Clear cell sarcoma of the parotid region

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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 sarcomas. It occurs typically as a deep lesion that arises in connection to tendons and aponeuroses, involving the skin only in advanced cases. It is observed more frequently in adolescents and young adults of both genders, and preferentially affects the lower extremities. It is rarely seen in the head or neck.

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 basaloïd 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 spared. Histopathology tests showed the tumor was a basaloid neoplasm with low rates of cell proliferation. The histology of the tumor was consistent with the diagnosis of CCS.

Eight months after surgery the neck tumor recurred, and the patient was submitted to a radical neck clearance procedure and adjuvant radiotherapy (6600 cGy). The patient has been followed for six months since, and no relapsing tumors have been found.

DISCUSSION

Clear cell sarcoma was first described in 1965 and has been known as malignant melanoma of the soft parts because of the histological and immunohistochemical similarities it bears with melanomas. However, molecular analysis revealed they are distinct tumors, as CCS presents translocation $(t(12;22)(q13;q12))$ that results in chimeric gene EWSR1-ATF1, which is not seen in melanomas. This alteration is also seen in hyalinizing clear cell carcinomas of the salivary glands, angiomatoid fibrous histiocytomas, and in few cases of the recently described gastrointestinal subtype of CCS. In this case, morphology and absence of melanocytic markers match the diagnosis of this variant.

Only 1.2% of the approximately 500 reported cases of CCS involved the head or neck. The parotid region was compromised in only three cases reported in the literature. CCS generally appears preferentially in regional lymph nodes. Five and ten-year survival rates are approximately 47% and 36% respectively. Tumors larger than five centimeters and presence of tumor necrosis mean worse prognosis. The better therapy appears to be broad excision of the tumor followed by adjuvant radiotherapy. Given the limited number of reported cases, the role of the neck clearance procedure and systemic adjuvant therapy are still uncertain.

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