**INTRODUCTION**

Malignant schwannomas are tumors that arise from the Schwann cells of the peripheral nerves. They represent 10% of all soft-tissue sarcomas, and are rare in the head and neck region. The extremities, trunk and thorax are the locations most commonly affected. When their origin is in the parotid sinus, tumor-related symptoms are often present before the lesion is identified, with a relatively small size. This means that they remain unsuspected and are often diagnosed late.

**CASE REPORT**

A 57-year-old smoker and heavy drinker, who was a truck driver, came to the Otorhinolaryngology and Head and Neck Surgery Service of Brasilia University Hospital presenting nasal obstruction, rhinorrhea and epistaxis of three years duration in the right nasal cavity (RNC), along with ipsilateral facial pain. Two months earlier, he noticed epistaxis and epoxiphthalia in his right eye with unaffected visual acuity, but with hearing loss in his right ear. Physical examination showed a bony nose, with left sinus deviation of the nasal septum, and a friable mass inside the RNC, showing hemorrhagic spots. Computed tomography (CT) on the paranasal sinuses revealed a voluminous mass in the RNC, extending as far as the pterygopalatine fossa, measuring approximately 60 x 70 x 20 mm and eroding the medial wall of the maxillary sinus, the lamina papyracea, the anterior and posterior ethmoid cells and part of the wing of the sphenoid bone (Figure 1A-B).

Endoscopic resection of the mass was attempted and a friable brownish lesion with its epidermis clearly痨able in the posterior ethmoid cells was removed, along with infiltration of adjacent tissue. A biopsy was performed and the histo-pathological examination revealed a malignant neoplasm represented by atypical fusiform cells in a fascicular arrangement, with great alternation of cellular density and areas with intense collagen deposition, and with frequent mitotic figures and focal areas of necrosis. Immunohistochemical analysis showed positivity of S-100 protein and type IV collagen (Figure 1C-D). The histological and immunohistochemical findings were compatible with malignant tumors of the peripheral nerve sheath.

The patient then underwent another procedure to check on the surgical margin status, using an open approach via lateral rhinotomy. The omentum was preserved. There wasn’t tumor infiltration beyond the margin of resection. Postoperative radiotherapy was administered to reduce the risk of the disease recurrence. Up to the present moment (24 months after the operation), the patient has not presented any signs of tumor recurrence or distant metastases.

**DISCUSSION**

Malignant schwannomas are sarcomas that arise from Schwann cells of the peripheral nerves. They represent approximately 10% of soft-tissue sarcomas, and are unusual in the head and neck region (8% of the cases). The most common locations are the extremities, trunk, chest and retroperitoneum. Several studies have shown that malignant schwannomas in the nasal cavity, paranasal sinuses and nasopharynx are rare. In the latter region, the epithelial and maxillary sinuses are mostly affected, the nasal fossa and sphenoid sinuses come next and the frontal sinus is rarely affected. There are few cases of benign schwannomas in national literature. Just Pirio et al. cited an unique case of epithelial malignant schwannoma in a study about nasal and paranasal-sinus malignant tumors.

Malignant schwannomas of the paranasal sinuses usually originate in the trigeminal nerve, usually from the ophthalmic or maxillary division and their terminal branches. Individuals in their fourth and fifth decades are more often affected, and there is no preference for sex or race. They can occur alone, but are associated with von Recklinghausen disease in 30% of the cases, and usually evolve from malignant transformation of neurofibromatosis.

Microscopically, malignant tumors of the peripheral nerve sheath are characterized by layers of fusiform cells, with indistinct outlines and a moderate amount of cytoplasm. The nuclei are ovoid or fusiform with cellular and atypical pleomorphism. The mitotic activity level is variable and indicates the aggressiveness of the tumor. Immunohistochemical analysis is positive for the S-100 protein. The differential diagnosis is made with fibrousomas, malignant fibrous histiocytomas, benign schwannoma, capillary hemangioma, hemangiopericytoma and other tumors.

**REFERENCES**

1. Ghosh BK, Ghosh L, Hovee AG, Fortune Jr. Malignant schwannoma. A clinicopathologic study. Cancer. 1975;35(12):184-90.
2. Herman AA, Singh M, Balaban B, Agustina M, Sharma MC. Solitary malignant schwannoma of the nasal cavity and paranasal sinuses: report of two rare cases. Ear Nose Throat J. 2003;82(5):304-6.
3. Perzin KH, Pannu H, Wechter S. Nephroblastomas of the nasal cavity, paranasal sinuses and nasopharynx. A clinicopathologic study. Schwann cell tumors (neurilemoma, neurofibroma, malignant schwannoma). Cancer. 1982;50(3):219-21.
4. Scharner JA, Mark BK, Tran L, Stroope I, Calcagno TC. Sarcomas of the nasal cavity and paranasal sinuses. Ann Otol Rhinol Laryngol. 1994;103(9):796-9.
5. Khademi B, Owji SM, Khodhari MJ, Momandipour M, Gandestani R. Description of a neural sheath tumor of the trigeminal nerve: immunohistochemical and electron microscopy study. Sao Paulo Med J. 2006;124(3):355-5.
6. Kiureghian JRS, Becker FRM, Silva CDL, Grossa PFTB, Ferreira JR, Salles PGO, et al. Schwannoma nasal com extensão intracraniana. Relato de Caso. Rev Bras Otorrinolaringol. 2000;66(3):279-82.
7. Pitto JA, Pinto HCF, Felippa A. Cirurgia combinada craneofacial para tumores malignos do nariz e seios paranasais. Rev Bras Otorrinolaringol. 1994;62(2):112-20.
8. Hamaker F, Kayabara H, Sumi Y, Esumi H. Mandibular malignant schwannoma with multiple spinal metastases: a case report and a review of the literature. J Oral Maxillofac Surg. 1998;56(10):1191-4.

**Keywords**: epistaxis, nasal obstruction, paranasal sinus neoplasms.