CASE REPORT

Leiomyosarcoma of the maxilla in a 24-year-old man who initially presented with odontalgia, and suffered from tumour mismanagement

Shokouh Taghipour Zahir,^{1,2} Naser Sefidrokh Sharahjin³

¹Department of Pathology, Shahid Sadoughi University of Medical Sciences, Yazd, Islamic Republic of Iran

²Department of Pathology, Mortaz General Hospital, Yazd, Islamic Republic of Iran

³Department of Radiology, Shahid Sadoughi University of Medical Sciences, Yazd, Islamic Republic of Iran

Correspondence to
Professor Shokouh Taghipour Zahir,
shokouh_zahir@yahoo.com

SUMMARY

Leiomyosarcoma is an uncommon malignant soft tissue tumour originating from smooth muscle cells. It generally occurs in the female genital tract, especially in the uterus, gastrointestinal tract, retroperitoneum and rarely in the head and neck. Here we report a case of leiomyosarcoma in a 24-year-old man with a 6-month history of toothache and extraction of three right upper molar teeth, that was complicated by fistula tract formation and facial swelling. Imaging studies demonstrated tumoural lesions involving the right maxillary sinus, nasal cavity, hard palate and orbit. The histopathological and immunohistochemical studies confirmed that it was leiomyosarcoma.

BACKGROUND

Leiomyosarcoma is an unusual soft tissue sarcoma mostly affecting the uterus, gastrointestinal (GI) tract and retroperitoneum.^{1–10} Head and neck involvement is rare, but a few cases have been reported in the regions of the nasal cavity, paranasal sinuses and orbit, respectively.^{2–4} The origin of these types of sarcoma may be smooth muscle cells located in the skin tissue such as the pilorector muscles, or in the wall of blood vessels.^{1–13} Primitive mesenchymal and myoepithelial cells of salivary glands, paranasal sinuses, scalp, nasopharynx, tongue and larynx, have the ability to become sarcomatous cells. Leiomyosarcoma of the head and neck may present as a slow growing tumour with symptoms such as epistaxis, pain, maxillary swelling, nasal obstruction and visual disturbances.^{1–13} We report a case of maxillary sinus leiomyosarcoma for its unusual presentation, mismanagement of the patient, delay in diagnosis and complications, followed by a brief discussion of its histopathological and radiological characteristics.

CASE PRESENTATION

A 24-year-old man was admitted to the otorhinolaryngology clinic with a history of weight loss, tinnitus, loss of appetite and serial extraction of his first, second and third right molar teeth, due to pain in the right upper maxilla without filling defects, for a period of 6 months. Recurrent hemifacial swelling and reduction in its size were noted by the patient. He also explained that whenever he pressed the extracted teeth region, a significant amount of retained pus was excreted from his mouth, and some necrotic soft tissues were expelled from his nasal cavity, due to a fistula tract.

The patient underwent fistula tract excisional biopsy and the histopathological result was actinomycosis. One month later, he came with decreased vision of his right eye, hemifacial paresthesia and also complete obstruction of the right nasal cavity.

The patient did not have a medical history of diabetes mellitus or cigarette smoking.

INVESTIGATIONS

During a physical examination, conjunctivitis of the right eye, a large defect in the right alveolar ridge and a mobile non-tender lymph node in the right side of neck, level II, were detected. A CT scan study on axial sections of the paranasal sinuses demonstrated, an ill-defined expansile, lobulated isodense large soft tissue mass lesion, measuring 5 cm in diameter, within the right maxillary sinus (figure 1). Destructive changes of the adjacent bony structures in particular, hard palate, walls of the right maxillary sinus, pterygoid, palatine and extension of the lesion to the soft tissue of the right masticator space, right parapharyngeal space, apex of the right orbit and skull, were also depicted. All findings could have been suggestive of a neoplastic lesion, or less likely a fungal infection, in addition, retained secretion in the right maxillary, ethmoid and sphenoid sinuses were seen. Extension of the lesion to the right nasal cavity showed complete obstruction of the right nasal cavity, maxillary ostium and right middle meatus. Brain MRI, with and without contrast, using axial, sagittal and

Figure 1 The CT scan shows an ill-defined expansile and lobulated isodense large soft tissue mass lesion, within the right maxillary with destruction of the bony structures.

CrossMark

To cite: Taghipour Zahir S, Sharahjin NS. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-200933

Figure 2 T1-weighted MRI following intravenous administration of gadolinium demonstrates heterogeneous enhancement and extension of the lesion to the right parapharyngeal space, right masticator space, right middle cranial fossa and right cavernous sinus.

coronal on T1-weighted images (T1WI), T2WI, proton density and fluid-attenuated inversion recovery showed a heterogeneous mass-like lesion in the right maxillary sinus and extension of the lesion beyond its walls. T1-weighted MRI following intravenous administration of gadolinium presented heterogeneous enhancement and extension of the lesion to the right parapharyngeal space, right masticator space, right middle cranial fossa and right cavernous sinus (figure 2). Mucosal thickening of the right mastoid air cells and left maxillary, sphenoid sinuses retention cysts were also reported. Grey and white matter signals, cerebral ventricles, major intracranial vascular structures, basal ganglia and brain stem were unremarkable. Based on MRI and CT scan findings the diagnosis was most likely suggestive of an adenoid cystic carcinoma, however, other differential diagnosis such as fungal infections were also considered. The patient underwent oroantral fistulectomy with marsupialisation of the retention cyst and excision of the right maxillary sinus polypoid lesion. A right anterior ethmoidectomy, along with a left and right

maxillary sinus antrostomy, were also arranged for the patient. As the tumour extended widely and seemed to be unresectable, no other surgical resection was performed. The specimen received from the maxillary lesion consisted of multiple pieces of greyish coloured tissues measuring $1.5 \times 2 \times 1$ cm. Histopathological examination showed a tumoural lesion composed of spindle cells with cigar-shaped nuclei arranged in fascicles and interlacing bundles with large areas of coagulative necrosis. Some of the neoplastic cells had large hyperchromatic nuclei and infiltration of inflammatory cells between them was also seen (figure 3A,B). Neoplastic spindle cells had a positive reaction to vimentin, α smooth muscle actin and caldesmon, but had negative results for pan cytokeratin, epithelial membrane antigen (EMA), desmin and CD34, in immunohistochemical (IHC) studies (figure 4A,B). Based on H&E staining with IHC findings, leiomyosarcoma was confirmed. According to *The American Joint Committee on Cancer Staging System*, the tumour was in stages G3, Td2, N0 and M0.

DIFFERENTIAL DIAGNOSIS

In a histopathological differential diagnosis of the maxillary sinus spindle cell tumours, leiomyosarcoma, fibrosarcoma, synovial sarcoma, malignant peripheral nerve sheath tumour, sarcomatoid carcinoma, metastatic lesions and fungal infections, might be considered. Imaging differential diagnosis included squamous cell carcinoma, adenocarcinoma, undifferentiated carcinoma, metastatic lesions and fungal infections. For a definitive diagnosis and differentiation of spindle cell sarcoma, IHC studies composed of CD34, vimentin, α smooth muscle actin, desmin, CK7, CK20, EMA, pan-cytokeratin, and caldesmon, should be examined. Neoplastic cells in leiomyosarcoma have a positive reaction to caldesmon, α smooth muscle actin, vimentin, and some of them also react with desmin. Fungal infections could be ruled out with a negative reaction to the periodic acid-Schiff staining method.

TREATMENT

Extension of the tumour towards the base of the skull and involvement of the right orbit prevented resection of the lesion, so the patient opted for chemotherapy. He received three courses of chemotherapy (1) intravenous injection of Adriamycin 35 mg + 100 cc NS, given over a 3-day continuous infusion (days 1, 2 and 3); (2) intravenous injection of ifosfamide 350 mg + 500 cc NS, given over a 3-day continuous infusion (days 1–3);

Figure 3 (A) Tumoural lesion composed of spindle cells arranged in a whorled pattern with inflammatory cells between them (H&E, objective $\times 20$). (B) Some of the tumoural cells have large hyperchromatic nuclei with mitotic figures (H&E, objective $\times 20$).

Figure 4 (A) By immunohistochemistry, neoplastic cells were positive for vimentin (objective $\times 20$). (B) Neoplastic cells strongly positive for α smooth muscle actin (objective $\times 20$).

(3) intravenous injection of Mesna (0–4–8), (400–800–800), given over a 3-day continuous infusion (days 1–3); (4) intravenous injection of dacarbazine 500mg+500 cc NS, given over a 3-day continuous infusion (days 1–3) and afterwards a head and neck MRI was carried out to estimate his response to chemotherapy. The tumour did not completely respond to chemotherapy, but reduced slightly in size, so radiotherapy was recommended. At the present time, he has undergone radiotherapy and subsequent treatment planning will involve surgical resection.

OUTCOME AND FOLLOW-UP

After 4 months his general condition, facial swelling and the vision in his right eye had improved.

DISCUSSION

Leiomyosarcoma belongs to the heterogeneous group of cancers which originate from smooth muscle cells located in the uterus, GI tract or other soft tissues, such as the retroperitoneum.^{1–13} It is a rare soft tissue tumour with aggressive behaviour and accounts for approximately 5–10% of all soft tissue sarcomas, with only 3% of these tumours occurring in the head and neck.^{5–10} Treatment of this type of lesion is difficult because of high rates of recurrence, metastatic rates and poor prognosis.^{1–13} Leiomyosarcoma of the soft tissue often presents as an enlarging, painless, slow growing mass and a variety of symptoms may appear based on its location.³ In the head and neck especially when maxillary or other paranasal sinuses are involved, common presentations may include nasal obstruction, epistaxis, local pain or facial swelling.^{2–4} Tooth complications have been rarely described, however, our case presented with unusual serial toothaches in addition to loosening of the teeth that led to their extraction with complications of nasal obstruction, swelling and epistaxis. Hideki *et al* reported a case of gingival leiomyosarcoma which was similar to our case.⁴ Unusual presentation of the salient lesion also caused a delay in diagnosis of the patient's disorder. In the differential diagnosis of leiomyosarcoma other spindle cell sarcomas such as fibrosarcoma, malignant peripheral nerve sheath tumours, synovial sarcoma and malignant melanoma (spindle cell type) should be considered, and IHC studies need to be conducted. The efficacy of chemotherapy and radiotherapy was not clearly described in literature.^{1–13} In some reported cases, it seems that wide surgical resection with chemotherapy could control the disorder, but definitive treatment guidelines were not published.¹ Our patient was mismanaged

because prior to the diagnosis through imaging by the radiologist, three of the his molar teeth were extracted by the dentist after which a fistula tract was formed, therefore treatment was delayed. The patient initially received three courses of chemotherapy and then radiotherapy was considered to reduce the size and extent of the tumour before surgery. The general medical condition of the patient was satisfactory and both facial swelling and right eye vision improved after 4 months.

Learning points

- ▶ Although leiomyosarcoma of the head and neck is a rare malignancy, it should be considered in a differential diagnosis of head and neck mass lesions.
- ▶ Leiomyosarcoma in the maxilla might present as a nasal obstruction, pain or epistaxis, but an unusual presentation such as odontalgia that does not resolve itself by tooth extraction is evidence of a problem, therefore it is wise to rule out an underlying cause by imaging.
- ▶ In young adults, loss of visual acuity without a history of trauma or diabetic mellitus, should be considered as a serious symptom and a comprehensive workup should be performed.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Chew YK, Noorizan Y, Khir A, *et al*. Leiomyosarcoma of the maxillary sinus. *Med J Malaysia* 2009;64:174–5.
- 2 Prasad KC, Alva TB, Khadilkar U, *et al*. Leiomyosarcoma of the maxillary sinuses: report of two cases. *Ear Nose Throat J* 2004;83:122–6.
- 3 Montgomery E, Goldblum JR, Fisher C. Leiomyosarcoma of the head and neck: a clinicopathological study. *Histopathology* 2002;40:518–25.
- 4 Lo Muzio L, Favia G, Farronato G, *et al*. Primary gingival leiomyosarcoma. A clinicopathological study of 1 case with prolonged survival. *J Clin Periodontol* 2002;29:182–7.
- 5 Keck T, Mattfeldt T, Kühnemann S. Leiomyosarcoma of the ethmoidal cells. *Rhinology* 2001;39:115–17.
- 6 Sumida T, Hamakawa H, Otsuka K, *et al*. Leiomyosarcoma of the maxillary sinus with cervical lymph node metastasis. *J Oral Maxillofac Surg* 2001;59:568–71.

- 7 Tanaka H, Westesson PL, Wilbur DC. Leiomyosarcoma of the maxillary sinus: CT and MRI findings. *Br J Radiol* 1998;71:221–4.
- 8 Ohashi Y, Nakai Y, Muraoka M, *et al.* Asymptomatic leiomyosarcoma of maxillary sinus accompanied by primary mucocele. *Arch Otorhinolaryngol* 1984;240:73–8.
- 9 Richter HJ, Steinert W, Mahn B, *et al.* Leiomyosarcoma of the nasal cavity and paranasal sinuses. *HNO* 1981;29:17–21.
- 10 Weitzner S. Leiomyosarcoma of the anterior maxillary alveolar ridge. *Oral Surg Oral Med Oral Pathol* 1980;50:62–4.
- 11 Yamaguchi S, Nagasawa H, Suzuki T, *et al.* Sarcomas of the oral and maxillofacial region: a review of 32 cases in 25years. *Clin Oral Investig* 2004;8:52–5.
- 12 Eppsteiner RW, DeYoung BR, Milhem MM, *et al.* Leiomyosarcoma of the Head and Neck: a population-based analysis. *Arch Otolaryngol Head Neck Surg* 2011;137:921–4.
- 13 Shruti D, Ashwani S, Ishwar S, *et al.* Leiomyosarcoma of the maxillary sinus: an unusual site of occurrence. *Internet J Head Neck Surg* 2008;2:10.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow