Clear cell sarcoma of the parotid region
Evandro Maccarini Manoel, Rafael Reiser, Fábio Brodskyn, Marcello Franco, Márcio Abrahão, Onivaldo Cervantes

Case Report

INTRODUCTION

Clear cell sarcoma (CCS), also referred to as malignant melanoma of the soft parts, is a rare aggressive tumor that accounts for less than 1% of all soft tissue sarcomas1. It occurs typically as a deep lesion that arises in connection to tendons and aponeuroses, involving the skin only in advanced cases2. It is observed more frequently in adolescents and young adults of both genders, and preferentially affects the lower extremities3. It is rarely seen in the head or neck1.

CASE PRESENTATION

A 43-year-old Caucasian female came to our service complaining of a lump that had been growing in her right parotid region for a year and four months. She had no other symptoms. The patient had well-managed systemic high blood pressure and asthma. Physical examination revealed a tumor in her right parotid region with a diameter of five centimeters. The tumor was hard, barely mobile, ulcerated, hyperemic, and painless to palpation.

Fine-needle aspiration (FNA) revealed a basoloidal neoplasm with low rates of cell proliferation. Head and neck CT scans showed a tumor located in the patient’s right parotid region (Figure 1A).

The patient was referred to surgery and underwent a superficial parotidectomy with neck clearance on level II; the accessory nerve was removed as it had been involved by the tumor. Histopathology tests showed the tumor was a tumor with five centimeters in its greater diameter enhanced with contrast in the patient’s right parotid region. B: optical microscope; H&E stained slide (magnification 100x). C: immunohistochemistry assay with diffuse positive result for protein S-100 (magnification 200x). D: FISH test showing rearrangement in gene EWSR1 resulting from translocation (12;22) (q13;q12). Eight months after surgery the neck tumor recurred, and the patient was submitted to a radical neck clearance procedure and adjuvant radiotherapy (6600 Gy). The patient has been followed for six months since, and no relapsing tumors have been found.

DISCUSSION

Clear cell sarcoma was first described in 19654, and has been known as malignant melanoma of the soft parts because of the histological and immunohistochemical similarities it bears with melanomas5. However, molecular analysis revealed they are distinct tumors, as CCS presents translocation (12;22)(q13;q12) that results in chimeric gene EWSR1/ATF1, which is not seen in melanomas6. This alteration is also seen in hyalinizing clear-cell carcinoma of the salivary glands, angionoidoid fibrous hystiocytomas, and in few cases of the recently described gastrointestinal subtype of CCS7. In this case, morphology and absence of melanocytic markers match the diagnosis of this variant8. Only 1.2% of the approximately 500 reported cases analyzed positive for protein S-100 (Figure 1C), and negative results for malignant melanoma markers HMIB45, Melan-A, MART-1 and MITF. A FISH (fluorescence in-situ hybridization) test showed translocation (t(12;22) (q13;q12) (Figure 1D) and changed the diagnosis to CCS.

While measuring under five centimeters7. It presents high local recurrence and late metastasis rates and, contrary to most sarcomas, its metastases appear preferentially in regional lymph nodes. Five and ten-year survival rates are approximately 47% and 36% respectively9. Tumors larger than five centimeters and presence of tumor necrosis mean worse prognosis10. The better therapy appears to be broad excision of the tumor followed by adjuvant radiotherapy11. Given the limited number of reported cases, the role of the neck clearance procedure and systemic adjuvant therapy are still uncertain.

REFERENCES


1 MD (Resident ENT Physician at EPM-UNIFESP).
2 MD, Head and Neck Surgeon (MD, Head and Neck Surgeon at EPM-UNIFESP).
3 MD, ENT, Head and Neck Surgeon (MD, ENT, and Head and Neck Surgeon at EPM-UNIFESP).
4 MSc on Pathology at EPM-UNIFESP (Professor in the Department of Pathology at EPM-UNIFESP).
5 Adjunct Professor in the Department of Otorhinolaryngology and Head and Neck Surgery at EPM-UNIFESP (Vice-Head of the Head and Neck Surgery Course at EPM-UNIFESP).
6 Adjunct Professor in the Department of Otorhinolaryngology and Head and Neck Surgery at EPM-UNIFESP (Head of the Head and Neck Surgery Course at EPM-UNIFESP and Chairman of the Brazilian Association of Head and Neck Surgery).
7 Universidade Federal de São Paulo (EPM-UNIFESP).

Send correspondence to: Evandro Maccarini Manoel, Rua Dr. Diogo de Faria, nº 539, apto. 67, Vila Clementino, São Paulo - SP, CEP: 04037-001. Paper submitted to the BJORL-SGP (Publishing Management System - Brazilian Journal of Otorhinolaryngology) on June 6, 2011; and accepted on June 3, 2012. cod. 8058.