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Paget's Disease of the Breast. Presentation of a Case

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Introduction

Paget's disease (PD) of the breast (also known as Paget's disease of the nipple and Paget's disease of the breast) is a rare type of cancer that affects the skin of the nipple and usually the darker circle of skin on the nipple. around it which is called areola. (1)

PD can be located in the breast and extramammary. (2) Paget's disease of the breast is named after the 19th century British physician, Sir James Paget, who, in 1874, observed a relationship between nipple changes and breast cancer. (2,3)

The average age of onset is over 50 years and constitutes 2% of all types of cancer that appear in the breast.(2,3,4) It is described that an underlying breast cancer can be present up to the age of 97% of cases, generally without a palpable mass or associated mammographic abnormality. PD is more frequent in postmenopausal women, with a higher incidence between the sixth and seventh decade of life, unlike invasive breast carcinoma, whose maximum incidence is 5 to 10 years later. (5)

In our country, breast cancer occurs with a rate of 146.1 per 100,000 women. In 2019 It was found that the incidence of Paget's disease in the province of Matanzas was 1.03% among 586 patients treated for breast cancer between 2015 to 2021 with an average age of 60 years. During the same period, our hospital has not had any patient with this diagnosis.

Clinically it is described as a type of eczematous lesion that is located on the nipple, initially as a dry eczema with a thick scab that later vacuolizes and when detached leaves an erosion that slowly invades the nipple and later progresses to the areola in surface and to the terminal lactiferous ducts in depth, conditioning the destruction of these structures. (2,3,4)

In the progression of the malignant invasion towards the interior of the breast, a tumor with the clinical characteristics of breast cancer is produced, which in the initial stages is difficult to palpate although it exists in an incipient form in the ducts, until the evolution of the disease allows clinical identification when its volume increases. (23)

Doctors don't fully understand what causes Paget's disease of the breast. The most widely accepted theory is that cancer cells from a tumor inside the breast travel through the milk ducts to the nipple and areola. This would explain why Paget's disease of the breast and tumors within the same breast are almost always found together. A second theory is that the cells in the nipple or areola become cancerous on their own. This would explain why some people get Paget's disease of the breast without having a tumor inside the breast. (1,6)

Because the early symptoms of Paget's disease of the breast can suggest a benign skin condition, and because the disease is rare, the initial diagnosis may be wrong. People with Paget's disease of the breast often have had symptoms for several months before receiving the correct diagnosis. (1.5)

These symptoms may be the following:

-itching, tingling, or redness of the nipple or areola

scaly, crusty, or thickened skin on or around the nipple

-flattened nipple

-discharge from the nipple that may be yellowish or bloody

The definitive diagnosis is made by excision biopsy of a fragment of the nipple where the histopathologist can see the typical Paget cells.(2,3,4) (Annex #1)

We present the case of a Paget's Disease of the Breast, attended by the Gynecology service of the Provincial Gynecology-obstetric Hospital of Matanzas in the year 2023. All the information about the case is found in the patient's clinical history and was used previously. informed consent.

Presentation of a case: Paget's disease of the breast

NETH patient, 75-year-old female, with personal pathological history of Arterial Hypertension with stable treatment with Enalapril (20mg) half a tablet in the morning and one tablet at night orally. Obstetric history of gestations 1, deliveries 1 eutocic. Menopause at age 51. He attended the consultation because it began about a year ago with increased volume and discharge from the right nipple and several months ago he noticed an ulcerated lesion on the nipple of said breast (Annex #2), for which it was decided to admit him for study and treatment.

Physical exam:

Temperature 360C

mucosa Moist and normal colored

respiratory system Normal breath sounds, no rales

Respiratory rate: 16 x '

cardiovascular system Rhythmic heart sounds, good tone, no murmur.

Heart rate: 78 x '

Blood pressure: 120/80 mmHg

Breast Right: ulcerated area is observed that compromises more than half of the nipple, the rest of the skin is normal, smooth, painless, and there is no palpable mass.

Breast Left: with good nipple-areola relationship, soft, not painful, no palpable mass

Abdomen Soft, depressible, not painful on superficial or deep palpation, no visceromegaly.

Airborne noise present.

TCS Not infiltrated.

genitourinary system Without modifications

Endocrine system Without modifications

Central Nervous System Patient oriented in time, place and person, not meningeal signs

Complementary exams:

Hemoglobin: 128 g/l

Hematocrit: 0.42

Leukogram: 6.9x109/l

Neutrophils: 0.61

Lymphocytes: 0.39

Blood glucose: 5.6 mmol/l

Clotting time: 7 min

Bleeding time: 1 min

Group and factor: O positive

Creatinine: 64umol/l

Imaging exams:

Ultrasound: symmetrical breasts with heterogeneous breast pattern with a predominance of fatty tissue, no nodules, no cysts, no distortion of breast tissue. BI RADS cat I.

Mammography: small symmetrical breasts with a heterogeneous pattern with a predominance of fatty tissue without macro or microcalcifications, no tumor, no distortion of the breast pattern. BI RADS CAT I.

Conduct to follow: Admission for surgical medical treatment.

During admission, excision of the area affected by the ulcer and paraffin biopsy were performed, which reported Paget's disease of the nipple associated with intraductal comedo carcinoma, periductal granulomatous inflammatory response.

Diagnostic impression: Paget's disease of the nipple,

Arterial hypertension.

With the same preoperative diagnosis, it was decided to perform conservative surgery on the right breast plus axillary dissection, sending pieces to the Pathology laboratory.

The total surgical time was 1 hour and 20 minutes and the postoperative diagnosis remained Paget's disease of the nipple.

Her postoperative evolution was good, and she was discharged on the 3rd day of her intervention.

Pathological anatomy:

The definitive histopathological diagnosis was intraductal carcinoma.

Macroscopic features:

The hallmark of PD is the presence of intraepithelial adenocarcinoma cells (Paget cells) either singly or in groups within the epidermis of the teat. (2,3,4)

Microscopically, it is a neoplastic lesion of the squamous epithelium of the skin of the areola and/or nipple, which presents cells with broad pale and clear cytoplasm with enlarged and prominent nucleoli, a displaced atypical nucleus, and which are located in the epidermis along the entire length of the epidermis. basement membrane, which characterize it and are known as Paget cells

The patient was sent for follow-up by the multidisciplinary mastology group, with the application of postoperative oncospecific treatment. She has evolved satisfactorily, and is currently 7 months after surgery alive.

Discussion

The average age of onset of this special variety of cancer is over 50 years. 50% present an underlying palpable mass, in 95% of cases there is an associated carcinoma in the breast or nipple. (1,6)

There are 2 theories about the origin of Paget's cells:

Epidermotropic theory: states that Paget's cells are intraductal carcinoma cells that migrate from the teat ducts through the basement membrane to reach the epidermis.

In situ transformation theory: postulates that Paget's cells are keratinocytes that transform and are considered as independent carcinoma in situ.

Today the most accepted theory is the first and it is even known that there is a factor that causes the mobilization of the cells of the glandular ducts towards the epidermis. This factor called heregulin-a is produced by normal epidermal keratinocytes, where Paget's cells express the heregullin factor receptor (HER2/NEU) to bind to keratinocytes and thus migrate and infiltrate the epidermis. (7)

PD usually presents as an ulcerative lesion of the nipple-areola complex, which causes patients to be treated ignoring the possibility of a neoplastic origin. When it is suspected, it is necessary to perform additional tests to rule out other diagnostic possibilities: Mammography, breast ultrasound, hematological tests.

The differential diagnosis includes benign dermatological alterations such as eczema of the nipple, contact dermatitis, lichen simplex chronicus, adenoma of the nipple, psoriasis and some malignancies such as Bowen's disease, basal cell carcinoma and in patients with pigmentation, malignant melanoma. (8)

The diagnosis of Paget's disease of the breast was confirmed by nipple skin biopsy, considered the study of choice, which also includes evaluation of the milk ducts.(9) Initially, in the case reported here, mammography was requested, which is considered the radiological study of choice to detect any invasive carcinoma but with limitations for intraductal in situ. (8,9) the sensitivity of mammography for the detection of breast neoplasms without a palpable mass is 50% (10).

Ultrasound represents a complementary study to detect mammographic findings or a negative result. (9) In our case, an ultrasound was requested with a negative result.

For many years, mastectomy, with or without axillary lymph node dissection on the same side of the chest, was considered the standard surgery for Paget's disease of the breast (9,11,12). This type of surgery was performed because patients with Paget's disease of the breast were almost always found to be associated with multicentric carcinoma (11,12). It is currently performed in patients with ductal carcinoma in situ of more than 2cm in diameter or when it is impossible to be certain that negative margins can be reached due to extension (13).

However, studies have shown that breast-conserving surgery, which includes removal of the nipple and areola, followed by radiation therapy to the whole breast, is a safe option for people with Paget's disease of the breast who do not have a lump. palpable in the breast or whose mammograms do not show a tumor (11). After reviewing the case studies presented here, it was decided to perform conservative surgery.

Several authors are in favor of conservative surgery (9), and suggest it as an alternative treatment for women with PD and intraductal cancer in situ. (eleven)

Prognostic factors include: coexistence of a palpable mass, lymph node involvement, tumor enlargement, invasive disease, age greater than 60 years, and stage II disease (14).

The patient in this study corresponded to the Tis, N0 M0, clinical stage 0 classification. Today she is being treated and monitored by specialists from the Oncology service.

Conclusion

PD represents a rare condition, which should be considered in the differential diagnosis of benign lesions of the nipple-areola complex and whose manifestation implies carrying out the necessary studies to rule out the diagnosis of breast carcinoma.

The complementary study of greatest diagnostic value is mammography. Ultrasonography often fails to report changes underlying a nipple-areola complex lesion.

Currently, the most accepted treatment is breast-conserving surgery, which includes removal of the nipple and areola, followed by radiotherapy.

The definitive diagnosis is by biopsy of the lesion and the presence of Paget's cells is a pathognomonic sign.

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