

Case report

Inflammatory leiomyosarcoma of the head and neck: Case report

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ABSTRACT

Introduction and importance: Primary sarcomas in the head and neck region are rare. Inflammatory leiomyosarcoma was first described in 1995. The case reported herein is the first reported inflammatory leiomyosarcoma occurring in the head and neck.

Presentation of case: A 37-year-old male presented with a long history of an asymptomatic slowly enlarging neck mass. Examination revealed a firm mass in the lower third of the right sternocleidomastoid muscle. Computerized tomography and magnetic resonance imaging showed a lobulated, well-circumscribed tumour with malignant features. A wide local excision was performed and histopathological examination confirmed an inflammatory leiomyosarcoma.

Discussion: Inflammatory leiomyosarcoma is a recently described peculiar soft tissue tumour with histological features overlapping conventional leiomyosarcoma, and dense lymphocytic inflammation and immunohistochemical reactivity for both smooth and skeletal muscle markers. These are indolent tumours and wide local excision is curative.

Conclusion: This case highlights the importance of considering primary sarcomas in the differential diagnosis of asymptomatic head and neck masses.

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1. Introduction

The differential diagnosis of asymptomatic head and neck masses is broad and includes a wide range of inflammatory, benign and malignant conditions. Sarcomas account for only 1% of primary neoplasms occurring in the head and neck region, of which leiomyosarcomas account for approximately 4% [1]. Although inflammatory leiomyosarcoma (ILMS) was first described in 1995 [2], it was only recognised as a distinct entity by the WHO in 2020.

This case highlights the importance of considering non-epithelial neoplasms in the differential diagnosis when faced with an asymptomatic, slowly enlarging neck mass. Although uncommon, primary sarcomas should not be forgotten since they can cause fatal compressive complications. The case reported herein is the first reported inflammatory leiomyosarcoma occurring in the head and neck. Ethics approval has been obtained from the University of Cape Town, Human Research Ethics Committee. This case has been reported in line with the SCARE criteria [3].

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2. Case report

A 37-year-old male presented with a 4-year history of an asymptomatic lump in the right lower neck. His past medical history was unremarkable and he had no previous trauma or skin related malignancies. The patient was not on any chronic medication, and there was no family history of cancer. Clinical examination revealed a hard mass in the inferior third of the right sternocleidomastoid muscle. A fine needle aspiration was performed which revealed atypical spindle cells suspicious for malignancy.

A core biopsy showed a low grade spindle cell neoplasm with a fascicular growth pattern and moderate atypia. An initial diagnosis of low grade myofibroblastic sarcoma was favoured. Further investigations to determine tumour extent were performed. This included computerized tomography (CT) of the head and neck which showed a 44 × 27 × 24 mm well defined homogeneously enhancing mass in the right supraclavicular region at the inferior level of the thyroid gland. On magnetic resonance imaging (MRI) the mass appeared lobulated, mildly hyperintense on T1 and markedly hypertense on T2. Diffuse postcontrast enhancement and moderate restricted diffusion were seen. The tumour was confined to the SCM without evidence of locoregional spread. Chest X-ray was unremarkable.

A wide local excision of the mass with a liberal cuff of sternocleidomastoid muscle was performed by the third author (JJF), with the second author (GV) assisting during the procedure. Macroscopic examination of the resected specimen revealed a sharply circumscribed and encapsulated white to yellow intramuscular tumour with a whorled cut surface. Microscopic examination showed a well circumscribed spindle cell neoplasm surrounded by a thick fibrous capsule. The neoplastic cells were arranged in short fascicles with intermixed lymphocytes and collections of xanthoma cells. Moderate pleomorphism with nuclear hyperchromasia and conspicuous nucleoli were seen. The tumour cells diffusely expressed desmin and smooth actin. There was focal expression of myogenin in tumour cells. The following stains were negative: S100, AE1/AE3 (cytokeratin) and ALK1. The EBER in-situ hybridisation was also negative. Complete surgical excision with clear margins was achieved. As excision with clear surgical margins is currently the preferred management of these tumours, the patient did not receive adjuvant chemoradiation.

3. Discussion

Inflammatory leiomyosarcoma (ILMS) was first described as a distinct entity in 1995 by Merchant et al. [2]. Further research showed that these are rare soft tissue tumours with a distinct immunophenotype and genetic profile. Intramuscular sites of the back and lower limbs are the most common sites of occurrence [4]. They have an indolent behaviour, and the clinical presentation is often simply an enlarging soft tissue mass. These tumours generally have a favourable clinical outcome, although there is one case report of distant metastasis [5].

The histological picture is characterized by an encapsulated spindle cell tumour with nuclear pleomorphism, interspersed chronic inflammatory cells and aggregates of xanthoma cells. Recently it has been reported that it expresses both smooth and skeletal muscle markers indicating an origin from primitive myogenic precursors; these authors proposed naming it a low-grade inflammatory myogenic tumour [4].

Inflammatory leiomyosarcoma has not been reported in the head and neck region, although a case of "histiocyte-rich rhabdomyoblastic tumour" has been reported in the parapharyngeal space [6]. This tumour apparently shares morphological features with inflammatory leiomyosarcoma and the authors proposed that such lesions be reclassified as inflammatory rhabdomyoblastic tumours [6]. It is clear that the classification and nomenclature for this tumour requires further investigation and discussion.

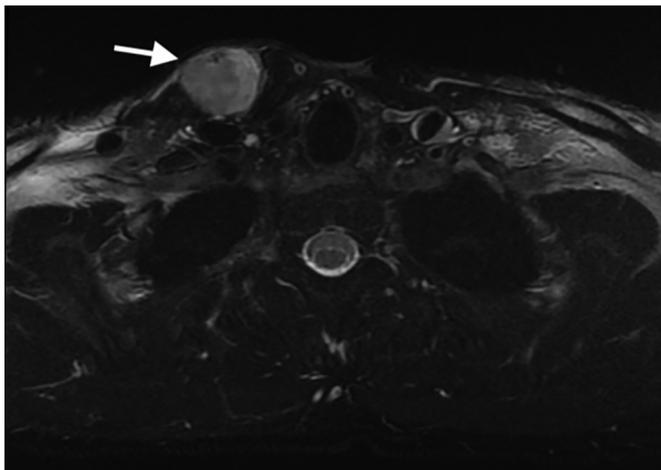


Fig. 1. MRI indicated a well-circumscribed, hyperintense lesion (white arrow) within the inferior aspect of the right sternocleidomastoid muscle.



Fig. 2. Wide local excision of the mass with a liberal cuff of sternocleidomastoid muscle.

In our case the differential diagnoses considered for a spindle cell sarcoma expressing desmin and smooth muscle actin included low grade myofibroblastic sarcoma, inflammatory myofibroblastic tumour and EBV-related smooth muscle tumour. The focal expression of myogenin and the collections of foam cells are useful diagnostic features of inflammatory leiomyosarcoma.

Although fine needle aspiration of ILMS is non-specific, it is the preferred initial investigation to direct further workup and management. A core biopsy of the lesion is essential to make a reliable and accurate diagnosis prior to surgery [4].

Wide local excision of ILMS with clear resection margins is reported to be curative. Large ILMS can be fatal if vital organs are compressed [4].

4. Conclusion

This case highlights the importance of considering primary sarcoma as a cause of an asymptomatic mass in the head and neck, and is the first reported case of ILMS in the head and neck region (Figs. 1–5).

Provenance and peer review

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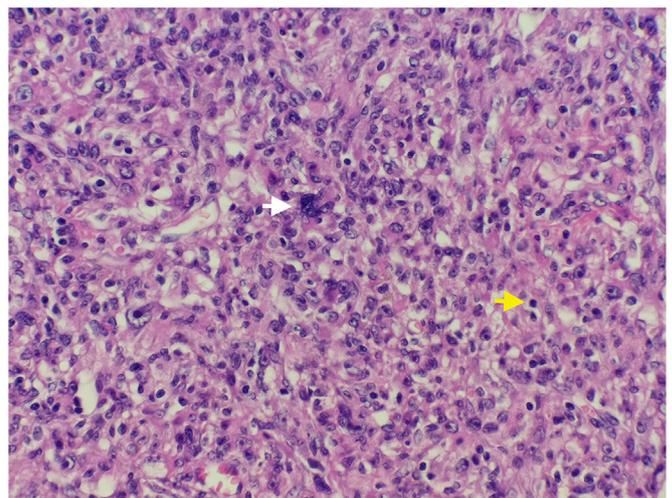


Fig. 3. The H&E (hematoxylin and eosin) stain demonstrates moderately atypical spindle cells (white arrow) arranged in short fascicles with admixed lymphocytes (yellow arrow) and xanthoma cells. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

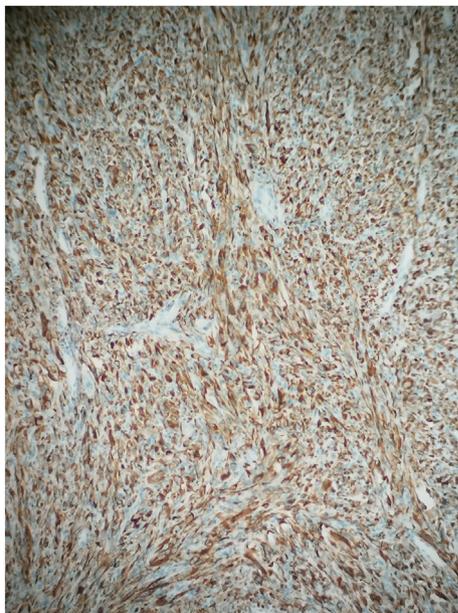


Fig. 4. Smooth muscle actin was diffusely positive in the atypical spindle cells.

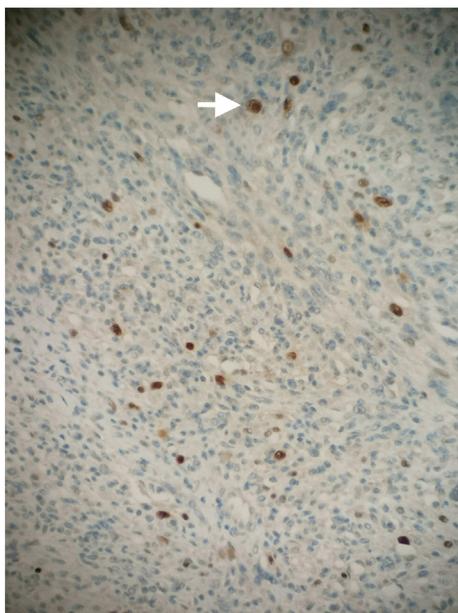


Fig. 5. Myogenin was focally expressed in the nuclei of the neoplastic cells (white arrow).

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethics approval

Ethics approval obtained from University of Cape Town, Human Research Ethics Committee.
HREC Ref: 186/2021.

Registration of research studies

None.

Guarantor

Gerrit Viljoen.

CRediT authorship contribution statement

N. Viljoen - Concept, design, writing the paper.
G. Viljoen - Design, writing the paper.
Fagan - Concept, design, writing the paper.
Govender - Concept, design, writing the paper.

Declaration of competing interest

The authors declare no conflict of interest.

References

- [1] J. Yadav, J. Bakshi, M. Chouhan, R. Modi, Head and neck leiomyosarcoma, *Indian J. Otolaryngol. Head Neck Surg.* 65 (1) (2013) 1–5, <https://doi.org/10.1007/s12070-011-0305-8>.
- [2] W. Merchant, E. Calonje, C.D.M. Fletcher, Inflammatory leiomyosarcoma: a morphological subgroup within the heterogeneous family of so-called inflammatory malignant fibrous histiocytoma, *Histopathology.* 27 (6) (1995) 525–532, <https://doi.org/10.1111/j.1365-2559.1995.tb00323.x>.
- [3] R.A. Agha, T. Franchi, C. Sohrabi, et al., The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.
- [4] M. Michal, B.P. Rubin, D.V. Kazakov, et al., Inflammatory leiomyosarcoma shows frequent co-expression of smooth and skeletal muscle markers supporting a primitive myogenic phenotype: a report of 9 cases with a proposal for reclassification as low-grade inflammatory myogenic tumor, *Virchows Arch.* 477 (2) (2020) 219–230, <https://doi.org/10.1007/s00428-020-02774-z>.
- [5] A. Chang, S.M. Schuetze, E.U. Conrad III, K.L. Swisshelm, T.H. Norwood, B.P. Rubin, So-called “inflammatory leiomyosarcoma”: a series of 3 cases providing additional insights into a rare entity, *Int. J. Surg. Pathol.* 13 (2) (2005) 185–195.
- [6] J.M. Cloutier, G.W. Charville, F. Mertens, et al., “Inflammatory Leiomyosarcoma” and “Histiocyte-rich Rhabdomyoblastic tumor”: a clinicopathological, immunohistochemical and genetic study of 13 cases, with a proposal for reclassification as “inflammatory Rhabdomyoblastic tumor.”, *Mod. Pathol.* 34 (4) (2021) 758–769, <https://doi.org/10.1038/s41379-020-00703-8>.