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Apocrine sweat gland adenocarcinoma: A rare case report and review

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ABSTRACT

INTRODUCTION: Primary apocrine sweat gland carcinoma (PASGC) is an extremely rare neoplasia whose management and treatment are still evolving. The only curative therapy is wide local excision. Many patients have metastasis at the time of the diagnosis, mainly because this neoplasm has been misdiagnosed as some benign skin lesions.
PRESENTATION OF CASE: We herein report a case of a 72-year-old-man with PASGC affecting the axilla and regional lymph nodes that underwent surgical resection and lymphadenectomy at our Institution. This is the first case reported in Brazil.
DISCUSSION: Our observation suggests just a MRI as necessary to study tumoral limits and lymph nodes and a full surgical excision with free margins is decisive for success.
CONCLUSION: Despite the PASGC be a rare cancer and require expensive tests, knowledge of this disease is critical to reduce costs in medical services without availability of investment.

1. Introduction

Primary apocrine sweat gland carcinoma is an extremely rare neoplasm with around 50 cases reported in the literature. The slow growing characteristic gives the clinical picture of benign tumor, but sometimes progress aggressively and with metastasis [1,2]. The median age of the patient is around 67 years, without race or gender preference, related in axilla and anogenital area, but can be found in forehead, wrists, ear canals, eyelids, trunk, feet, toes, and fingers. The incidence of PASGC is quite low as 0.0049–0.0173 cases/100,000 people per year [1–3]. Most of these lesions present indolent symptoms and slow growth rate that can delay diagnosis. After excisional biopsy, the treatment is wide local excision with clear margin of 1–2 cm, with axillary lymphadenectomy if clinically positive nodes are detected. The most controversial topic is adjuvant chemotherapy and local radiotherapy [3]. PASGC has a high incidence of local recurrence and lymph node metastasis. Some authors report patients with lungs, liver and bone dissemination [1]. We present a case of a 72-year, male, with axillary apocrine, papillary and acinar adenocarcinoma of sweat gland and review the few existing literatures about the disease and its management. We reported in line with the SCARE criteria [4].

2. Presentation of case

A 72-year-old man was referred to our Surgical Service from a primary health care with a 6-month history of a growing and palpable axillar mass. The lesion was painless, lobed, slightly mobile, hardened and without fistula to the skin. This patient had hypertension and was a smoker (30 years/pack).

The Magnetic Resonance Imaging (MRI) showed a left axillary mass with inaccurate shape, high capitation signal, lobulated and cystic-solid content (exam with better sensitivity and specificity available). Near this mass, there was a clearly defined and extended lymph node (Fig. 1). There was no contact with nervous or vascular structures.

Initially, the surgical team thought about a metastatic tumor and submitted this patient to an excisional biopsy performed in local anestesia, preserving nerve and vascular structures. Macroscopically, this tumor had two cystic cavities (one with a fetid black secretion and other with a doughy white secretion) (Fig. 2). The pathologist described this mass as an irregular tumor ($6.4 \times 3.9 \times 2.5$ cm, 7.0 g) compatible with apocrine, papillary and acinar adenocarcinoma, with free surgical margins (Fig. 3).

Possibly dealing with a primary sweat gland neoplasm, however, it doesn't exclude the possibility of metastasis. Immuno-

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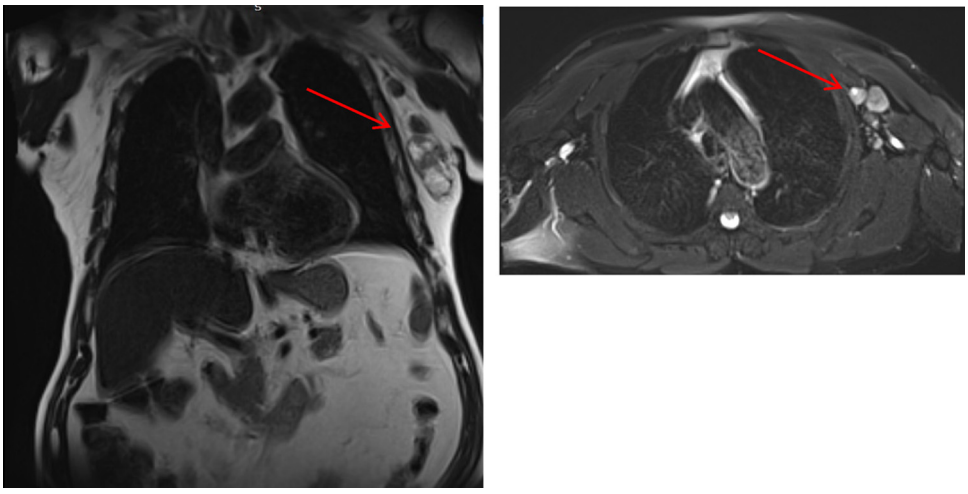

Fig. 1. Thoracic MRI.

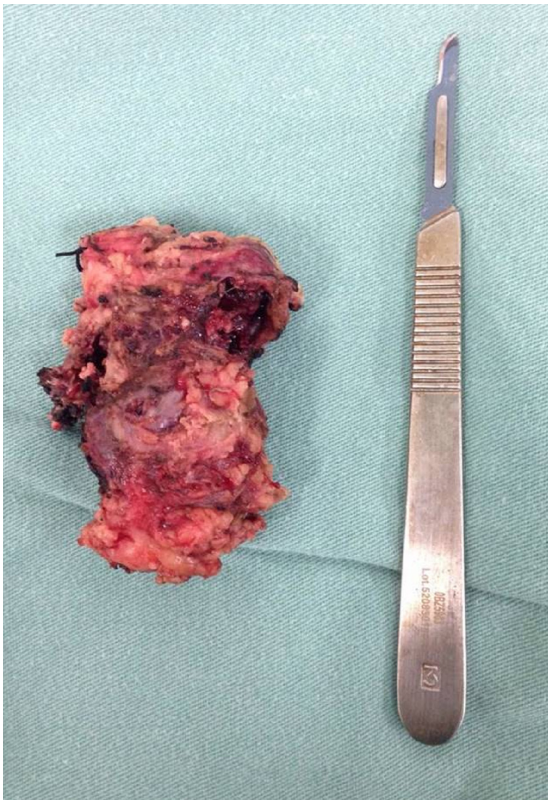

Fig. 2. Sweat Gland Adenocarcinoma (Excisional Biopsy).

histochemical study (Table 1) confirmed the diagnosis of sweat gland adenocarcinoma (apocrine, papillary and acinar).

After an extensive review of the literature, the patient was submitted to an classic axillary dissection preserving nerve structures, since this patient had a large lymph node next to the tumor, possibly positive to metastasis, but no other clinical manifestation or metastatic site (Fig. 4). In this surgery, it was found six macroscopic large lymph nodes around the major one and an Intercostobrachial nerve and a branch of Thoracodorsal vein involvement (that were sectioned and removed together).

The pathology analysis presented 17 lymph nodes, 8 described as adenocarcinoma metastasis, 9 as sinus histiocytosis and adenocarcinoma of fibroadipose tissue (Fig. 5).

Table 1
Immunohistochemical Study.

Antibody	Results
CK7 automated (SP52)	Positive (neoplasy)
GCDFP-15 automated (23 A3)	Positive (neoplasy)
RE automated (SP1)	Negative
CK20 automated (Ks20.8)	Negative
TTF1 automated (SPT24)	Negative
S100 automated (P)	Negative
Napsin A (TMU-AdO2)	Negative

On the postoperative period, the patient evolved without loss of local sensitivity, pain or motor disorders and was referred to oncological care to complete the treatment with chemotherapy (5-fluorouracil) and radiotherapy (50 Gy for 5 weeks). Patient maintained in clinical follow-up without return of disease.

3. Discussion

The PASGC is a rare tumor with a simply treatment when there is no metastasis. The restricted literature about this disease is a barrier to define the best way to treat the patient [1–3,5]. Maybe the most important predictor of survival is the lymph node status. Once the node is involved, and the lack of consensus about chemotherapy and radiotherapy impute many doubts about the best management [2,6]. The literature describes some optimistic results with chemotherapy drugs, like Methotrexate, Bleomycin and 5-Fluorouracil, but there is no consensus. Nevertheless, the local radiotherapy seems to be right for most authors among medical services with great results in long-term progression-free survival [7,8].

Some studies say that the prognosis of patients with or without lymph node involvement is similar to breast carcinoma and steroid receptor expression should be investigated in these tumors [9]. The axillary situs is the most common local of PASGC and the differentiation of metastasis (breast, colorectal, synchronic tumors, etc.) is fundamental to the correct and early treatment [10].

Many services support the idea to make a lymphatic mapping and sentinel lymph node biopsy to detect early metastasis from PASGC and this routine procedure is proving to be effective, but, as adjuvancy and other therapeutic procedures, there are no sufficient studies to prove that this routine is always beneficial and must be implemented as protocol [1–3,11].

Our observation about this case suggests just a MRI to study tumoral limits and compromised lymph nodes. The next step