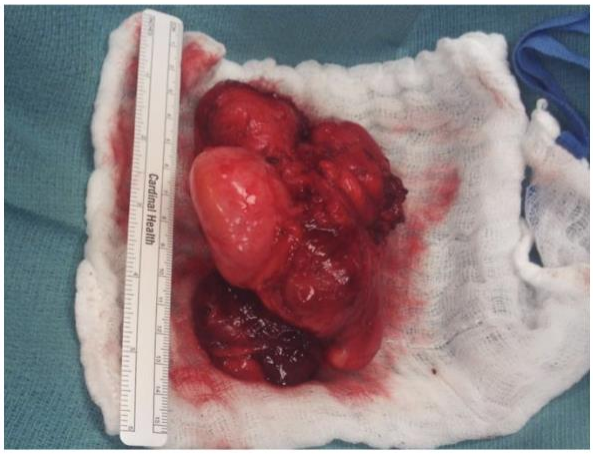


Figure 2: (a) Intraoperative view, showing the mass is separate from thyroid gland with stretching of the recurrent laryngeal nerve. (b) The biggest dimension of the tumor is measuring 10 cm.

The microscopic examination revealed a multinodular lesion consisting of loosely arranged plump, spindled myofibroblasts set in a myxoid background and associated with abundant blood vessels and a mixed inflammatory cell infiltrate consisting of lymphocytes, plasma cells and rare eosinophils (Figure 3). The plump myofibroblasts had vesicular nuclei with small nucleoli and focally demonstrate a ganglion-like appearance. Immunohistochemistry was performed and the spindle cells were positive for SMA (focal weak), desmin (focal) and CD34. The spindle cells were negative for MUC-4, h-caldesmon, SOX10, S100 protein, MSA, STAT6, CD15, CD30 and GFAP. ALK protein demonstrated only weak staining. ArcherDx sarcoma fusion panel (v1) was performed but did not detect an ALK gene arrangement. Despite the absence of an ALK fusion, the overall morphologic features were consistent with an inflammatory myofibroblastic tumour (IMT). The patient had uneventful postoperative recovery. During the 12 months follow-up period there were no signs of recurrence with no adjuvant therapy.

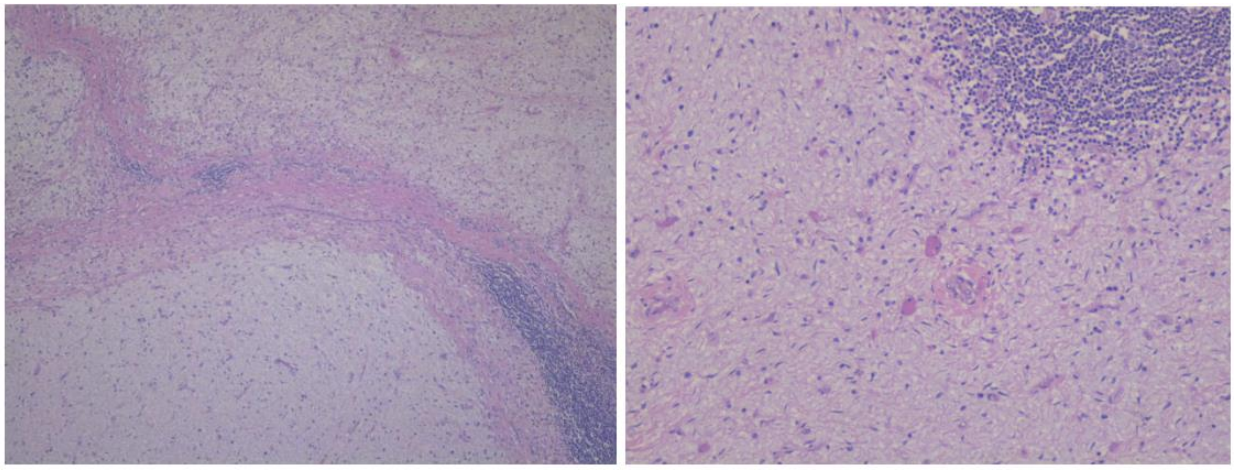


Figure 3: Inflammatory myofibroblastic tumour. Low power (a) and medium power (b) shows a multinodular myxoid lesion, consisting of loosely arranged plump, spindled myofibroblasts set in a myxoid background and associated with abundant blood vessels and a mixed inflammatory cell infiltrate consisting of lymphocytes, plasma cells and rare eosinophils (H&E stain. (a) 4X objective (b) 10X objective). Myofibroblastic phenotype was

confirmed with patchy SMA staining (not show). ALK immunohistochemistry showed weak, non-specific staining (not shown). No *ALK* gene alterations were identified using the ArcherDx sarcoma fusion panel.

This case report was exempted from requiring an ethics review and a waiver/exemption for informed consent was obtained from The Ottawa Hospital Integrated Research Information System ethics committee (IRIS).

Discussion

The clinical presentation of IMT is variable and depends on the involved anatomical location. In most cases, IMT involves the lung or orbit.[11] In our case, our patient presented with stridor, dysphagia, and the sensation of neck fullness. IMT can affect a patient at any age (from 9 days to 88 years), but the highest occurrence is among middle aged patients (18 to 40 years), as was the case for our patient.[1,2]

The pathogenesis of IMT remains unclear but various etiological agents have been investigated including prior surgery, autoimmune diseases, inflammation, infection, virus-induced trauma and abnormal response to a persistent external stimulus, which results in myofibroblast proliferation.[11] IMT is strongly correlated with *ALK* gene rearrangement in almost 50% of cases. In our case, despite the weakly positive stain with the *ALK* protein on immunohistochemistry, no *ALK* gene rearrangements were detected. Several studies have confirmed that the dysregulation of *ALK* leads to tumorigenesis by promoting abnormal phosphorylation.[11,12] The expression of *ALK* is considered a poor prognostic factor. That said, *ALK* gene-targeted drug therapy for *ALK*-positive IMT may prove to be a successful treatment in the future.[11] Other, less common gene rearrangements including *ROS1* and *NTRK3*, have been described in small subset of IMTs (~ 5-10%) and very rare cases show *RET* or *PDGFRB* gene rearrangements.[13] In our case, there were no identifiable etiologic risk factors to explain the development of IMT.

The treatment for IMT is primarily surgery via complete resection, balanced with the preservation of organs and function. A combined treatment approach is used when the tumour is not amenable for complete surgical resection. In these situations, post-operative radiotherapy and chemotherapy are indicated. Other treatment approaches including the use of corticosteroids and non-steroidal anti-inflammatory drugs have not proven to be beneficial.[11,14] Overall, there is no clear consensus regarding the best treatment modality of IMT in head and neck region. In our case, we were successful with complete resection without the need for adjuvant therapy.

Recent studies have explored the relationship between IMT and IgG4-related disease (IgG4-RD). IgG4-RD is an autoimmune disease characterized by multi-organ involvement. Some studies have identified IgG4-positive plasma cells in the mixed inflammatory cell component of IMT in various anatomic sites including the head and neck. Thus far, corticosteroids are the mainstay of treatment.[12] However, other immunosuppressive therapies could be used to prevent long term side effects of corticosteroid treatment and preventing high relapse rate including azathioprine, methotrexate, mycophenolate mofetil, and rituximab.[15]

This report has several limitations. As a single case report, the findings may not be generalizable to all patients with this condition. Additionally, the observations are based on an individual clinical course, and variations in patient characteristics, disease stage, and treatment approaches may lead to different outcomes in other settings. Therefore, while this case highlights important clinical considerations, larger studies are needed to further validate these observations and determine their broader applicability.

Conclusion

The diagnosis, prognosis, and treatment of IMT remain a challenge. Here we present a rare case of IMT in the retropharyngeal space with successful surgical resection without recurrence after a follow-up period of 1-year. Further investigation into the use of gene-targeted drug therapy, anti-inflammatories and immunosuppression are warranted particularly for cases for which surgical resection is not feasible.

Disclosure

The authors declare no conflict of interest in preparing this article. All contribution has been acknowledged and included in authorship. No non-author contribution to be noted. No data sharing agreement with any other party is currently in place. All relevant and significant data has been included in the manuscript.

References

1. Fu GX, Xu CC, Yao NF, Gu JZ, Jiang HL, Han XF. Inflammatory myofibroblastic tumor: A demographic, clinical and therapeutic study of 92 cases. Math Biosci Eng 2019 Jul;16(6):6794-6804.

2. Kovach SJ, Fischer AC, Katzman PJ, Salloum RM, Ettinghausen SE, Madeb R, et al. Inflammatory myofibroblastic tumors. J Surg Oncol 2006 Oct;94(5):385-391.

3. Honda K, Kadowaki S, Kato K, Hanai N, Hasegawa Y, Yatabe Y, et al. Durable response to the ALK inhibitor alectinib in inflammatory myofibroblastic tumor of the head and neck with a novel SQSTM1-ALK fusion: a case report. Invest New Drugs 2019 Aug;37(4):791-795.

4. Baranov E, Hornick JL. Soft Tissue Special Issue: Fibroblastic and Myofibroblastic Neoplasms of the Head and Neck. Head Neck Pathol 2020 Mar;14(1):43-58.

5. Kansara S, Bell D, Johnson J, Zafereo M. Head and neck inflammatory pseudotumor: Case series and review of the literature. Neuroradiol J 2016 Dec;29(6):440-446.

6. Segawa Y, Yasumatsu R, Shiratsuchi H, Tamae A, Noda T, Yamamoto H, et al. Inflammatory pseudotumor in head and neck. Auris Nasus Larynx 2014 Jun;41(3):321-324.

7. Maruya S ichiro, Miura K, Tada Y ichiro, Masubuchi T, Nakamura N, Fushimi C, et al. Inflammatory pseudotumor of the parapharyngeal space: a case report. Auris Nasus Larynx. 2010 Jun;37(3):397–400.

8. Kourtidis S, Saravakos P, Fiehn C, Preyer S. Inflammatory Pseudotumor in the Parapharyngeal Space: Is It Possible to Diagnose by Exclusion? Cureus [Internet]. 2021 Oct 19 [cited 2022 Aug 29]; Available from: https://www.cureus.com/articles/73948-inflammatory-pseudotumor-in-the-parapharyngeal-space-is-it-possible-to-diagnose-by-exclusion

9. Lee JH, Kim K, Chung SW, Choi YC, Lee A. A case report of inflammatory pseudotumor involving the clivus: CT and MR findings. Korean J Radiol 2001;2(4):231-234.

10. Fanous MM, Margo CE, Hamed LM. Chronic idiopathic inflammation of the retropharyngeal space presenting with sequential abducens palsies. J Clin Neuroophthalmol 1992 Sep;12(3):154-157.

11. Zhao J, Han D, Gao M, Liu M, Feng C, Chen G, et al. Inflammatory myofibroblastic tumor of the neck with thyroid invasion: a case report and literature review. Gland Surg 2020 Aug;9(4):1042-1047.

12. Tao J, Zhou ML, Zhou SH. Inflammatory myofibroblastic tumors of the head and nec. Int J Clin Exp Med 2015 Feb;8(2):1604-1610.

13. Hidetaka Y, Hornick JL. WHO Classification of Tumours Editorial Board. Soft tissue and bone tumours. Inflammatory Myofibroblastic Tumour. [Internet]. 5th ed. Vol. 3. Lyon (France): International Agency for Research on Cancer; 2020. Available from: https://tumourclassification.iarc.who.int/chapters/33

14. Cheng KJ, Wang SQ, Zhou SH. A case report of an inflammatory myofibroblastic tumor of the neck: A focus on the computed tomography and magnetic resonance imaging findings. Oncol Lett 2015 Jul;10(1):518-522.

15. Behzadi F, Suh CH, Jo VY, Shanmugam V, Morgan EA, Guenette JP. Imaging of IgG4-Related Disease in the Head and Neck: A Systematic Review, Case Series, and Pathophysiology Update. J Neuroradiol 2021 Sep;48(5):369-378.