CASE REPORT

Malignant peripheral nerve sheath tumour of the tongue

Anacélia Mendes Fernandes, Aline Cristina Batista Rodrigues Johann, João Batista da Silveira-Júnior, Maria Cássia Ferreira de Aguiar, Maria Auxiliadora Vieira do Carmo, Ricardo Alves Mesquita

Oral Surgery, Medicine and Pathology Department, Dentistry School, Federal University of Minas Gerais, Av. Antônio Carlos, 6627 sala 3204, Pampulha 31.270-901, Belo Horizonte, MG, Brazil

Received 5 December 2005; accepted 6 December 2005

Summary Malignant peripheral nerve sheath tumour (MPNST) is a rare malignant neoplasm in the maxillofacial region. This study reports on a sporadic case of MPNST located on the tongue and provides a review of existing literature regarding this lesion. Data analyses show four well-documented cases of MPNST on the tongue.

KEYWORDS Malignant peripheral nerve sheath tumour; Malignant schwannoma; Tongue; Neurogenic sarcoma

Introduction

Malignant peripheral nerve sheath tumour (MPNST) is the coined term used by the World Health Organization and corresponds to the malignant proliferation of any cell of the nerve sheath: Schwann cell, perineural fibroblast or endoneural fibroblast.\(^1\) MPNST represents 5–10% of soft tissue sarcomas and is manifested mainly associated to the type I neurofibromatosis.\(^2\) About 8–16% MPNST develops in the head and neck region.\(^3\) There are reports of only four sporadic cases involving the tongue in the English literature.\(^4\)\(^,\)\(^6\)\(^,\)\(^8\)\(^,\)\(^9\)\(^,\)\(^12\) The purpose of this paper is to report a case of MPNST of the tongue occurring in a 37-year-old man and to review the literature about this lesion.

Case report

A 37-year-old black man was referred to the Oral Medicine Clinic of the Federal University of Minas Gerais (UFMG), in April 2004, complaining of swelling in the tongue with an evolution of one week. The intraoral physical exam showed a painful, ulcerated, exophytic, purple tumor measuring \(2 \times 2 \) cm in the posterior region of the tongue (Fig. 1(A)). The clinical diagnoses were pyogenic granuloma and mesenchymal neoplasm. An incisional biopsy was taken and the specimen fixed in 10% neutral formalin. Microscopic examination showed a solid neoplasm alternating within hyper and hypoplastic areas (Fig. 1(B)). The cells revealed spindle form with wavy nuclei and elongated cytoplasmic process. In focal area a Antoni A pattern was observed (Fig. 1(C)). In the hypocellular area, representing a undifferentiated area, the cells exhibited large and pleomorphic nuclei and elongated cytoplasmic process (Fig. 1(D)). Foci of necrosis

---

* Corresponding author. Tel.: +55 31 34992478; fax: +55 31 34992472.

E-mail address: ramesquita@ufmg.br (R.A. Mesquita).
and hemorrhaging were present. Immunohistochemical reactions were performed with streptavidine–biotin protocol. The neoplastic cells were S-100 positive (Dako, clone: Z0311, dilution 1:100, no antigen retrieval) (Fig. 1(E)). Reactivity for Ki-67 (Dako, clone: A0047, dilution 1:75, antigen retrieval with 0.01 M citric acid, 95°C, 30 min) was 6.1% in hypercellular areas and 5.9% in hypocellular areas (Fig. 1(F)). A diagnosis of MPNST was performed. The patient was re-evaluated; and type I neurofibromatosis was discarded. The patient was treated using surgical resection and still remains free-disease after seventeen months (Fig. 1(G)).

A review of the English literature about MPNST of the tongue that not associated with type I neurofibromatosis demonstrated four well-documented cases over the last 29 years (Medline since 1966). Relevant data are presented in Table 1.

Discussion

MPNST is a rare malignant lesion, and the current case report represents the fifth case located in the tongue. In the maxillofacial region, MPNST can be found in the jaws or in soft tissues. In soft tissues, the lesion usually appears with indistinct margins, and may be ulcerated, painful or cause paresthesia. This case report revealed an ulcerated and painful lesion. Also, as the case reported, patients with MPNST on the tongue are generally young.1,2,6,8–10,12

MPNST have arise "de novo" from a neuroectodermal tissue, being considered a sporadic cases.2,3,8 This case represents a "de novo" form of the lesion, as it did not detect any alteration or signs of type I neurofibromatosis. Histopathologically, it was characterized as a conventional type containing an area of schwannoma differentiation. The area of schwannoma differentiation is present in 10% of MPNST.2,4 Fifty to seventy percent of MPNST are S-100 immunopositive. Although S-100 expression is not exclusive of MPNST, it is indicative of the neural differentiation. Other proteins such as glial fibrillary acidic protein (GFAP), Leu-7, myelin basic protein, neuron-specific enolase (NSE) and neurofilament may be evaluated in the diagnosis of the MPNST.5,7

The MPNST is treated by surgical excision, but local recurrence is common. Hematogenous metastasis occurs in at least half of all cases.2,3,8,9 The patient’s survival is correlated to the size of the lesion, adequacy of margins, association or not with type I neurofibromatosis, and immunohistochemical findings. Watanabe et al.11 correlated a high Ki-67 labeling index (>25%) with a reduced survival rate. Our case had a Ki-67 labeling index below that of 25% in more differentiated and undifferentiated areas. Overall, the five-year survival rate for MPNST is 40–75%.3 Follow-up on cases of MPNST of the tongue without type I
neurofibromatosis (Table 1) ranged from one to three years, with one case presenting metastase.6,8,9,12 The prognosis of our patient is good, because it is a sporadic case, of small size and was management with wide surgical excision.

**Acknowledgments**

This study was supported by grants from CAPES, FAPEMIG and CNPq. Mesquita RA and Aguiar MCF are research fellows of the National Council for Scientific and Technological Development (CNPq).

**References**