

Metachronous Paget's disease of the breast: case report

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SUMMARY: Metachronous Paget's disease of the breast: case report.

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Paget breast disease is a kind of intraductal carcinoma that through an intracanalicular diffusion invades the basal epidermal layer, reaching the areola and nipple, producing a typical erythematous desquamative eczematous-like lesion. This neoplasia can remain undetected for a long time and inadequately treated as a dermatological affection. Synchronous or metachronous lesions are very uncommon.

Surgical choice is conditioned by the presence of a tumor below the epidermal lesion, by its dimensions, and by the possible lymph node involvement. Surgical therapy can be radical or conservative.

From our experience we think that lesion biopsy is always necessary to formulate a correct diagnosis and to schedule an appropriate therapeutic approach. In our case, a biopsy was performed first, then on the basis of the frozen section analysis a radical mastectomy with axillary third level lymph nodes dissection, because of the large dimensions of the lesion and the previous history of a methachronous lesion.

RIASSUNTO: Malattia di Paget metacrona della mammella: caso clinico.

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La malattia di Paget della mammella è un carcinoma intraduttale che attraverso la diffusione intracanalicolare invade lo strato basale epidermico, raggiungendo l'areola e il capezzolo e producendo lesioni eritematose desquamative simileeczematose. La neoplasia può rimanere per lungo tempo misconosciuta e inadeguatamente trattata, spesso come una patologia dermatologica. Lesioni sincrone o metacrone sono molto rare.

La scelta chirurgica è condizionata dalla presenza del tumore al di sotto della lesione epidermica, dalle sue dimensioni e dal possibile coinvolgimento linfonodale. La terapia chirurgica può essere radicale o conservativa.

Dalla nostra esperienza riteniamo che la biopsia della lesione sia sempre necessaria per formulare una corretta diagnosi e programmare un adeguato approccio terapeutico. Nel nostro caso, in primo luogo è stata eseguita una biopsia, quindi, sulla base dell'esame istologico, una mastectomia radicale, con dissezione dei linfonodi ascellari di terzo livello, a causa delle grandi dimensioni della lesione e della storia di una pregressa lesione metacrona.

KEY WORDS: Paget disease - Breast cancer - Surgery.
Malattia di Paget - Cancro della mammella - Chirurgia.

Introduction

Paget disease, described by James Paget in 1874 (1), is a carcinoma developing from a mammary duct (invasive ductal carcinoma), that has a specific affinity to the nipple and areolar skin so that it has been called "intra-

ductal epidermotropic breast cancer" (Tab. 1) (2). The tumor grows along the galatophorous ducts to the areola and nipple area producing a typical erythematous, crustous and eczematous-like area that grows into the skin infiltrating the basal layer and looks like hanging drops. The cells contain a lot of mucopolysaccharides that can be recognized microscopically by a surrounding light halo, thereby distinguishing them from the skin cells. Sometimes the disease is not related to an underlying nodule and in these cases the symptomatology can feign a dermatologic affection that could lead the physician to treat it in the wrong way for a long time (2).

TABLE 1 - WHO HISTOLOGICAL BREAST CANCER CLASSIFICATION.

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| <p>1. NON INVASIVE</p> <p>a. Intraductal</p> <p>b. Lobular carcinoma in situ</p> <p>2. INVASIVE</p> <p>a. Invasive ductal carcinoma</p> <p>b. Invasive ductal carcinoma with a predominant intraductal component</p> <p>c. Invasive lobular carcinoma</p> <p>d. Mucinous carcinoma</p> <p>e. Medullary carcinoma</p> <p>f. Papillary carcinoma</p> <p>g. Tubular carcinoma</p> <p>h. Adenoid cystic carcinoma</p> <p>i. Secretory (juvenile) carcinoma</p> <p>j. Apocrine carcinoma</p> <p>k. Carcinoma with metaplasia</p> <p> I. squamous type</p> <p> II. spindle-cell type</p> <p> III. cartilaginous and osseous type</p> <p> IV. mixed type</p> <p>l. Others</p> |
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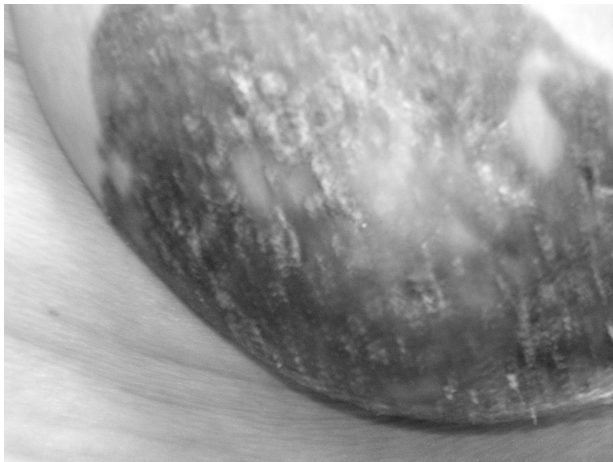


Fig. 1 - Paget's disease of the left breast.

Case report

A 73-year old woman came to our clinic for an erythematous microulcerated lesion of nipple, areola and periareolar skin of the left breast (Fig. 1). The patient referred that the lesion was seven years old and it had enlarged in the last two months. The patient had undergone a hysterectomy when she was 43 years old and a radical right mastectomy at 61 yrs for Paget-associated ductal carcinoma (Fig. 2).

Mammography showed a mammary fibroadipose involution

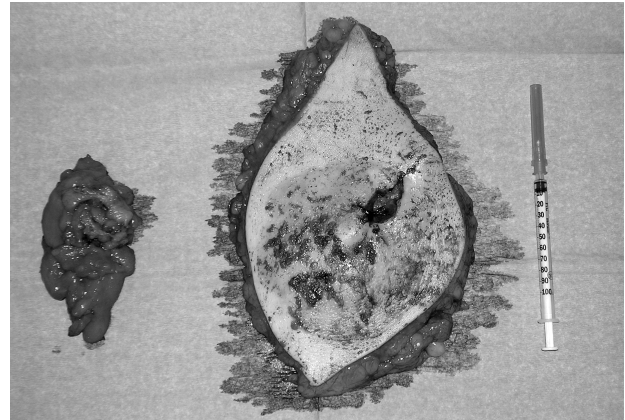


Fig. 2 - Specimen.

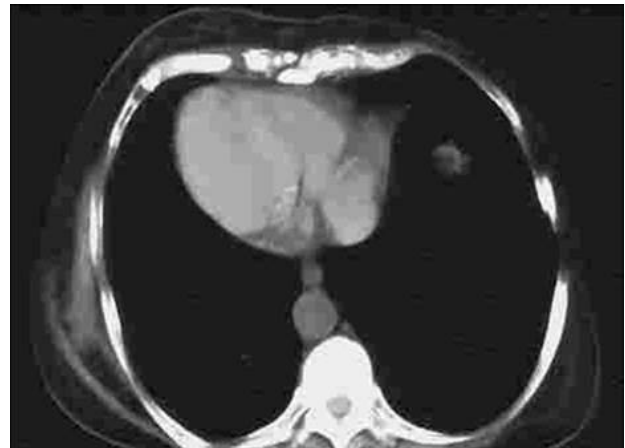


Fig. 3 - Preoperative TC.

with no focused lesions or microcalcifications, but with the axillary nodes enlarged. US scan confirmed the mammography report. CT scan showed an increase in tissue thickness in the mammary extension up to the axillary area of the left breast (Fig. 3). The bone nuclear study revealed no involvement. Tumoral markers were negative.

The patient underwent skin biopsy that revealed "invasive ductal undifferentiated Paget-associated breast carcinoma (pT4b G3)"; immunohistochemical analysis was positive for CK-LMV and CEA, Ki-67 (pos 50%), c-erbB2 (3+).

The patient underwent a radical mastectomy with third level lymph node dissection (Madden). The postoperative course was complication-free and the histology of the specimen confirms the biopsy. The final diagnosis was intraductal carcinoma with residual Paget focus, with metastasis-free regional resected lymph nodes.

Discussion

Paget disease represents 1-4% of all breast cancer (3), appearing frequently in the 5th-6th decade of life (4); synchronous (5) and metachronous Paget cases are very rare (6). Diagnosis is reached by clinical examina-

tion, cytologic analysis, US scan, mammography, and CT scan.

Depending on the stadiation, therapeutic options can include radiotherapy, surgery and endocrine therapy (5). Reports in the literature vary widely. Surgical treatment is used most frequently; someone use radiotherapy only, others prefer a conservative surgical approach reserving mastectomy only for large-dimension tumors. In any case, the parameters conditioning therapeutic options are the presence of underlying lesional nodule and its dimensions, enlarged lymph nodes, and hormone responsiveness. Radiotherapy alone is sometimes used for low dimension tumors (< T2) or performed with quadrantectomy when the lesion is nodule-associated. It can also be used with palliative intent in advanced disease.

Chemotherapy with radiotherapy and hormone therapy can be performed as adjuvant treatments (8).

Surgery can be conservative or radical; quadrantectomy, with radio- and/or chemotherapy, is indicated when a palpable nodule is present. In the absence of palpable nodule, a radical mastectomy with axillary dissection should be performed when the lymph nodes are enlarged (9) or, as in our case, when the erythema area is very large. Our surgical choice was conditioned by histological findings of the biopsy (pT4, G3), positive anamnesis for Paget-associated intraductal carcinoma, and axillary lymph node enlargement.

This case report is a simple contribution to an uncommon disease, even more rare when we consider the metachronous nature of the lesion.

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