CASE REPORT

Leiomyosarcoma: a rare sinonasal malignancy

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SUMMARY

Leiomyosarcomas (LMS) are rare sinonasal malignant tumours with 20 cases of nasal LMS previously reported in the literature. An 81-year-old man presented with an incidental left nasal lesion associated with left nasal obstruction and occasional left-sided blood-stained rhinorrhoea. Nasendoscopic examination showed green coloured polyps filling the left nasal cavity with biopsies showing LMS. CT imaging did not show any bony erosion. No metastases were found on positron emission tomography imaging. The patient underwent endoscopic resection and all surgical margins were clear of disease. The patient was declined postoperative radiotherapy, and serial follow-up has not shown any recurrence to date. Nasal LMS are rare sinonasal smooth muscle tumours and symptoms and radiological findings are often non-specific. Therefore, diagnosis is made on histopathological and immunohistochemical analysis. Due to their invasive nature, the treatment of choice is complete surgical resection, with adjuvant therapy reserved for patients with locally advanced or metastatic disease.

BACKGROUND

Leiomyosarcomas (LMS) are malignant tumours of smooth muscle which are commonly found in the uterus and intestinal tract. Although LMS can occur in the sinonasal region, they make up only less than 3% of all non-epithelial sinonasal tumours¹ and less than 10% of all sarcomas are found in the head and neck region.² Its rarity had been postulated to be due to the minimal amount of smooth muscle in the sinonasal region, which is mostly located in the tunica media of blood vessels.³ Prompt diagnosis and treatment is often difficult to achieve due to its rarity and a lack of specific symptom as well as clinical and radiological findings. Patients therefore often present late with more advanced disease, and hence nasal LMS is associated with poorer prognosis compared with LMS from other regions. We present a case of nasal LMS arising from the inferior turbinate. Theories of pathogenesis and management strategies will be explored.

CASE PRESENTATION

An 81-year-old man presented with an incidental left nasal lesion found during investigations for recurrent falls. On questioning, this was associated with intermittent nasal obstruction, epiphora and occasional left-sided blood-stained rhinorrhoea, which had been present for a few years. He has not had any previous sinonasal symptoms or sinonasal surgery and his active medical issues

include ischaemic heart disease, hypertension, type 2 diabetes, gastro-oesophageal reflux disease and recurrent pulmonary emboli. There was no associated family history of sinonasal diseases or occupational carcinogenic exposure such as wood dust or textiles. Flexible nasendoscopic examination showed a green coloured polypoidal lesion with clear mucous filling the entire left nasal cavity with an unremarkable right nasal cavity. The discolouration was likely related to an overlying infection; however, no specific bacteria was isolated on microscopy and culture. He underwent rigid endoscopic biopsy of the polyp under general anaesthetic which was suspicious for LMS. Clear mucus was found in the maxillary sinus without any tumour extension into the paranasal sinuses.

INVESTIGATIONS

CT imaging showed a soft tissue mass filling the left nasal cavity, extending through the posterior choana to abut the posterior pharyngeal wall ([figure 1A, B](#)). The lateral nasal wall was slightly bowed, but did not show any bony erosion or extension into the paranasal sinuses. No nodal or distant metastases were found on positron emission tomography imaging. The left maxillary sinus opacity was found to be clear mucus on rigid endoscopy.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses include benign polyps such as inflammatory nasal polyps and inverting papillomas. Malignant differential diagnoses include sarcomas such as rhabdomyosarcomas, fibrosarcomas, liposarcomas, angiosarcomas and osteosarcomas, as well as adenocarcinomas, squamous cell carcinomas and mucoepidermoid carcinomas.

TREATMENT

The patient underwent endoscopic resection with medial maxillectomy with resection of the uncinate, lateral nasal wall, inferior turbinate and preservation of the lacrimal bone and nasolacrimal duct. The tumour was found to be originating from the left inferior turbinate. Histopathology showed moderate to markedly pleomorphic spindle cells with enlarged irregular hyperchromatic nuclei ([figure 2](#)) which, on immunohistochemistry, was strongly positive for desmin ([figure 3](#)) and smooth muscle actin ([figure 4](#)), and negative for myogenin, cytokeratin MNF116, AE1/AE3 and S100/Melan-A. The proliferative marker Ki-67 showed that approximately 30%–40% of cells were positive. This is consistent with LMS. All surgical margins were clear of disease.

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Figure 1 CT imaging showing a polypoid lesion filling the left nasal cavity.

OUTCOME AND FOLLOW-UP

Although the patient did not have advanced or metastatic disease, due to the aggressive nature of this condition, the results were discussed at the unit head and neck cancer multidisciplinary meeting with the recommendation of offering postoperative radiotherapy. However, this was declined by the patient, and three monthly follow-up with nasendoscopic examination had not shown any recurrence to date (9 months).

DISCUSSION

Nasal LMS are rare sinonasal smooth muscle tumours with a current literature search showing 20 cases reported in the literature.¹⁻⁵ The first case of nasal LMS was described by Dobben in 1958 in a 69-year-old woman presenting with LMS at the floor of the posterior choanae.³ Reported cases show a mean age of onset of 53 and a slight predominance in males.² Although there is no agreed theory on pathogenesis, they are thought to arise from the smooth muscle in the tunica media of the blood vessels in the nasal cavity or from undifferentiated mesenchymal tissue.^{4,6} The scarcity of smooth muscle in the nasal cavities may explain the low incidence of this condition. Several predisposing risk factors had been postulated for LMS, including prior exposure to radiation, as well as exposure to chemotherapy such as cyclophosphamide.⁷⁻¹⁰ Although occupational exposure to wood, leather and textile dust as well as formaldehyde, chlorophenol and cutting oils had been associated with sinonasal malignancies and LMS,^{11,12} the patient in this report has not had any such exposure in his various employments.

Figure 2 H&E stain at 400× magnification showing spindle cell neoplasm composed of cells in orderly broad fascicles with cigar-shaped nuclei.

Figure 3 Positive immunohistochemical staining for desmin at 400× magnification.

The diagnosis of nasal LMS is often difficult as patients often present late due to the non-specific symptoms as well as clinical and radiological findings. These symptoms include unilateral or bilateral nasal obstruction, rhinorrhoea, epistaxis, facial pressure, pain, postnasal drip or a reduction or loss in smell sensation.⁵ Macroscopic clinical examination findings on nasendoscopy are also non-specific but include grey-coloured unilateral polypoid or pedunculated sinonasal lesions, which may have additional features such as necrosis, ulceration or haemorrhage.^{4,7} Vascular metastases are often more common than lymph node metastases,⁴ but patients presenting with distant metastases had also been reported.¹³

Likewise, imaging findings on CT are often non-specific and include expansile cystic or necrotic heterogenous lesions of soft tissue density.¹⁴ Similarly, nasal LMS show only moderate hyperintensity on both T1 and T2 weighted MRI which does not provide additional diagnostic information.¹⁴ Imaging therefore may be more helpful in surgical planning than for the diagnosis of nasal LMS.

Similar to other smooth muscle tumours, histopathological findings include interlacing spindle cells with eosinophilic cytoplasm and hyperchromatic irregularly shaped nuclei.⁷ Therefore, immunohistochemical staining is crucial in the differentiation of

Figure 4 Positive immunohistochemical staining for smooth muscle actin at 400× magnification.

nasal LMS from other tumours such as rhabdomyosarcomas, melanomas and nerve sheath tumours. Nasal LMS stain positive for smooth muscle actin and desmin and negative for myogenin, cytokeratin MNF116 and AE1/AE3 as well as S100/Melan-A.^{4 15}

Because of their invasive nature, the treatment of choice is complete surgical resection, with adjuvant chemotherapy or radiotherapy reserved for patients with locally advanced, recurrent or metastatic disease.^{3 7} Late recurrence is uncommon for all subsites of LMS¹⁶ and Papoian *et al* reported a case of recurrent nasal LMS 4 years after treatment.¹⁷ Prognosis is often poor due to the late presentation because of the lack of specific symptoms. Moreover, achieving wide surgical margins for locally advanced disease is often difficult in the sinonasal region.¹⁷ Papoian *et al* reports a case of recurrent nasal LMS despite clear surgical clearance with postoperative adjuvant radiotherapy.¹⁷ Several factors associated with poor prognosis had been identified, including the size of the tumour, higher histological grading and positive surgical margins.¹⁸ Wu *et al* reports a 5-year survival of 88% for patients with well-differentiated tumours, which falls to 53% for poorly differentiated tumours.¹⁹

Due to its rarity, nasal LMS needs to be considered when investigating sinonasal lesions. We would recommend careful consideration of clinical symptoms, as well as radiological and histopathological findings in a multidisciplinary setting with surgical resection as the first-line treatment modality to maximise the outcome of patients with nasal LMS.

Learning points

- ▶ Nasal leiomyosarcomas (LMS) are rare malignant nasal cavity tumours.
- ▶ Symptoms, clinical and radiological findings are often non-specific and may not help in diagnosis.
- ▶ Histopathology is crucial in the diagnosis of nasal LMS.
- ▶ Complete surgical excision is the recommended first-line treatment modality.
- ▶ Prognosis is often poor due to late presentation and difficulty in achieving wide surgical margins.

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