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Case Report

A Giant Primary Leiomyosarcoma of the Neck: Report of One Case

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ABSTRACT

Leiomyosarcomas are rare malignant tumors of smooth muscles, accounting for 4% of sarcomas of head and neck sarcomas. They are characterized by slow growth with metastatic potential, hence the value of early diagnosis and management. Through a case of primary cervical leiomyosarcoma, we illustrate clinical, therapeutic and evolutionary particularities of this rare tumor.

We report a case of a 47-year-old patient who consulted for a recent painful right paramedian cervical tumefaction with dysphagia. Examination objectified a hard cervical mass with no clear limits, fixed to superficial and deep planes with inflammatory skin. Cervical CT and MRI were performed and objectified a right paratracheal cystic mass measuring 78mm long axis. Peroperatively the mass was adhering to pre-vertebral muscle planes, trachea and the right carotid artery. Resection was impossible and the biopsy had concluded a grade III leiomyosarcoma.

Leiomyosarcoma is a rare tumor of the neck region. An early diagnosis and aggressive initial treatment remains the mainstay of therapy for a good prognosis.

INTRODUCTION

Leiomyosarcoma (LMS) is a malignant tumor of smooth muscle commonly found in the pelvic area or gastrointestinal tract [1]. Its neck involvement is rare, accounting for less than 4% of head and neck sarcomas and mostly described in the thyroid gland [2]. LMS is a slow growing tumor with an important metastatic potential with high mortality [3]. The positive diagnosis requires anatomopathologic examination with immunohistochemical analysis. Through an original observation of a rapidly evolving neck leiomyosarcoma in a 47-year-old patient, we emphasize the need for early diagnosis and adequate treatment based on complete surgical excision to improve prognosis. Resection was impossible and the biopsy concluded a grade III leiomyosarcoma.

CASE REPORT

We report a case of a 47-year-old female patient with a medical history of thyroidectomy for multinodular goiter. She consults for a recent painful right paramedian cervical tumefaction with dysphagia. Clinical examination showed a paramedian hard fixed cervical swelling of 5 cm long with healthy looking skin.

Cervical ultrasound showed a right paramedian solid mass of 52mm*35mm*28mm associated with satellite adenopathies. Cervical CT and MRI were performed and objectified a right paratracheal cystic mass measuring 78mm long axis (Figure 1). The decision to operate was made, but the patient refused and was lost to follow-up. She reconsult, two months later, for worsening of the symptomatology and an apparition of a dyspnea. Examination objectified a hard cervical mass with no clear limits, fixed to superficial and deep planes with inflammatory skin.

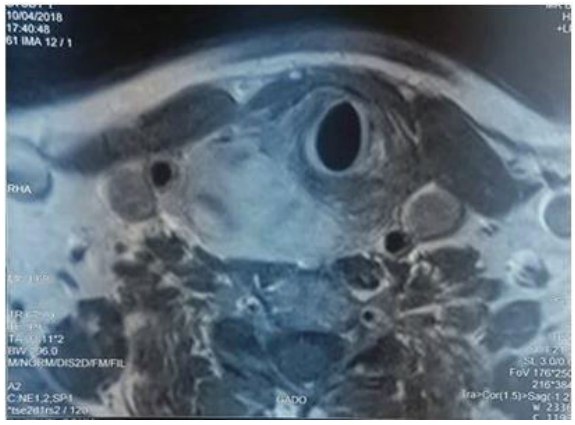

Figure 1: Cervical axial MRI cut illustrating a tissue mass developed in the posterior cervical region with intense enhancement after gadolinium injection.

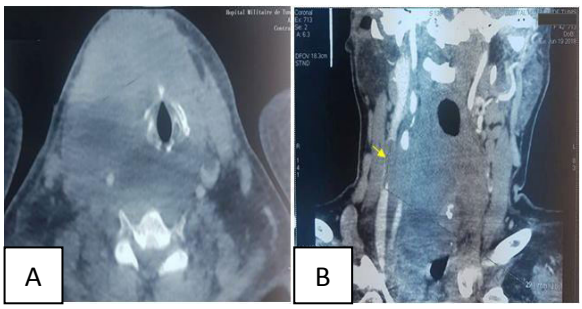

Figure 2: Axial (A) and coronal (B) images showing a tissular mass of the posterior cervical region extended from C3 to C7; with a thrombosis of the jugular vein.

New radiological examination (Scan CT) showed a solid mass encompassing digestive tracts with a mass effect on the respiratory and a thrombosis of the jugular vein (Figure 2). Peroperatively the mass was adhering to pre-vertebral muscle planes, trachea and the right carotid artery. Resection was impossible and the biopsy had demonstrated a grade III leiomyosarcoma of NCI classification (Figure 3). Metastases have been objectified in the liver, lungs and pelvis.

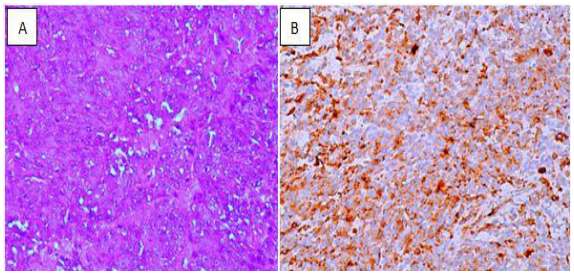

Figure 3: Histopathological slides showing (A)Tumor proliferation arranged in a diffuse layer of tumor cells presenting many features of mitosis (HE*400) with diffuse and intense cytoplasmic positivity to caldemol in immunohistochemical study (B).

Radiochemotherapy was not achieved because of the rapid evolution with exteriorization of tumor (Figure 4). Death occurred after a few days in a context of cerebral embolism.

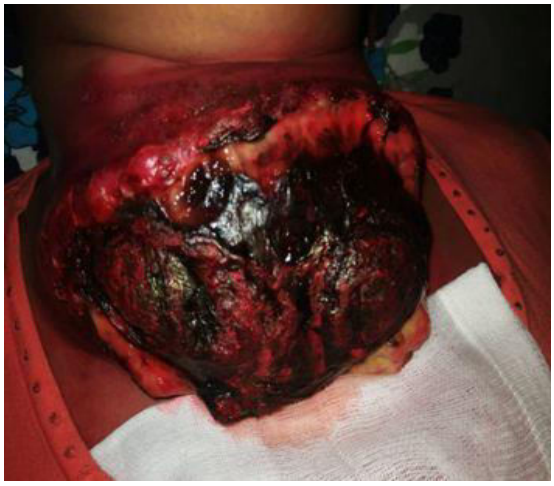

Figure 4: Exteriorization of tumoral mass.

DISCUSSION

Leiomyosarcoma is a rare malignant tumor of smooth muscle, which accounts for only 4% of head and neck sarcomas [4,5]. While the arrector pili muscle is believed to be the most probable source of cutaneous leiomyosarcoma, some authors have suggested that the tunica media of the blood vessels are the source of deep primary leiomyosarcoma [5,6]. Head and neck leiomyosarcomas are very aggressive and associated with a poorer prognosis and are preferentially treated with

surgery. Magnetic resonance imaging is the preferred modality for the evaluation of this type of tumor [5]. Histopathologic exam is essential for the diagnosis with moderate-to-high histologic grade in most cases. Complete surgical excision with wide negative margins remains the mainstay of treatment for leiomyosarcoma [4,5]. A review of head and neck leiomyosarcoma cases by Eppsteiner et al found that patients who underwent surgery had better survival rates than did those who received primary radiotherapy [6]. The classic surgical approaches to the parapharyngeal space are via the transoral, transcervical, transparotid, or transmandibular route. Newer treatment strategies were recently introduced, including an endoscopic transnasal-transmaxillary-transpterygoid approach Achieving safe surgical margins is of paramount importance with respect to overall survival [4,7]. Risk factors of failure of surgical treatment are large tumor size, high grade, deep location, and recurrent disease at presentation [4,8]. In our case surgery treatment was very difficult due to multiple adherences and rapid local progressive of the tumor. Adjuvant chemotherapy has been recommended by some authors, but its impact on overall survival is unclear [6]. Adjuvant postoperative radiotherapy, particularly for high-grade tumors, can effectively improve survival, although its role appears to be quite limited [4,9].

CONCLUSION

Leiomyosarcoma is a rare tumor of the neck region. It has a poor prognosis despite the development of surgical techniques. Chemotherapy and radiotherapy did not show good results. An early diagnosis and aggressive initial treatment remains the mainstay of therapy for a good prognosis.

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