CASE REPORT

Leiomyosarcoma of the Maxillary Sinus

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SUMMARY

Leiomyosarcoma is a malignant smooth-muscle tumour that has a predilection for the gastrointestinal and female genital tract. It is locally fast-spreading and highly aggressive, and the prognosis is poor. We report a rare case of leiomyosarcoma of the maxilla in a patient who sought treatment for maxillary swelling, nasal obstruction with no epistaxis, orbital involvement or cervical lymph node metastasis. The patient underwent subtotal maxillectomy followed by radiotherapy. At present, he is symptom free with no recurrence and under regular follow up.

KEY WORDS:
Leiomyosarcoma, Maxillary sinus

INTRODUCTION

Smooth-muscle tumours are rare in the head and neck. Most arise in the gastrointestinal and female genital tract because of the preponderance of smooth muscle at these sites[1]. Leiomyosarcomas of the head and neck are believed to originate in the tunica media of the blood vessels or in pluripotential mesenchymal cells. Clinically, these tumors are very aggressive, and the prognosis is poor. Leiomyosarcomas of the paranasal sinuses are rare and distinct entities[2]. We report a rare case of leiomyosarcoma of the maxillary sinus.

CASE REPORT

A 36 year-old man sought treatment for a 3-month history of painful swelling over the left maxillary area that was accompanied by nasal obstruction. There was no epistaxis, proptosis, and loose teeth.

Clinical examination revealed a diffuse swelling measuring 5 x 4 cm on the left maxilla. The tumour extended inferiorly to the angle of the mouth, laterally to the front of the tragus, and medially to the dorsum of the nose. It obliterated the nasolabial fold and involved the skin at the root of the nose. The skin over the swelling appeared to be stretched. On anterior rhinoscopy, the mass could be seen filling the entire nasal cavity. Findings on postnasal examination were normal. Clinical examination of the neck revealed no enlarged neck nodes.

A clinical diagnosis of a malignancy of the left maxilla was made. Computed tomography (CT) revealed a large soft-tissue density in the left maxillary sinus measuring 3.4 x 3.3cm and a soft tissue mass measuring 2.7 x 2.9 that had destroyed all of the maxillary walls except the posterior wall with involvement of the orbit. There was no intracranial extension. A biopsy from hard palate was taken under local anaesthesia which was reported as inconclusive. The patient underwent subtotal maxillectomy and skin was preserved. The surgical wound was closed primarily without any flap. An obturator plate was placed at the defect of the hard palate. The patient was then referred to a oncologist and radiotherapy treatment was commenced. The patient is currently well and under regular follow up for the past three years. Repeated CT scan and nasal endoscopy on post operative follow up revealed normal mucosa with no sign of recurrence.

Analysis of the biopsy specimen revealed that the tumour tissue comprised of fascicles of spindle-shaped cells with eosinophilic cytoplasm and oval to elongated and blunt-ended vesicular nuclei. Mitosis was conspicuous. Areas of necrosis, fibrosis, hyalinization, and inflammatory infiltrates were seen. The reticulin stain showed abundant reticulin fibers around individual tumour cells. The van Gieson's and Masson's trichrome stains showed smooth-muscle differentiation in the spindle cells. The tumour cells showed strong vimentin and smooth muscle actin (SM A) stains. These features were suggestive of leiomyosarcoma.

DISCUSSION

Leiomyosarcomas account for 6.5% of all soft-tissue sarcomas, and only 3% of them arise in the head and neck[3]. When they occur in the sinonasal tract, the most common sites are the nasal cavity, the maxillary sinus, and the ethmoid sinus, in that order. Therefore, this patient is a rare case of leiomyosarcoma of left maxillary sinus.

Leiomyosarcomas of the sinonasal tract are more common in men than in women. The average age of diagnosis is 50 years Initial symptoms in order of decreasing frequency include nasal obstruction, epistaxis, facial pain, and facial swelling. Sinonasal tract leiomyosarcoma is characterized as locally aggressive with a low metastatic potential[3]. Our patient presented with nasal obstruction and facial swelling with absence of epistaxis and locally invasive features.

On CT, leiomyosarcomas appear as bulky masses, and are frequently associated with extensive necrotic or cystic changes. These tumours do not contain calcifications, but they do cause frank bony destruction, as occurred in our patient[3]. Magnetic Resonance Imaging (MRI) was not employed in this case due to the unavailability of the service in this centre.

This article was accepted: 13 April 2009
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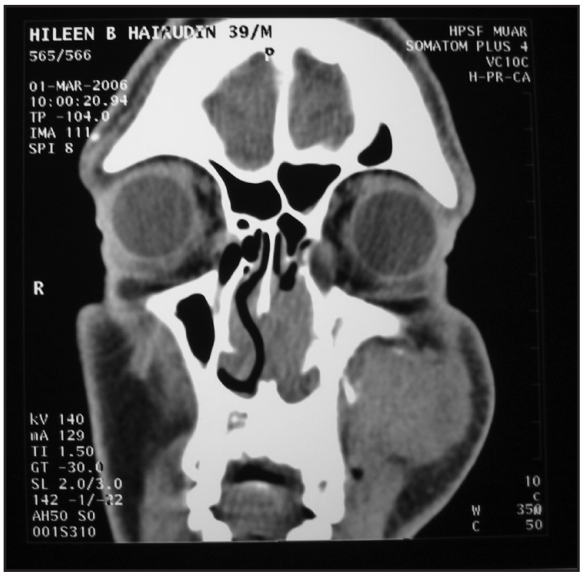

Fig. 1: CT scan coronal view showing the tumour involving the left maxillary sinus

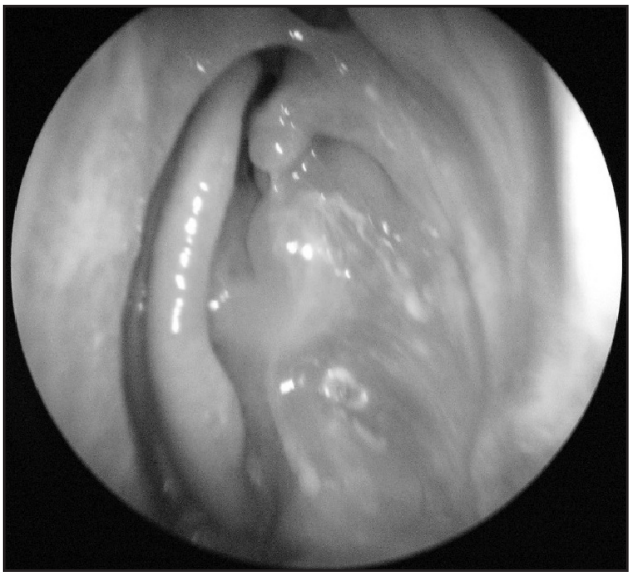

Fig. 2: Nasal endoscopy finding on post operative follow up showing normal mucosa with no sign of recurrence

The histopathological finding in this case showed the typical appearance of leiomyosarcoma. Paranasal sinus leiomyosarcomas are extremely rare. Therefore, it is essential that immunohistochemical techniques be employed to accurately diagnose sarcomas. Reactivity to desmin and muscle-specific actin in spindle-cell sarcomas confirms a diagnosis of leiomyosarcoma[4,5]. Further confirmation can be obtained by electron microscopy but this was not done for our patient.

Resection of the primary tumour with wide margins and neck dissection is the treatment of choice when there is regional lymph node occurrence. Surgical resection with adequate margins of uninvolved tissue is the recommended treatment. Although many tumours appear to be well circumscribed, the high incidence of local recurrence and the persistence of leiomyosarcomas despite initial wide resection illustrate the infiltrative and aggressive nature of this malignancy. The postoperative course is unpredictable, and long-term follow-up is mandatory.

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