Case Report

A Rare Case of Atypical Lipomatous Tumour of Larynx

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DOI: http://dx.doi.org/10.21276/ijcmsr.2021.6.1.7

How to cite this article: Sufyan Ibrahim, Venkatesh Manchikanti. A rare case of atypical lipomatous tumour of larynx. International Journal of Contemporary Medicine Surgery and Radiology. 2021;6(1):A34-A36.

A B S T R A C T

Introduction: Atypical lipomatous tumour of the larynx is an unusual entity and hence may masquerade as other tumours clinico-radiologically

Case report: 40 year-old female presented with complaints of dysphagia and dysphonia was evaluated with ultrasound initially and underwent CT and MRI of neck for better delineation and characterisation of lesion which revealed fat-containing soft tissue lesion in the left infra-hyoid region, involving the left paraglottic space.

Conclusion: We highlight this case for the rarity of the neoplasm leading to dilemma in establishing a diagnosis and delay in management.

Keywords: Atypical Lipomatous, Tumour of Larynx

INTRODUCTION

Atypical lipomatous tumour (ALT), otherwise also referred to as well-differentiated liposarcoma, is a low-grade, malignant, mesenchymal neoplasm commonly seen affecting the extremities, trunk or retroperitoneum.[1] ALT of the larynx is an unusual entity and hence may masquerade as other tumours clinico-radiologically.[2]

CASE REPORT

40 year-old female presented with complaints of dysphagia and dysphonia. On examination soft tissue swelling was palpable not moving with the protrusion of tongue. Ultrasound examination of the neck showed well-encapsulated hyperchoic lesion in the left infra hyoid region of neck with no significant internal vascularity(Figure 1B).

Contrast enhanced CT scan of the neck revealed a heterogeneously enhancing, well-defined, hypodense, fat-containing soft tissue lesion measuring 4x2cm in the left infra-hyoid region, involving the left paraglottic space and abutting the left vocal cord as well as the left submandibular gland anterolaterally(Figure 1A,C and D).

MRI a well defined heterogenous T1,T2 hyperintense lesion showing fat suppression on PDFS (C) noted in the left infra hyoid region in the left paraglottic space abutting the left vocal cord medially and submandibular gland laterally. Features were suggestive of Lipoma(Figure 2 A,B and C).

Patient underwent wide surgical excision of the tumour(Figure 2D) with 1 cm free margin. Microscopic histopathologic examination showed scattered atypical cells with few lipoblasts suggestive of lipoma-like subtype of atypical lipomatous tumour of the larynx, with free deep margins.

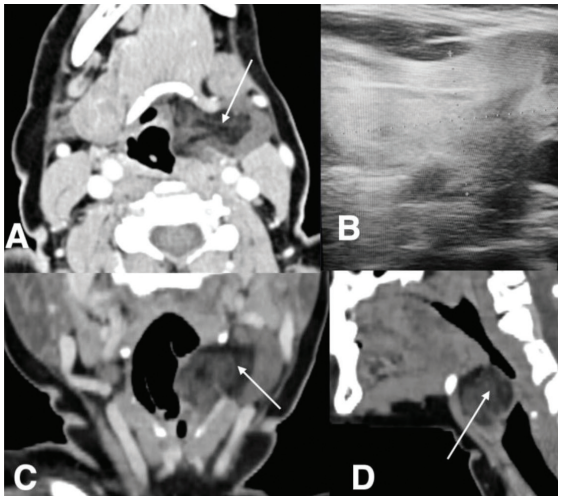

Figure-1: Axial contrast CT images(A, C and D) shows a well defined hypodense heterogenous fat containing lesion(white arrow) noted in the left infra hyoid region in the left paraglottic space abutting the left vocal cord medially and submandibular gland laterally. Axial USG image (B) showing hyper echoic lesion in the left infrahyoid region

DISCUSSION

Non-epithelial tumours are a small subset of laryngeal neoplasms, accounting for only upto 5% of all primary malignant laryngeal tumours, most commonly localising on the vocal cords or sub-glottis.[3] Chondrosarcoma, lymphoma, leiomyosarcoma, fibrosarcoma are more predominant malignant non-epithelial lesions, while liposarcoma, the second most common soft-tissue sarcoma, is extremely rare in the larynx.[4] Liposarcoma is a malignancy of adipocytic origin

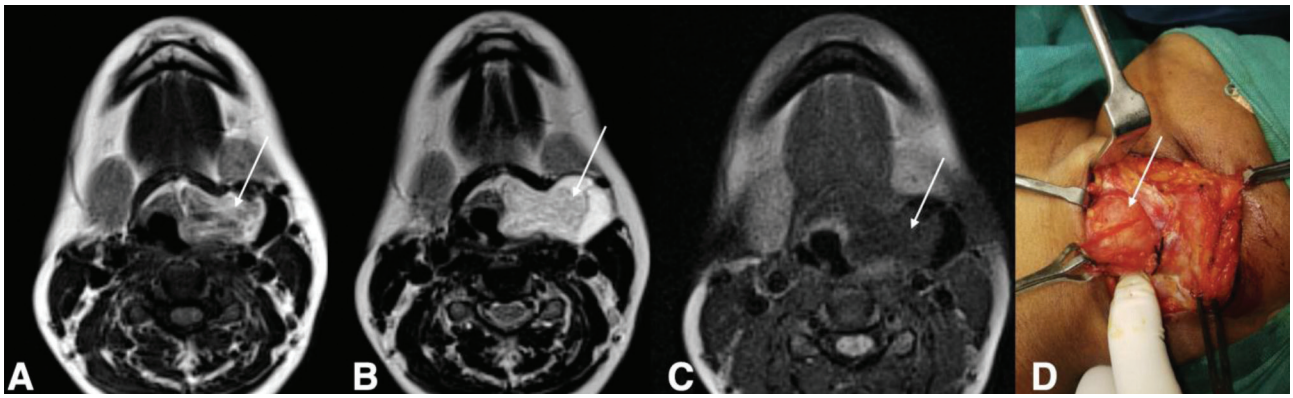

Figure-2: Axial contrast MRI images(A-T1, B- T2 and C-PDFS) shows a well defined heterogenous hyperintense lesion (white arrow)showing fat suppression on PDFS (C)noted in the left infra hyoid region in the left paraglottic space abutting the left vocal cord medially and submandibular gland laterally. Intraoperative photograph showing the lesion.

that can further be sub-classified into well-differentiated/ALT, myxoid/round-cell, dedifferentiated, pleomorphic or mixed variants.[1]

ALTs show a male preponderance, with elderly population being affected more commonly, in concordance with liposarcomas located elsewhere.[5] Usual clinical manifestations include hoarseness of voice (dysphonia), foreign-body sensation in the throat (globus), dysphagia, difficulty in breathing and stridor.[6] Macroscopically they may mimic retention cysts, benign lipomas, and giant fibrovascular or inflammatory polyps.[7] Secondary changes in the form of infection, haemorrhage, calcification or fat necrosis is not uncommon.[8] Most ALTs have an indolent course but multiple tumor recurrence is frequent, whereas dedifferentiation is exceptionally uncommon and distant metastasis is almost non-existent.[9]

CECT is essential for delineating the true extent of the disease, which shows inhomogenous attenuation of the fatty mass with significant amounts of soft-tissues within, and based on the amount of fat distributed in the tumor, liposarcomas can have solid, pseudocystic or mixed pattern.[10] MRI features such as deeper location, large size (>10cm), low fat. percentage (<75%), multiple thick septations (>2mm), nodular and globular areas, prominent enhancement and high T2 signal foci favour the diagnosis of ALT over lipoma.[11]

On histopathological examination of ALT, variable sized adipocytes containing mature fat, with bands of fibrotic stroma containing spindle cells with enlarged, heterochromatic nuclei are appreciated, and can be of lipoma-like, sclerosing, inflammatory or mixed subtypes.[12] MDM2 amplification and CDK4 overexpression are molecular characterisation, employed to differentiate from histologic mimickers.[8]

Superficially located ALT have a good prognosis with 5-year-survival rates of 100%, especially in solitary lesions with complete surgical excisions including free margins, but those in deeper locations have greater recurrence rates, due to incomplete resections, and are more at risk for transforming into dedifferentiated liposarcoma.[12] Despite this, conservative surgical approach is preferred mode of management due to the rarity of grade-progression and lack of significant morbidity with recurrences.[13] For inadequately excised tumors, post-operative radiotherapy may be beneficial.[12]

CONCLUSION

We highlight this case for the rarity of the neoplasm leading to dilemma in establishing a diagnosis and delay in management. Multimodality imaging as described in our case plays a major role for better delineation and characterisation of this tumour.

REFERENCES

1. Nouri H, Hassani R, Aderdour L, Raji A. The well-differentiated liposarcoma of the hypopharynx. Eur Ann Otorhinolaryngol Head Neck Dis. 2011;128(3):143–5.
2. Pajaniappane A, Farzan J, Green DM, De M. Well-differentiated liposarcoma of the epiglottis. J Laryngol Otol 2014;128(3):296–8.
3. Fasunla A, Ogundoyin O, Onakoya P, Nwaorgu O. Malignant tumors of the larynx: Clinicopathologic profile and implication for late disease presentation. Niger Med J 2016;57(5):280.
4. Saraydaroglu O, Narter S, Ozsen M, Coskun H. Non-epithelial tumors of the larynx: case series of 12 years. Eur Arch Oto-Rhino-Laryngology [Internet]. 2019
5. Han Y, Yang LH, Liu TT, Wang J, Li H, Yu G, et al. Liposarcoma of the larynx: Report of a case and review of literature. Int J Clin Exp Pathol 2015;8(1):1068–72.
6. Meis JM, Mackay B, Goepfert H. Liposarcoma of the Larynx: Case Report and Literature Review. Arch Otolaryngol Neck Surg. 1986;112(12):1289–92.
7. Okromelidze L, Middlebrooks EH, Bhatt AA. Lipoma of the Larynx and Other Differential Diagnoses. J Clin Imaging Sci [Internet]. 2019;9(1):51.
8. Zhu H, Sun J, Wei S, Wang D, Brandwein M. Well-Differentiated Laryngeal/Hypopharyngeal Liposarcoma in the MDM2 Era Report of Three Cases and Literature Review. Head Neck Pathol [Internet]. 2017;11(2):146–51.
9. Muddaiah A, Zaffarullah W, Tewary A. Recurrent well-differentiated liposarcoma of the larynx: A case report and review of literature. Eur Arch Oto-Rhino-Laryngology [Internet]. 2010;267(7):1163–5.
10. Nishida J, Morita T, Ogose A, Okada K, Kakizaki H, Tajino T, et al. Imaging characteristics of deep-seated lipomatous tumors: Intramuscular lipoma, intermuscular lipoma, and lipoma-like liposarcoma. J Orthop Sci [Internet]. 2007

ISSN (Online): 2565-4810; (Print): 2565-4802 | ICV 2019: 98.48 |

11. Kransdorf MJ, Bancroft LW, Peterson JJ, Murphey MD, Foster WC, Temple HT. Imaging of fatty tumors: Distinction of lipoma and well-differentiated liposarcoma. Radiology [Internet]. 2002;224(1):99–104.
12. Fritchie K, Ghosh T, Graham RP, Roden AC, Schembri-Wismayer D, Folpe A, et al. Well-Differentiated/Dedifferentiated Liposarcoma Arising in the Upper Aerodigestive Tract: 8 Cases Mimicking Non-adipocytic Lesions. Head Neck Pathol 2020;14(4):974–81.
13. Shi HY, Wei LX, Wang HT, Sun L. Clinicopathological features of atypical lipomatous tumors of the laryngopharynx. J Zhejiang Univ Sci B 2010

Source of Support: Nil; **Conflict of Interest:** None

Submitted: 19-12-2020; **Accepted:** 24-01-2021; **Published online**: 17-03-2021