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Abrikossof's Tumor: Report of a Case

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Abstract

The granular cell tumor of the breast is a rare tumor and a usually benign disease that appears clinically and radiologically like a malign tumor. We report a case of GCT in a woman of 41 years, who present a clinically and radiologically suspect later. The anatomopathological examination showed the benign tumor proliferation and a large tumorectomy was realized. The immunostaining with PS100 and NSE protein confirm the nature of the tumor.

Keywords

Abrikossof, Breast, Benign

1. Introduction

Granular cell tumor (GCT) of the breast or Abrikossof's tumor is a benign conjunctive proliferation of cells with a probable schwannian neurogenic origin [1] [2] [3].

It is a rare tumor first described in 1926 by Abrikossof occurring most often in black women aged 20 - 60 years [1] [2] [3]. The localization is ubiquitous with the oral cavity, head and neck cells but also the breast as the main localizations [1] [4] [5]. It is a benign tumor, most often asymptomatic with occasional inflammatory flare-ups [1]. The diagnosis is made by histology with additional immunohistochemical examination [2]. There is often a discrepancy between the clinical aspect, imaging and histological examination of the lesion, which may lead to the suspicion of a suspicious lesion [2]. Their malignancy is exceptional (1% to 3% of breast GCTs) with variable malignancy criteria. For Jardines, only the presence of metastases with anatomopathological characteristics similar

to those of the primary tumor can confirm the malignant character [2]. The management is usually surgical, consisting of a wide resection with a favorable evolution in case of complete resection [6] [7].

We report the case of a granular cell tumor in its breast location.

2. Observation

We report the case of a 41-year-old female patient, multigestate, with no personal or family history of breast pathologies, who consulted for a nodule of the right breast associated with mastodynia that had been progressing for more than one year, discovered by self-examination.

The physical examination revealed a single nodule in the inferolateral quadrant of the right breast, 2 cm in circumference, close to the sub mammary fold, attached to the superficial indurated plane, without inflammatory signs. The lymph nodes were free.

Mammography with complementary ultrasound showed a mass of irregular shape and contours, with a hyperechoic linear center with attenuation of posterior echoes, measuring 7.6 mm * 5.7 mm of the right inferolateral quadrant of the breasts classified ACR3.

Surveillance was recommended with a follow-up mammogram one year later, which revealed a focal solid image with spiculated contours measuring 13.7 mm * 10.4 mm * 10.7 mm located in the inferolateral quadrant of the right breast, attached to the superficial plane without any thickening of the skin covering and with a small homolateral inflammatory axillary lymph node. This nodule was classified ACR4 and a microbiopsy of the mass was performed (Figure 1). Histological examination showed a benign mesenchymal proliferation of histiocytic cells with abundant eosinophilic granular cytoplasm centered by a pycnotic nucleus in favor of an abrikossof tumor.

The patient had had a large tumorectomy (Figure 2).

The final histological examination showed a benign tumor proliferation. It

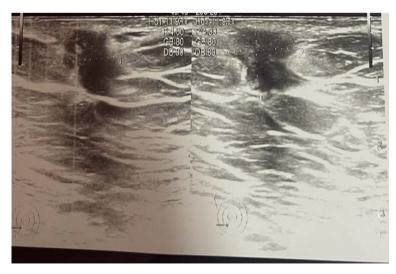


Figure 1. Ultrasound appearance of Abrikossof's tumor.

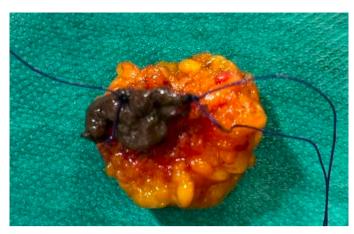


Figure 2. Intraoperative view of the breast mass after removal with resection margins in the healthy zone.

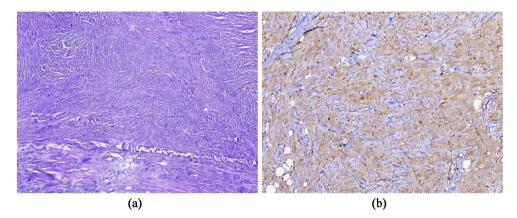


Figure 3. View of histological blade (a) high magnification GCT HE staining ×400; (b) high magnification GCT immunostaining PS100.

was formed by large polygonal cells with abundant eosinophilic granular cytoplasm and moderately atypical nuclei, arranged in sheets infiltrating the breast adipose tissue. The resection margins were healthy.

The immunohistochemical study showed diffuse positivity of PS100 and CD68 antibodies on the tumor cells, confirming a granular cell tumor (**Figure 3**).

3. Discussion

The histogenesis of Abrikossoff's tumor has long been controversial. When Abrikossoff first described this tumor in Moscow in 1926, he thought it was a degeneration of striated muscle fibers and renamed it "granular cell myoblastoma" in 1931 [8]. In 1949, Frest and Custer found myelin degradation products in granules suggesting a nervous origin of this tumor; they then reconsidered it as a granular cell neurofibroma. Fischler and Wechsler in 1962, after electronic and histochemical studies, considered that the tumor was derived from Schwann cells and that the granules reflected altered hyaline. The latest studies using immunohistochemistry show a constant marking by the S100 protein, in favor of a

Schwanian origin of this tumor. It would appear to be a tumor of nervous origin. Currently, the nervous origin is well established thanks to ultrastructural studies (lysosomal nature of the intracytoplasmic granules and sometimes presence of myelin bodies) and immunohistochemical studies (tumor cells expressing neurogenic markers: PS100 and NSE) [2] [8].

Abrikossoff's tumor is most often found in African-American women who are not postmenopausal, with an average age of 38 years [2].

The most common way to discover this tumor is by self-examination of a painless nodule in the breast. The clinical presentation is that of a nodule without particular aspect, centimetric, and most often not exceeding 3 cm, of slow growth. It is asymptomatic or more rarely painful, with a hard consistency [9]. It is ubiquitous and occurs more frequently on a mucous membrane (oral, digestive, respiratory, genital) than on the skin. Most often unique and benign, it can be multifocal in 3% to 25% of cases [8].

There are few cases of malignant GCTs of the breast in the literature (1% - 2% of GCTs of the breast). Malignant GCTs are often larger and faster growing, and the occurrence of metastases remains one of the most important criteria for defining malignancy [10]. Gomard-Menesson *et al.* described a case of a benign granular cell tumor that undergoes malignant transformation after a 7-year free interval [4]. For Jardines *et al.* only the presence of metastases with anatomopathological characteristics similar to those of the primary tumor can affirm the malignant character.

The mammographic features of GCT are often suggestive of a malignancy. The most common characteristics are a solid, heterogeneous image, with deep development, vascularization, especially in the periphery [10]. Magnetic resonance imaging with gadolinium injection can help differentiate between malignant and benign GCTs. Malignant GCTs rapidly take up the contrast medium in the first minute after injection, with a plateau as early as the second minute because of the greater vascularity. Contrast with a peripheral corona is frequently found and may suggest malignancy [10].

Ultrasound microbiopsies or biopsy-exereses provide a histological diagnosis of certitude. Anatomopathological examination reveals large polygonal cells with fragile membranes and abundant eosinophilic, finely granular cytoplasm [2]. The immunohistochemical study confirms the diagnosis. The tumor cells are positive for neurogenic markers (PS100 and NSE) [10].

The treatment is surgical, consisting of a wide excision without associated axillary curage [2]. It allows a diagnosis of certainty, to appreciate the surgical limits and to search for possible criteria of malignancy. The prognosis is often favorable. However, the evolutionary follow-up can be enmeshed of local recurrence or secondary metastatic localization particularly pulmonary or bone. The complementary and systematic analysis of a cell proliferation index (Ki67) could be a useful aid to define the presence of this evolutionary turning point [2] [4] [8] [10].

Aggressive surgical treatment, including when there are several pulmonary metastases, associated with a large lymph node curage is recommended by most authors in the management of malignant forms. The value of adjuvant radiotherapy is much debated and has been proposed by some authors. Like chemotherapy, it has not been proven to be effective [4].

4. Conclusions

Abrikossof's tumor is a rare and mostly benign tumor that may be suggestive of a malignant breast tumor.

Surgical excision must be complete and long-term surveillance must be carried out, especially if the proliferation index is high, in order to watch for a potentially malignant tumor.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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