SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Plastic Surgery

Primary Cutaneous Mucinous Carcinoma: A Case Report and Review of the Literature

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DOI: https://doi.org/10.36347/sasjs.2025.v11i03.006 | **Received:** 04.03.2024 | **Accepted:** 14.04.2024 | **Published:** 05.03.2025

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Abstract **Case Report**

Primary cutaneous mucinous carcinoma is a rare adnexal tumor arising from the sweat glands. It is usually located at the cephalic extremity, especially the periorbital region. We report the case of a patient who presented with primary mucinous carcinoma of my axillary region. The histological study showed a poorly differentiated and infiltrating carcinoma requiring an immunohistochemical study, which subsequently showed an appearance in favor of a malignant adnexal tumor of the primary mucinous cutaneous carcinoma type with Cytokine 7 positivity, ER at 60%, PR at 40%, androgen receptor positivity at 30% and negativity of Cytokine 20. Primary cutaneous mucinous carcinoma is difficult to differentiate from breast or gastrointestinal metastasis. The presence of carcinoma in situ or myoepithelial cells is in favor of a primary cutaneous origin. It is a slow-growing tumor, with exceptional metastasis. The treatment is surgical and the recurrence rate is very high. Through these two observations, the authors highlight the main clinical, histological and therapeutic aspects of this entity with a review of the literature.

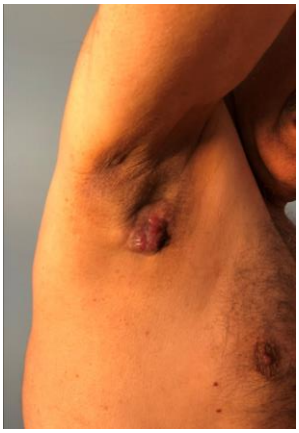
Keywords: adnexal tumor, mucinous carcinoma, Cytokine.

INTRODUCTION

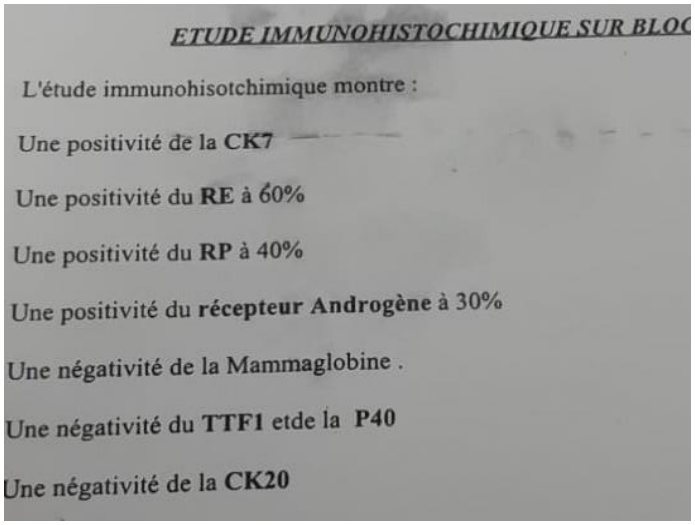
Cutaneous primary mucinous carcinoma is a rare malignancy. We report the case of a 61-year-old patient, in whom the anatomopathological study showed a primary cutaneous mucinous carcinoma. He required an extension workup (CT scan / ultrasound / endoscopy) to rule out a primary tumor, because the primary mucinous carcinoma of the skin was morphologically similar to the cutaneous metastases of a mucinous carcinoma of breast or gastrointestinal origin.

PATIENT ET OBSERVATION

The patient is a 61-year-old diabetic patient on insulin with a nodular lesion in the right axillary fossa that had been evolving for 2 years. On clinical examination, the swelling was reddish, nodular, measuring 6 cm in long axis, painless and non bleeding on contact. It was mobile in relation to the deep plane and fixed in relation to the superficial plane. With presence of palpable homolateral adenopathies.



The biopsy of the lesion revealed a dermal-hypodermal localization of a poorly differentiated and infiltrating carcinomatous proliferation. The immunohistochemical study showed CK7 positivity, ER at 60%, PR at 40% and Androgen receptor positivity at 30%.



ETUDE IMMUNOHISTOCHIMIQUE SUR BLOC

L'étude immunohisotchimique montre :

Une positivité de la **CK7**

Une positivité du **RE** à 60%

Une positivité du **RP** à 40%

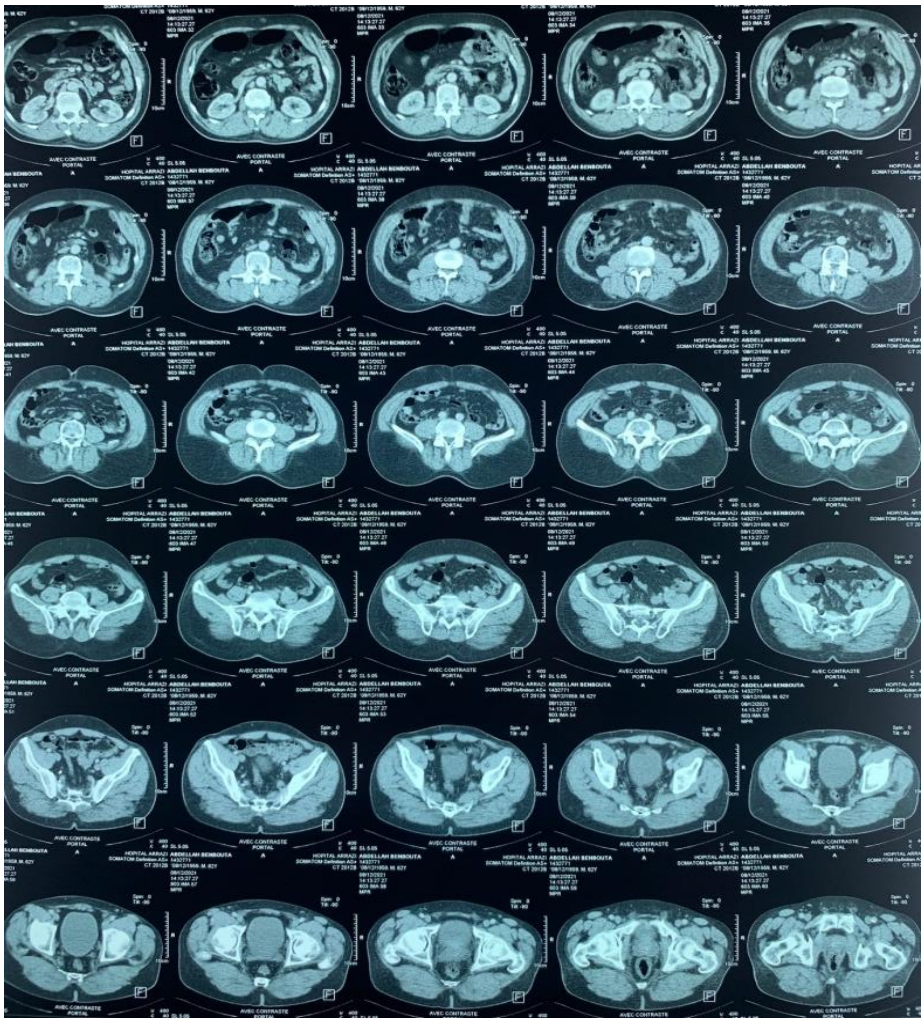
Une positivité du **récepteur Androgène** à 30%

Une négativité de la Mammaglobine .

Une négativité du **TTF1** etde la **P40**

Une négativité de la **CK20**

In order to retain the primary character, the patient had benefited from a CT scan which showed: axillary subcutaneous mass associated with homolateral axillary ADP. An endoscopy with gastric biopsy having revealed no abnormalities.



Based on these histological, immunohistochemical, radiological and fibroscopic data, the diagnosis of primary cutaneous mucinous carcinoma was retained.

DISCUSSION

Primary cutaneous mucinous carcinoma is a very rare adnexal tumor of the sweat glands. It occurs on average between 50 and 60 years of age, with extremes ranging from 8 to 84 years, with a male predominance (sex ratio of 2:1) [3]. It affects the cervicofacial region in 80% of cases, particularly the face and scalp, with a predilection for the periorbital region, which is affected in 40% of cases, but all locations have been reported [7]. The apocrine or eccrine origin is still debated.

Primary cutaneous mucinous carcinoma has a morphological similarity with cutaneous metastases of mucinous carcinoma of predominantly breast or gastrointestinal origin [8]. Differentiating a primary cutaneous mucinous carcinoma from a metastasis is histologically difficult. This must be based on an exhaustive clinical investigation, nevertheless some histological and immunohistochemical features allow to orientate towards the primary nature [8].

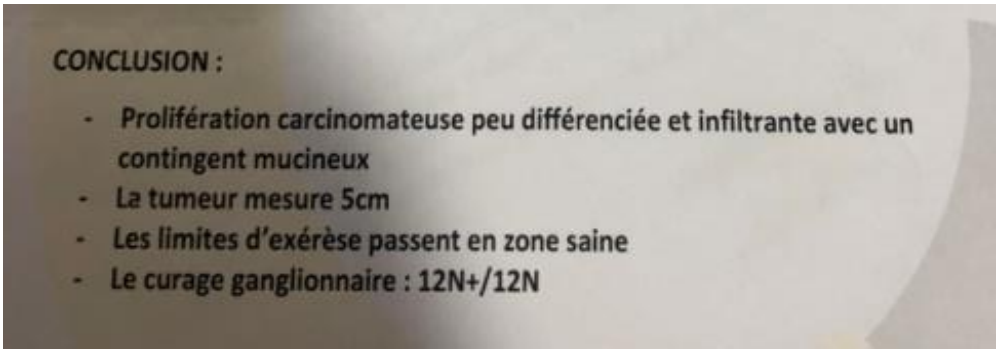
Metastases from mucinous colorectal adenocarcinomas localize to the abdominal wall, whereas breast carcinomas reach the chest wall [4]. In a large study of skin metastases, Brownsten et al found that only 6% of cases had a metastasis localized to the face [4]. Some authors base this on the fact that in the secondary tumor, more numerous and atypical tumor cells, mucus puddles are poorly represented and septa are rare [8]. Its usual clinical presentation is that of a solitary, asymmetric, nodular, flesh-colored, painless lesion, with a mean size of 2cm and slow growth [8], but rapidly progressive forms have been reported [4].

Histologically, it is a dermohypodermal tumor separated by fibrous septa with trabeculae, clusters and cribriform masses within a mucoid substance. The tumor cells are cohesive, cubic with a sometimes vacuolated cytoplasm. The nucleus is central, vesicular and not very atypical. Mitoses are rare or absent [3]. On immunohistochemical examination, primary mucinous carcinoma of the skin expresses low molecular weight cytokeratins, EMA, CEA (carcinoembryonic antigen), GCDFP-15 (gross cystic disease fluid protein) and rarely S100 protein, whereas cytokeratin 20 is always negative, thus eliminating a digestive origin [3]. Some authors have reported a positivity of primary carcinoma cells for hormone receptors underlining their similarity with carcinoma cells of breast origin [8].

Our case expressed cytokeratin 7, EMA and cytokeratin 20 was negative. Qureshi et al suggest that the search for an in situ component points to the primary cutaneous adnexal origin, such as atypical intracanal hyperplasia lesions to ductal carcinoma in situ [9]. The demonstration of myoepithelial cells by immunostaining with P63, cytokeratin 5/6, or smooth muscle actin may be contributory to the diagnosis [10]. The in situ component was observed in only one case and the primary origin was retained. In the absence of this in situ component, confrontation with clinical and paraclinical data is necessary [8].

The treatment is surgical, with wide margins, or surgery according to the Mohs technique [11]. Regional lymph node dissection is necessary if there are palpable adenopathies.

Our patient underwent a large excision with axillary curage and the peripheral surgical borders were healthy. The local recurrence rate is nearly 50% [12] and distant metastases occur in 2.7% [14]. Our patient presents a classic form of primary cutaneous mucinous carcinoma, successfully treated by an extended excision with axillary curage.



CONCLUSION

Primary mucinous carcinoma is a low-grade malignant adnexal tumor with low metastatic potential and a very high recurrence rate.

It can simulate a metastasis of breast or digestive origin, hence the need to compare with clinical and paraclinical data.

The management is surgical based on extended excision with curage in case of adenoapathy.

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