MALIGNANT PERIPHERAL NERVE SHEATH TUMOR: A RARE HEAD AND NECK MALIGNANCY

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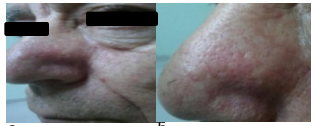
Background: Malignant peripheral nerve sheath tumor (MPNST) is an uncommon sarcoma with an incidence of 1:100,000/ year. It is encountered mostly in patients with neurofibromatosis type I, while in the general population it has an incidence of 0.001%. MPNSTs are infrequently found in the head and neck region. A case of MPNST of the nose is reported.

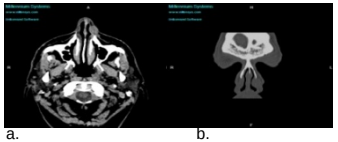
Methods: A-65-year-old male presented with a painless, enlarging, subcutaneous mass on the left side of the nose (Fig. 1a, b). Nasal endoscopy revealed no pathology inside the nose. The noncontrast CT scan of the paranasal sinuses showed a subcutaneous, oval-shaped, well-defined mass measuring 13x 12 mm. The lesion was confined to the left nasal cavity, mostly at the base of the left nostril (Fig. 2a, b). The mass was subjected to excisional biopsy. Histologic examination of the specimen revealed the presence of spindle cells arranged in bundles, with a mitotic rate of 14/10HPF (Fig. 3). Immunohistocheminal analysis showed that the tissue was positive for Vim, S-100, CD34 and negative for SMA, desmin, CD99, C-kit, MART-1, HMB-45, GFAP, p63, AE1/AE3 and CK34bE12 (Fig. 4). It was concluded that the findings were consistent with a Grade 2 sarcoma according to FNCLCC Grading System, possibly MPNST. Post- surgical head and neck MRI scan with paramagnetic contrast agent depicted neither residual tumor (Fig. 5) nor pathological lymph nodes, while chest CT scan with contrast and abdominal ultrasound showed no evidence of metastatic spread. The patient was referred to a regional oncology hospital for further management. Based on pathology report, he was subjected to a wide excision of the site of the lesion by performing medial maxillectomy and reconstruction of the defect by using the reverse nasolabial flap. The histologic examination showed no residual disease.

Fig. 1a, b: Subcutaneous mass on the left side of the nose



a. b.

Fig. 2: Noncontrast CT scan. a: axial, b. coronal. Subcutaneous, well-defined mass of the left nostril.



a. b.

Fig. 3 (H-Ex 400)**:** Grade 2 Spindle–cell sarcoma

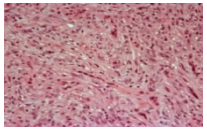


Fig. 4 (x 200): S-100

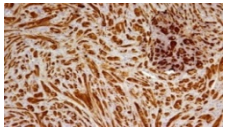
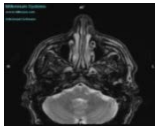
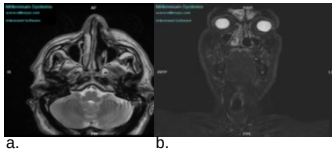


Fig. 5: T2-weighted axial MR image after excisional biopsy without evidence of residual tumor.



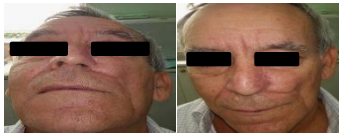
Results: There was no recurrence till date, 6 months postoperatively (Fig. 6a, b). Aesthetic and functional result is considered very satisfactory (Fig. 7a, b).

Fig. 6: T2-weighted MRI. a. axial, b: coronal. Six months after medial maxillectomy without sign of recurrence.



a. b.

Fig. 7a, b. Post- surgical result after 6 months.



a. b.

Conclusions: MPNST is a rare, aggressive tumor with a high ratio of local recurrence. Lymphatic spread is rare. Distant metastasis can occur. Radical tumor excision with wide margins of normal tissue is the mainstay of treatment.