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Primary ductal carcinoma of ectopic breast[☆]

*Dear Editor,*

Ectopic breast carcinoma accounts for approximately 0.3% to 0.6% of all breast cancers, with 95% arising from aberrant breast tissue.^{1,2} Clinical diagnosis may be delayed due to the atypical location, similarity to other diseases, and flawed laboratory tests.³ This case report describes a patient with primary ductal carcinoma of the ectopic breast in the axilla, diagnosed and treated as hidradenitis suppurativa (HS).

A 62-year-old female patient presented with an erythematous, well-defined nodule measuring 1.5×2.0 cm, with a retracted center, painful and hard on palpation in the left axilla (Fig. 1).

The clinical diagnosis suggestive of HS was confirmed by ultrasound, which showed an area of subcutaneous hypoechoic thickening and a nodular area. Topical and oral antibiotics were prescribed for 14 days, in addition to intralesional corticosteroid injections, without improvement.

Two 0.5 cm elliptical incisional biopsies were performed and anatomopathological analysis showed preserved epidermis and the presence of rows and clusters of atypical epithelial cells in the dermis (Fig. 2), while the immunohistochemistry showed positivity for pankeratin, suggestive of cutaneous metastasis.

The neoplastic screening did not disclose a primary site. Therefore, total excision of the lesion was performed, which showed dermal infiltration of carcinomatous cells over ectopic mammary gland tissue (Fig. 3). Immunohistochemistry was positive for estrogen and negative for progesterone and human epidermal growth factor receptor-type 2 (HER-2). It was concluded that it was a primary ectopic breast carcinoma with characteristics of invasive ductal carcinoma.

The patient was referred to the mastology and oncology service, where surgical margins were enlarged and an ipsilateral axillary lymphadenectomy was performed, due to the presence of lymph node metastasis. She underwent complementary treatment with radiotherapy and anastrozole. There has been no recurrence one year on outpatient follow-up and control mammography.

Ectopic breast tissue is subject to the same pathophysiological processes as the topical breast, but malignant changes are more frequent than benign ones.¹ It can consist of glandular tissue, nipple and areola. It is subdivided into

supernumerary breast or aberrant breast tissue. The latter is characterized by the presence of an isolated mammary gland, close to the topical breast and without communication with overlying skin.¹

Ectopic breast cancer predominates in the female sex. The axillary region is most frequently affected and infiltrating ductal carcinoma accounts for 79% of cases.^{1,4} The most common clinical manifestation is the presence of a unilateral, subcutaneous, irregular, erythematous, indurated nodule showing progressive growth, with or without a nipple and areola.⁵ Ultrasonography is the initial preferential examination, which may show an irregular, hypoechoic, heterogeneous, poorly-defined nodule; the accessory mammary gland may be detected.⁵ The diagnosis is usually a late one, with an average delay of 40 months, which can lead to worse prognosis. The confirmation is achieved with histopathological analysis.^{3,5} The management of ectopic breast carcinoma follows conventional breast cancer treatment and staging. There is no consensus on the prophylactic excision of the ectopic breast tissue.^{3,5}

Although it is a rare condition, it is essential to recognize it and consider tumors along the milk line. It is important to emphasize the importance of the histopathological analysis, even in the presence of a probable benign lesion.

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Figure 1 Erythematous, mobile nodule with retraction area.

[☆] Study conducted at the Hospital Federal de Bonsucesso, Rio de Janeiro, RJ, Brazil.

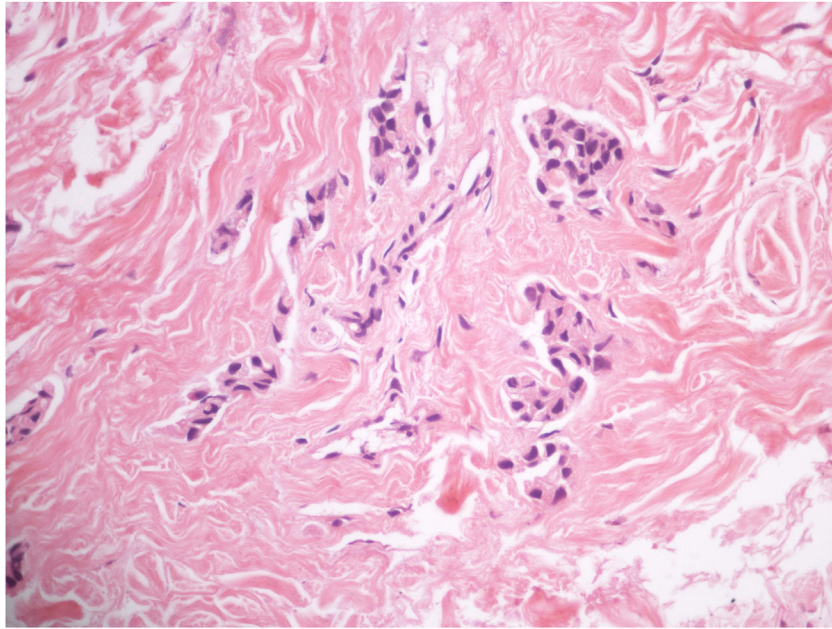


Figure 2 Histopathology of the incisional biopsy showing epithelial aggregates of atypical cells (Hematoxylin & eosin, $\times 400$).

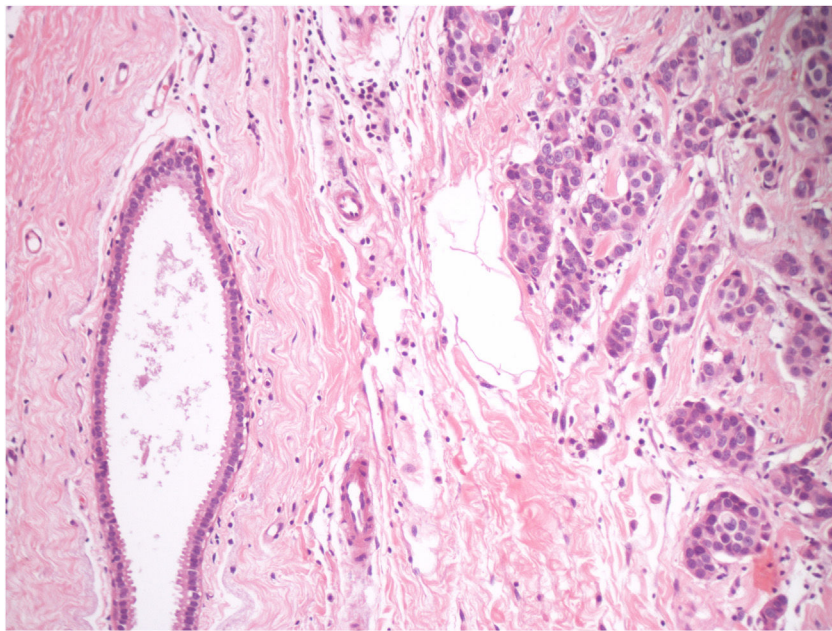


Figure 3 Histopathology of the excisional biopsy showing ectopic mammary glandular tissue and dermal infiltration by carcinomatous cells (Hematoxylin & eosin, $\times 400$).

Authors' contributions

Ariane Sponchiado Assoni: Design and planning of the study; drafting and editing of the manuscript; critical review of the manuscript.

Beatriz Baptista Abreu da Silva: Design and planning of the study; drafting and editing of the manuscript; critical review of the literature.

Aline Sponchiado Assoni: Drafting and editing of the manuscript; critical review of the literature.





Felipe Mauricio Soeiro Sampaio: Approval of the final version of the manuscript; effective participation in research orientation; critical review of the manuscript.

Conflicts of interest

None declared.

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Spontaneous regression of Merkel cell carcinoma with positive detection of Merkel cell polyomavirus by PCR and immunohistochemistry[☆]



Dear Editor,

Merkel cell carcinoma (MCC) is a rare cutaneous neoplasm, characterized by the proliferation of anaplastic cells, with an aggressive clinical course. It is more frequently diagnosed in caucasian males after the seventh decade of life and in immunosuppressed individuals.¹

In 2008, Feng et al. observed the DNA of a new polyomavirus in 8 of 10 MCCs, named Merkel cell polyomavirus (MCPyV). The viral DNA was integrated into the DNA of the tumor cells in a clonal pattern, suggesting that the viral infection preceded the clonal expansion of these cells.²

A 76-year-old patient reported fast-growing nodules on the leg, with eight weeks of evolution. The physical examination showed a firm, erythematous, semispherical nodule measuring 4 cm on the left leg, surrounded by similar satellite lesions (Fig. 1A). These findings regressed considerably three weeks after a shave biopsy of the main lesion was performed (Fig. 1B).

Histopathological analysis showed a dermal tumor with extensive proliferation of small basophilic cells, with large, ovoid, hyperchromatic nucleoli and finely dispersed chromatin (Fig. 2A). Immunohistochemistry was positive for CK20, with a perinuclear, dot-like pattern, and chromogranin A (Fig. 2B,C), and negative for TTF-1 and CK7, confirming the diagnosis of MCC. MCPyV DNA was detected by PCR and the major viral T-antigen was detected by nuclear positivity in immunohistochemistry using CM2B mon-

oclonal antibody (Fig. 2D). Despite the observed partial regression, surgical excision was performed with wide margins, and there was no recurrence of the condition after two years of follow-up. Histopathology of the surgical specimen revealed residual neoplasia circumscribed by connective tissue strands and dermal fibrosis.

Although there is no standard protocol, the treatment is based on the excision with wide margins for localized or locoregional disease, with adjuvant radiotherapy for large tumors. When there are distant metastases, radiotherapy and adjuvant chemotherapy are combined.¹

The pathogenesis of MCC is considered multifactorial. Studies have reported a P53 mutation and high levels of bcl2 proto-oncogene expression in tumor cells, supporting rapid tumor expansion and growth.^{1,3}

Spontaneous regression of MCC is rare and was described in 1986, with fewer than 40 similar cases being reported since then. Regressions after biopsy or incomplete excision have also been described and may be due to the activation of the T-cell-mediated immune response after surgical trauma, although the exact mechanisms remain unknown.⁴

Unlike melanoma cases, the reported cases of MCC with spontaneous regression usually had a better prognosis and progressed to cure.⁵

The presence of MCPyV in MCC is thought to stimulate the triggering of an immune response against viral antigens and tumor cells.⁵ Considering the presence of MCPyV in the present report, it is postulated that viral antigen exposure after the biopsy may have triggered host immune activation and tumor regression.

In conclusion, the present report aims to draw attention to the rare possibility of spontaneous regression of MCC and its association with MCPyV.

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