Peripheral primitive neuroectodermal tumor: a rare case of peripheral facial paralysis

INTRODUCTION

The primitive neuroectodermal tumor (NEDT) was first described in 1973 by Hart and Earle in order to characterize an undifferentiated neoplasia of the central nervous system, which represents between 90 and 95% of undifferentiated cells and does not fulfill diagnostic criteria for any other type of neoplasia. The peripheral NEDT on the head and neck is extremely rare, and in a recent literature review we did not find any report of such tumor involving the temporal bone and the facial nerve canal. Our goal with the present paper is to report on a patient who presented peripheral facial paralysis as initial manifestation of the peripheral NEDT involving the temporal bone.

CASE REPORT

R.A.O., a 32 year old female, complaining of 30 days of facial movement impairment, associated to right side otalgia. Otolaryngological exam showed grade IV (House-Brackmann scale) peripheral facial paralysis on the right side, and normal otoscopic exam. Temporal bone CT scan showed enlargement of the facial nerve canal within the mastoid (Figure 1).

DISCUSSION

Temporal bone and facial nerve involvement by neoplasia is not common, it represents about 5% of peripheral facial paralysis cases, and malignant tumors are even rarer. Temporal bone metastases are extremely rare, not even having a notified real incidence.

Peripheral NEDT is a neoplasia of primitive neuroectodermal cells, of rare occurrence. It comes from the peripheral nervous system and bears considerable predilection for limbs and pelvis, regardless of age range. Most NEDT cases that involve the spinal cord represent metastatic lesions of an intracranial primary tumor; however, the spine may be its primary site, especially in young adults. In this case, it is very likely that the primary lesion started on the facial nerve, followed by metastasis to the spinal cord and lung, as we can see in the clinical evolution of symptoms. We believe the lesions located in the lung and the spinal cord to be metastasis, because in blood borne spread these are the preferred sites due to their intense vascularization.

In the literature we found numerous series showing varied primary sites affected by the NEDT. It is thought that the NEDT comes from the neoplastic transformation of primitive neuroepithelial cells. The fact that such cells may remain anywhere in the nervous system may explain the diverse origins of this tumor. In our bibliographic review (MEDLINE and Lilacs) we found no report of temporal bone involvement by NEDT primary or metastatic lesions. Diagnostic confirmation for this tumor is carried out through immunohistochemistry, and it is positive for Vimentin and Mib2/CD99. Despite combined treatment (surgery, chemo and radiotherapy) prognosis is still poor, with average survival between 6 and 42 months.

REFERENCES