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Case Report

Nipple-Sparing Mastectomy is Feasible Even for Male Breast Cancer? Case of a Young Klinefelter Syndrome Man Treated with Bilateral Nipple Sparing Mastectomy

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- Modified radical mastectomy
- Lymph node surgery
- Klinefelter syndrome

Abstract

Male breast cancer (MBC) is a rare pathology. The known risk factors include radiation exposure, estrogen administration, and diseases associated with hyperestrogenism, such as cirrhosis or Klinefelter syndrome. An increased risk of male breast cancer has been reported in families in which the BRCA2 mutation on chromosome 13q has been identified. In these report we describe a case of a 40 year-old Klinefelter Syndrome man who underwent Nipple sparing mastectomy, sentinel lymph node biopsy, axillary dissection and contra lateral prophylactic NSM for an invasive mixed ductal-lobular carcinoma of the breast.

ABBREVIATIONS

MBC: Male Breast Cancer; FBC: Female Breast Cancer; SM: Simple Mastectomy; MRM: Modified Radical Mastectomy; NSM: Nipple Sparing Mastectmomy; NAC: Nipple-Areolar Complex

INTRODUCTION

Male breast cancer (MBC) is a rare pathology [1], less than 1% of all breast carcinomas occur in men [2,3]. However, as female breast cancer (FBC), the incidence of MBC has increased over the past 25 years [4]. The mean age at diagnosis is between 60 and 70 years, though in all ages men can be affected.

The known risk factors [5] include radiation exposure, estrogen administration, and diseases associated with hyperestrogenism, such as cirrhosis or Klinefelter syndrome [6]. Definite familial tendencies are evident with an increased incidence seen in men who have a number of female relatives with breast cancer. An increased risk of male breast cancer has been reported in families in which the *BRCA2* mutation on chromosome 13q has been identified [7,8].

Today, most men undergo simple mastectomy (SM) or modified radical mastectomy (MRM) [9]. Nipple sparing mastectomy in men is describe only for benign disease like gynecomastia.

CASE PRESENTATION

A 40 year-old Klinefelter Syndrome man, visited the outpatient clinic of the Breast Unit at the Humanitas Cancer Center. During clinical examination in the upper outer quadrant on the left breast there resulted a 2.5 cm mass. No pathological palpable lymph nodes in the bilateral axillae. He had mammogram, which showed 15 mm suspicious left irregular lesion in the middle of the outer quadrants, far away from the nipple. He had ultrasound scan which showed between the outer quadrants of the left

breast, at a distance of about three centimeters to the nipple, a focal hypoechoic lesion with irregular shape and with faded edges to a major axis of 1.5 cm and pathological significance. The core needle biopsy (tru-cut) showed infiltrating carcinoma. The patient underwent a left nipple-sparing mastectomy (NSM), sentinel lymph node biopsy, axillary dissection and contra lateral prophylactic NSM. According to our technique, during surgery, frozen section examination of the retroareolar tissue was performed to ensure that the nipple-areola complex was free of disease. Also sentinel node biopsy was performed and intraoperative histopathological examination (OSNA), showed presence of macro metastases. Complete axillary lymph node clearance was performed.

Final left specimen report showed an invasive mixed ductal-lobular carcinoma of the breast, Histological grade (Nottingham Score): G3. DCIS present and focal. Peritumoral vascular invasion: present and focal. Margin status: free of cancer. High hormone receptor positivity for both estrogen and progesterone (>90%). Nuclear proliferation (Ki67): 20-25% and HER-2-Neu negative. Two out of 14 dissected lymph nodes were positive for macrometastases, including the sentinel node. Histopathologic staging (TNM) was pT2 (2.5 m) N1a (2/14). The Right Breast specimen was negative for cancer.

Adjuvant treatment recommended, after the MDM meeting, was 4 cycles of adriamycin and cylophosphamide, followed by hormonal therapy with Tamoxifen for 5 years. He had also left locoregional radiotherapy.

DISCUSSION

Men who undergo mastectomies frequently complain unsatisfactory aesthetic results because of the excision of the nipple-areolar complex (NAC) and the alteration of the normal male chest contour [9,10]. Although this disease differs from female breast cancer, all treatments are based on its analogy. Current literature is suggesting low recurrence rates following NSM: Sacchini et al. [11], reported only 2 local recurrences in a series of 123 patients who had NSM. In the reported case the surgical approach chosen was nipple-sparing mastectomy with sentinel node biopsy and axillary dissection. No data are available in the literature concerning this type of conservative mastectomy in men: the choice was made considering the distance between the tumor and the NAC and once the surgical margins were found to be clear, taking into account the patient's wishes about the cosmetic result. The patient was satisfied with the cosmetic outcome and the surgical approach it's found to be feasible. At the moment we need more information about NSM in MBC to define a proper protocol and guide lines for surgical treatment.

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