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July 2011

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Naeem Sultan Ali
Aga Khan University

Montasir Junaid
Aga Khan University

Kanwal Aftab
Aga Khan University

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Recommended Citation

Ali, N., Junaid, M., Aftab, K. (2011). Malignant peripheral nerve sheath tumour of maxilla. *Journal of the College of Physicians and Surgeons Pakistan*, 21(7), 420-2.

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Malignant Peripheral Nerve Sheath Tumour of Maxilla

Naeem Sultan Ali¹, Montasir Junaid¹ and Kanwal Aftab²

ABSTRACT

Malignant peripheral nerve sheath tumour (MPNST) is a very rare entity in head and neck with high rate of recurrences and local invasiveness. This tumour is usually found in lower extremities and only 10-12% occur in head and neck region. The diagnosis is considered as the most elusive and difficult among soft tissue sarcomas because of its non specific presentation, both clinically as well as pathologically. This difficulty has now been overcome by immunohistochemistry. We report here a case of MPNST in a 50 years old male with a localized right maxillary growth.

Key words: *Malignant peripheral nerve sheath tumour (MPNST). Neurofibromatosis. Soft tissue sarcoma. S-100 protein.*

INTRODUCTION

Malignant peripheral nerve sheath tumours (MPNST) are rare soft tissue sarcomas of ectomesenchymal origin. They are considered highly malignant sarcomas which are locally invasive, associated with high rates of recurrences and metastatic spread.¹ This tumour is usually seen in lower extremities and retroperitoneal region and is rare in the head and neck region. The World Health Organization coined the term MPNST to substitute the previously heterogeneous and often confusing terms, such as malignant schwannoma, malignant neurilemmoma, neurogenic sarcoma, and neurofibrosarcoma. Cases of MPNSTs have been reported mostly in the soft tissues, less common in the bones and very rare in the maxilla.^{2,3} We report a case of MPNST in a 50 years old male with a right maxillary mass.

CASE REPORT

A 50-year-old male patient complained of swelling on the right side of face since 4 months. He gave a history of swelling, which was initially small, roughly marble sized and progressively increased in size over the past four months. It was not associated with any dental pain. There was no fluctuation in the size of the swelling. Patient also did not have any history of epistaxis, speech, chewing or breathing difficulty, numbness over the face or blurring of vision. Past medical, dental and surgical histories were insignificant. His personal history revealed that he was a chronic tobacco chewer and smoker.

Extra oral examination showed an ovoid swelling measuring 6x5 cm on the right middle third of face just

below the eye. The skin over the swelling was stretched and surrounding tissue appeared normal. This swelling was firm, non-compressible, nonfluctuant and non-reducible. Upon intra oral examination no visible ulcer or swelling or loose teeth identified. Mouth opening was adequate. Nasal endoscopy showed a soft tissue bulge on its lateral wall.

Based on history and clinical examination, a provisional diagnosis of antral malignancy was given and differential diagnosis included squamous cell carcinoma, mucoepidermoid carcinoma, adenoid cystic carcinoma, chondrosarcoma and osteosarcoma.

CT scan showed destruction of maxilla on right side, which was associated with soft tissue replacement (Figure 1). The lesion was extending into orbit, ethmoid sinus and pterygopalatine fossa. Incisional biopsy was taken via sublabial approach. Histopathological findings were suggestive of a spindle cell malignancy. The patient then underwent hemimaxillectomy. The tumour was visualized bulging out from the antrum extending posteriorly up to pterygoid muscles, superiorly orbital fat, laterally buccal fat and cheek muscles (not involving them) and medially eroding the medial wall of maxillary sinus. Floor of the sinus was found spared.

Histopathologic studies revealed a circumscribed cellular neoplastic lesion composed of spindle cells arranged in a storiform pattern. Lesion consisting of fusiform cells and epithelial cells with marked pleomorphism and multinucleated giant cells. The diagnosis was sarcoma of a high grade malignancy. Immunohistochemical studies showed tumour cells negative for cytokeratins, actin, desmin, myoglobin and factor VIII, however, it was positive to Protein S-100 (Figure 2). This led to the final diagnosis of malignant peripheral nerve sheath tumour of the maxilla.

The patient was then referred to radiation oncology service for adjuvant radiation. He is on regular follow-up for past 8 months and has presented no evidence of local recurrence.

Department of ENT¹ / Pathology², The Aga Khan University Hospital, Karachi.

Correspondence: *Dr. Naeem Sultan Ali, DD-115, Karimabad Colony, Block No. 3, F.B. Area, Karachi-75950.*

E-mail: naeem.sultanali@gmail.com

Received April 20, 2010; accepted May 10, 2011.

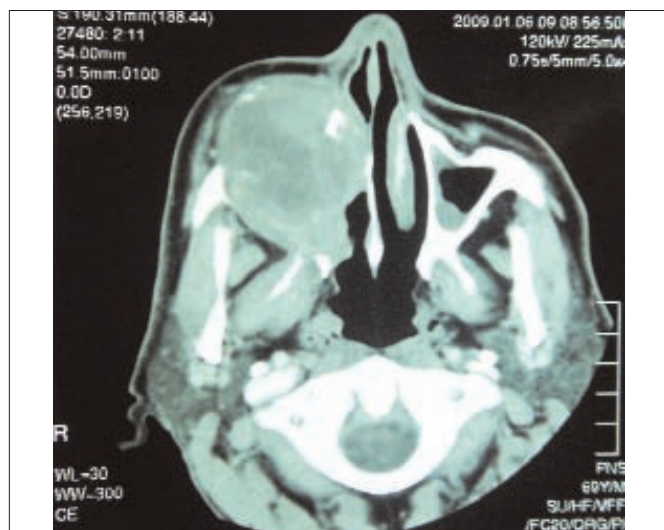


Figure 1: Pre-operative axial CT shows right maxillary mass.

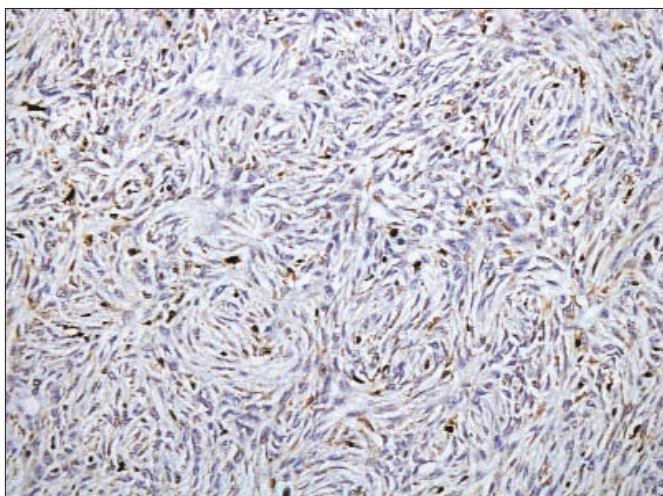


Figure 2: Patchy immunoreactivity for S-100 protein in the tumour cells (immunohistochemical stain, 20x).

DISCUSSION

MPNST is an aggressive sarcoma of neural origin showing close association with peripheral nerves or neurofibroma or may show features of neural differentiation. Most of these nerve sheath tumours in nose and paranasal sinuses arise from ophthalmic and maxillary branches of trigeminal nerve and its branches although it is difficult to identify exactly which nerve is involved.⁴ The most common sites involved are maxillary and ethmoid sinuses.⁵

Malignant peripheral nerve sheath tumours can arise de novo or as a malignant transformation of neurofibromatosis-1 (NF-1) gene. Approximately 50% of MPNST are associated with a family history of von Recklinghausen's disease. Since this patient denied of any such family history it was possibly a de novo case. The risk of developing MPNST in patients with NF-1 is approximately 2-4%. It is also seen to arise in patients previously treated with radiotherapy.⁶

These cases have been reported in all age groups, but peak incidence is seen commonly in second and fourth decade of life with an equal male and female predilection.⁷ MPNSTs are commonly seen in lower extremities and retroperitoneum followed by trunk, upper extremities and head and neck regions. In head and neck, frequent sites include nasopharynx, paranasal sinus, nasal cavity, orbit, oral cavity, parapharyngeal region, neck, thyroid gland and larynx.^{8,9}

The clinical presentations differ as per site involved. But in case of maxilla, they may present with facial and/or orbital swelling, paraesthesia of cheek, epiphora, nasal discharge or epistaxis (if involving nasal cavity). Since these tumours are aggressive they have tendency to erode into adjacent bones and tissue. Two third of these lesions are greater than 5 cm at the time of presentation. This tumour can spread through hematogenous route and by perineural extension. Lymph node metastasis is rare. Roof of orbit and floor of maxilla was not eroded in this case. However, the anterior wall was deficient completely. No nodal metastasis was found in this patient.

Histologically, these tumours have no defined, classic appearance. Commonly described findings are the presence of spindle cells with high mitotic rate and indistinct cytoplasmic borders arranged in bundles or fascicles. Immunohistochemistry plays an important part in the diagnosis and in excluding fibrosarcoma, synovial sarcoma and fibrous histiocytoma. MPNST specifically demonstrates S-100 positivity.

The recommended treatment of MPNST is the surgical extirpation including wide margins. In this patient hemimaxillectomy via Weber-Ferguson incision was opted. Elective neck dissection is not recommended unless there is positive radiological and/or clinical evidence of nodal metastasis. The role of radiotherapy and chemotherapy is still controversial. This tumour has been reported radioresistant, but some authors reported use of complementary radiotherapy.¹⁰ However, the prognosis is poor and even worse in patient with NF-1 disease.

REFERENCES

1. Patil K, Mahima VG, Ambika L. Malignant peripheral nerve sheath tumour: an elusive diagnosis. *Indian J Dent Res* 2007; **18**: 19-22.
2. Kar M, Deo SV, Shukla NK, Malik A, DattaGupta S, Mohanti BK, et al. Malignant peripheral nerve sheath tumours (MPNST) - clinicopathological study and treatment outcome of twenty-four cases. *World J Surg Oncol* 2006; **4**:55.
3. Lesic A, Bumbasirevic M, Atkinson HD, Maksimovic R, Sopta J, Atanackovic M. Malignant intraosseous peripheral nerve sheath tumour of the proximal femur: a case report. *J Orthop Surg* 2006; **14**:84-9.
4. Beegun I, Bottrill ID, Hollowood K. Malignant peripheral nerve sheath tumours of the infraorbital nerve: case report and

- literature review. *J Laryngol Otol* 2009; **123**:466-70. Epub 2008 May 20.
5. Kim HJ, Kim CH, Kim H, Yoon JH. Malignant peripheral nerve sheath tumour of the paranasal sinus. *J Otolaryngol* 2007; **36**:E9-11.
 6. Makimoto Y, Yamamoto S, Takano H, Motoori K, Ueda T, Kazama T, *et al.* Imaging findings of radiation-induced sarcoma of the head and neck. *Br J Radiol* 2007; **80**:790-7. Epub 2007 Oct 1.
 7. Martinez Devesa P, Mitchell TE, Scott I, Moffat DA. Malignant peripheral nerve sheath tumours of the head and neck: two cases and a review of the literature. *Ear Nose Throat J* 2006; **85**:392-6.
 8. Cheng SF, Chen YI, Chang CY, Peng Y, Liao SL. Malignant peripheral nerve sheath tumour of the orbit: malignant transformation from neurofibroma without neurofibromatosis. *Ophthal Plast Reconstr Surg* 2008; **24**:413-5.
 9. Sabesan T, Hussein K, Ilankovan V. Malignant peripheral nerve sheath tumour of the parapharyngeal space in a patient with neurofibromatosis type 1. *Br J Oral Maxillofac Surg* 2008; **46**:585-7. Epub 2008 Mar 18.
 10. Minovi A, Basten O, Hunter B, Draf W, Bockmühl U. Malignant peripheral nerve sheath tumours of the head and neck: management of 10 cases and literature review. *Head Neck* 2007; **29**:439-45.

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