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PRIMARY CUTANEOUS ADENOID CYSTIC CARCINOMA IN THE TRUNK: CASE REPORT AND LITERATURE REVIEW

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Primary cystic adenoid skin carcinoma is a rare and poorly documented neoplasm in literature worldwide, with just over 250 reports. This work describes a 52-year-old male patient, with no comorbidities, who presented this neoplasm in nodular format in the posterior thoracic region, associated with localized pain and erythema — symptoms that led him to seek medical help. The clinical findings, differential diagnosis and treatment particularities were reviewed and correlated with the clinical case. The choice of type of surgical treatment was done considering the characteristics of the primary lesion that are associated with a worse prognosis. Despite its rarity, this neoplasm is easily identified through histological examination, the correct choice of treatment and patient follow-up, essential to increase survival. Thus, this work contributes to diminish the scarcity of literature related to this topic, especially the form of treatment employed.

***Key Words*: adenoid cystic carcinoma, skin neoplasm, differential diagnosis, carcinoma, therapy.**

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The cystic adenoid carcinoma is an uncommon neoplasm, which develops in salivary glands and rarely originates in other anatomical sites [1]. When the diagnosis is performed on a skin lesion, it can only be considered a primary skin neoplasm, when the possibility of a metastatic lesion is discarded [2].

The first case of primary adenoid cystic skin carcinoma (PACSC) was documented in 1975 and, since then, just over 250 cases have been published [3, 4]. Despite the rarity of this neoplasm, the criteria used to define the diagnosis are well established in literature [5]. However, there is no consensus on the form of surgical treatment, mainly for PACSC originating in the trunk [3–5]. The purpose of this report is to describe the clinical and histopathological findings and the therapeutic option of a PACSC diagnosed in the chest region of a male patient.

Case history. A 62-year-old man, born in the western region of Santa Catarina, Brazil, with skin showing phototype III characteristics on the Fitzpatrick scale complained of a nodular lesion (+/- 1 cm in diameter), with a fast growth, where the emergence had occurred one month before. The lesion was located in the posterior region of the left thorax. He sought medical help due to localized pain and associated erythema. He had no comorbidities and a family history of colon cancer (father). He underwent antibiotic and non-steroidal anti-inflammatory treatment, due to the diagnostic hypothesis of an inflammatory lesion.

After 7 days, he was reassessed by the same doctor, and due to worsening symptoms, the excision of the lesion was the choice taken. Microscopic examination described a neoplasm with an infiltrative growth pattern, composed of basaloid cells, with little cytoplasm and uniform nuclei, and delicate nucleoli. The neoplastic cells were organized in tubules, nests and in a cribriform pattern forming cysts occupied by mucoid material. They exhibited positive PAS Alcian Blue coloring (Fig. 1). Two foci of incomplete perineural invasion were identified. The lateral surgical margins were compromised by the neoplasia. With this, the diagnosis of a cystic adenoid carcinoma was characterized.

The patient was referred to the oncology service, but he was waiting for a doctor's appointment to be scheduled. Three months later, the patient noticed a small nodule in the region of the previous scar, associated with local pain. He was then, forwarded again, with urgency, to oncology service due to the previous diagnosis and the possibility of local recurrence. Upon examination, there was a 2 cm long scar, with a small central nodular area that was palpable and painful to touch (Fig. 2, *a*, *b*). No clinically suspicious lymph nodes were found in the axillary and inguinal regions.

To assess whether the neoplasm was either a primary or a metastatic skin lesion, the patient underwent a positron emission computed tomography and laboratory tests which did not show signs of distant illness nor associated comorbidities. Since a local recurrence of a PACSC was considered, a wide resection (2 cm margin) and sentinel lymph node (SLN) biopsy was proposed. The preoperative lymphoscintigraphy exam marked an SLN in the left axilla. The anatomo-pathological examination of the previous operative site described a residual focus of cystic adenoid carcinoma next to the scar area, 0.5 cm (Fig. 3). No areas of metastasis were found in the axillary SLN, either with H&E staining or by immunohistochemistry (cocktail of cytokeratins with AE1/AE3, Flex and Dako antibody clones). The patient was examined in 6 and 12 months after surgical procedure and not presented disease local (Fig. 4) or systemic recurrence signs.

Discussion. PACSC is a neoplasm of slow growth, ranging from 10 to 25 years from the appearance of the skin lesion to the clinical manifestation that motivated the medical demand [1, 3]. It is prevalent (around 90% of cases) from 50 years of age, with an average

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Abbreviations used: PACSC – primary adenoid cystic skin carcinoma; SLN – sentinel lymph node; UFFS – Universidade Federal da Fronteira Sul.

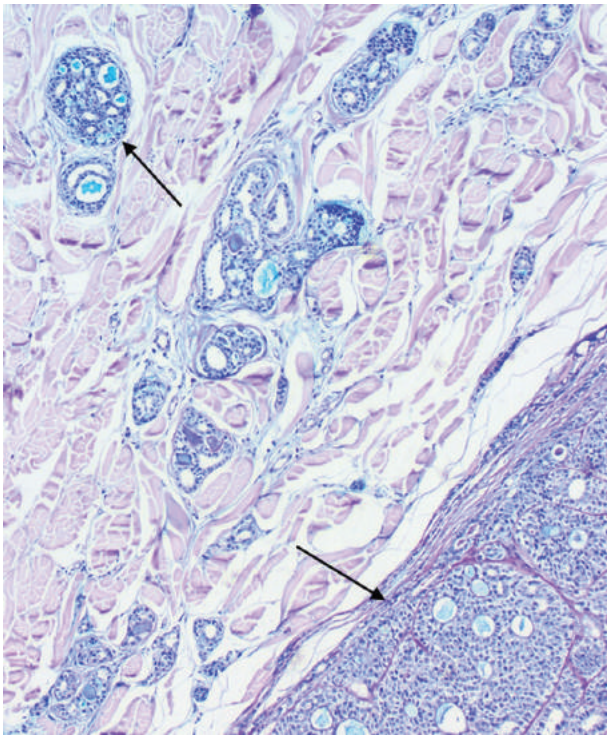

Fig. 1. Histopathological examination of PACSC (PAS Alcian Blue; ×10). Presence of cribriform areas with pseudoglandular spaces (arrows)

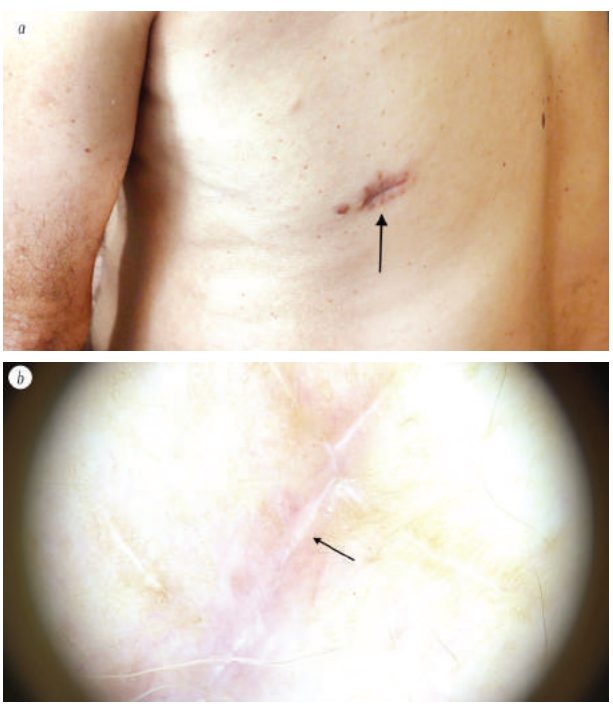

Fig. 2. Scar clinical aspect three months after the excisional biopsy (arrow) (*a*) and dermoscopy image of central area (*b*), where is possible to identify structureless white liner area (arrow) without signs of malignant patterns in the epidermis

of around 60, reaching a plateau in older ages [2, 3]. There are no prevalent differences between men and women, and it preferentially affects individuals with low phototypes. The head and neck are the most common areas where the primary lesion arises, whence the idea that the neoplasm originated in the salivary gland must be ruled out [3, 5].

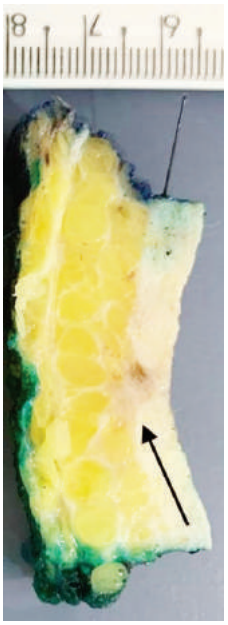

Fig. 3. Macroscopic examination of the surgical part of the margin enlargement, where it is possible to observe residual lesion between the dermis and hypodermis (arrow)

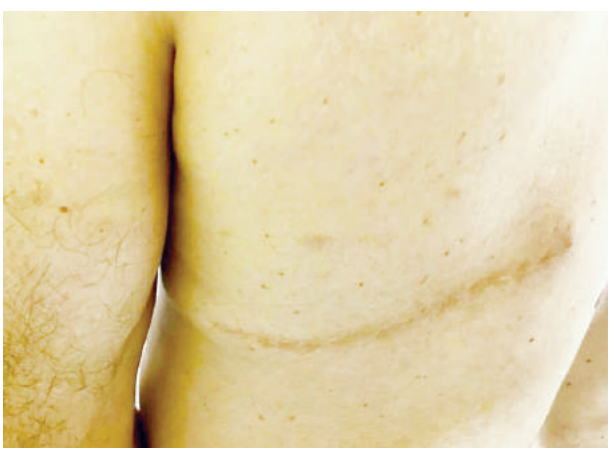

Fig. 4. Clinical aspect of the scar 12 months after the wide resection

Clinically, the primary PACSC lesion is described as a firm, subcutaneous, solitary nodule, or multiple associated nodules in an erythematous plaque. They can cause pain or discomfort, be ulcerated and cause alopecia at the primary lesion site [3, 5, 6]. The patient reported here is aged within the expected range for the development of PACSC. However, he presented a different clinical evolution, within a relatively short period from the appearance of the lesion to the first medical consultation. The fact of having associated inflammatory signs, where the pain symptom was highlighted, may have contributed to the definition of the diagnosis in an early manner.

The PACSC diagnosis is easily performed by histological examination [3]. It is a neoplasm composed of basaloid cells organized in solid and cribriform areas, where there are pseudoglandular spaces formed by the necrosis of neoplastic cells next to the basal cell mucin [5]. There may be true ductal structures with basal cuboid cells and myoepithelial cells. Epithelial cells have hyperchromatic

nuclei and are basophilic. In general, they have low mitotic activity and few atypias [5, 6].

During the investigation it is important to consider other differential diagnoses such as adenoid basal cell carcinoma, polymorphic carcinoma of the sweat gland, and primary mucinous carcinoma of the skin among others [3, 6]. In case there are doubts as to the histological examination, immunohistochemical markers may help in defining how cytokeratins AE1/AE3, 5, 5/6, 14 and 17 are positive in myoepithelial cells, and the 7 and 8/18s present in glandular luminal cells [7]. In the patient reported, the lesion was typical of PACSC, with immunohistochemical examination (cocktail of cytokeratins: AE1/AE3 antibody clones) being used only to discard micrometastases in SLN.

Regarding treatment, due to its rarity, there is no consensus on the best surgical method. However, wide margin excision (at least 2 cm) is used due to the high rate of local recurrence [1, 5]. PACSC has low metastatic potential for lymph nodes or at a distance [5]. A prevalence of regional and distant metastases ranging from 4% to 7% has been described [3, 6]. Even so, due to being a rare epithelial neoplasia, SLN biopsy technique can be considered, especially in patients with local disease recurrence, or, when there is invasion of the perineural space by neoplasia, local recurrence is estimated at 50% [1, 8]. Also, in cases of extensive skin lesions, the association of regional lymphadenectomy with the treatment of the primary lesion has been described [2].

Recurring cases are generally more aggressive, and, in many of them, the relapse occurs many years after the primary injury, which reinforces the need for clinical follow-up after treatment for early diagnosis of metastases [9]. Survival for patients who develop this neoplasm in the thoracic region is 75% in 5 years with a lower index of those with primary lesion in the head and neck area (90%) [3, 9, 10]. The option of enlarging margins with 2 cm associated with the biopsy of the SLN were adopted in the reported case, for three aspects related to a worse prognosis were considered: the localization of the primary lesion, neoplasia recurrence, and presence of foci of neoplastic cells associated with perineural spaces.

PACSC, despite being a rare neoplasm, has histological features that define diagnosis easily. Even though there is no consensus on the treatment of this disease, knowing the natural history as well as the factors related to a worse prognosis can contribute to the definition of the treatment and the type of clinical follow-up.

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ПЕРВИННА АДЕНОЇДНА КІСТОЗНА КАРЦИНОМА ШКІРИ ТУЛУБА: КЛІНІЧНИЙ ВИПАДОК ТА ОГЛЯД ЛІТЕРАТУРИ

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Первинна аденоїдна кістозна карцинома шкіри тулуба є рідкісним захворюванням. У світовій літературі нараховується не більш як 250 згадувань про нього. У цьому повідомленні наведено опис випадку хвороби у 52-річного чоловіка, у якого було виявлено нодулярну форму цього новоутворення в задній частині тулуба на рівні грудної клітки. Це новоутворення спричиняло місцевий біль і еритему, що й змусило хворого звернутися за допомогою. Наводяться клінічні дані, особливості диференційної діагностики та лікування у цьому клінічному випадку. Вибір хірургічного лікування зроблено з урахуванням особливостей первинного ураження, які асоціювалися з гіршим прогнозом. Незважаючи на те що це новоутворення виникає рідко, гістологічне дослідження дозволяє його ідентифікувати без труднощів, що важливо для вибору правильної стратегії лікування та подальшого спостереження і покращення показників виживаності. Це повідомлення має на меті зменшити прогалини в літературі з цього питання, особливо в тому, що стосується лікування пацієнтів із цією хворобою.

Ключові слова: аденоїдна кістозна карцинома, шкірні новоутворення, диференційний діагноз, рак, лікування.