An uncommon subtype of breast carcinoma in a man: The pure mucinous carcinoma

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ABSTRACT

Aim: Our purpose is to describe a new case of pure mucinous carcinoma in a man, to put emphasis on the rarity of this histologic subtype and the problems of management due to the fewer reported cases in the literature and to compare it with the female counterpart. Observation: We report the case of a 79-year-old man presenting as a nodular mass of the right breast. The clinical symptoms weren't specific and the diagnosis was based on histological and immunohistochemical findings. Conclusion: Male breast carcinoma is rare compared to its female counterpart representing less than 2% of male breast carcinomas and 1% of all breast carcinomas. Pure mucinous carcinoma in male is an extremely rare histological subtype. It is a variety of carcinoma of the breast which is characterized by the production of an abundant extra-cellular and/or intra-cellular mucin. It has been regarded as one of the more benign forms of breast carcinomas. To our knowledge, about 30 cases of mucinous breast carcinoma in male have been reported in the English literature and only 10 cases of pure mucinous carcinoma have been described. In general, pure mucinous carcinomas have a favorable prognosis. Numerous studies have shown that breast carcinomas in male behave more aggressively compared to those in females. Further research is needed on male carcinomas as they are becoming more frequent.

1. Introduction

Male breast carcinoma represents less than 2% of male breast carcinomas [1]. For invasive carcinomas, the female and male tumours are morphologically indistinguishable [2]. Mucinous carcinoma of the male, also termed colloid or gelatinous carcinoma, is an extremely rare histological subtype. It is a variety of breast carcinoma characterized by the production of an abundant extra-cellular and/or intra-cellular mucin. It has been regarded as one of the more benign forms of breast cancer [3]. To our knowledge, about 30 cases of mucinous breast carcinoma of the male have been documented in the English literature with only 10 cases of pure mucinous carcinoma.

2. Case report

A 79-year-old man with no particular past medical history was explored for a rapidly growing painless mass of the right breast. He has no family history of breast carcinoma. Physical examination revealed a retroareolar and firm mass of the right breast. The preoperative clinical examination revealed no axillary lymphadenopathy. Ultrasound examination showed an inhomogeneous, hypoechogenic nodule with irregular margins (Figure 1). The extemporaneous examination of the sentinel lymph node was negative so that the axillary nodal dissection seemed unnecessary. A segmental mastectomy was performed and we received a specimen of 4.5 x 3.5 x 2 cm covered by a spindle-shaped skin of 4 x 1.8 cm. The mass was well circumscribed and measured 3.5 cm. It had a soft consistency with a gelatinous appearance. The microscopic examination showed a lobulated and gelatinous mass composed of large mucin pools around tumour nests (Figure 2a). At a higher magnification, small, solid and papillary islets of tubular tumor cells were floating in an abundant extra-cellular...
mucin. Tumor cells were uniform and round with minimal amounts of eosinophilic cytoplasm (Figure 2b). Atypia and mitotic figures were occasionally seen. Tumoral stroma was fibrotic. There was no extensive intraductal spreading. Immunohistochemical examination revealed a diffuse expression of the estrogen and progesterone receptors by the tumour cells and the absence of the expression of the HER 2/neu antigens (Figure 3). The use of cytokeratin antigens confirmed the absence of micrometastasis in the sentinel lymph node.

**Figure 1.** Ultra-sound examination showing a breast hypo-echoic mass with irregular margins.

![Figure 1](image1)

**Figure 2.** a/ Large mucin pools around tumour cell nests (HE x 200), b/ Small, solid and papillary islets of tubular tumor cells floating in an abundant extra-cellular mucin (HE x 400).

![Figure 2](image2)

**Figure 3.** Nuclear expression of estrogen and progesterone receptors (HE x 400).

![Figure 3](image3)

3. Discussion

Male breast cancer was first described by John of Aderne in the 14th century. It is a rare disease representing less than 2% of male breast carcinomas and 1.5 to 5% of all primary breast carcinomas [1, 4]. Its incidence varies widely between countries with an associated mortality rate that accounts for less than 0.2% of all cancer-related deaths in men [5]. Recent epidemiological studies suggest that the incidence of male breast cancer has been steadily increasing [6]. It usually occurs at advanced age, with a peak incidence around 60 years of age. It is extremely rare in boys and young adults. According to the literature, our patient was aged 79. Major genetic factors associated with an increased risk of men breast cancer have been identified including BRCA2 mutations, Klinefelter syndrome and a family history of breast carcinoma.

Epidemiologic risk factors for male breast cancer include disorders relating to hormonal imbalances such as obesity, testicular disorders and radiation exposure. Suspected epidemiologic risk factors include prostate cancer, gynecomastia, professional exposures, dietary factors and alcohol intake [7]. The symptoms and physical findings of pure mucinous carcinoma are non specific. The typical clinical appearance consists in a palpable lump. The lesion is remarkably well delimited and sometimes fluctuant on physical examination [8]. Studies have shown that male breast carcinoma is more likely to be of high grade at presentation despite of number of points that enable its early detection including the frequent submammilary location, the thinner soft-tissue cover and the frequent skin manifestation [9]. Mammography shows usually a round, mutlobulated and well circumscribed lesion. On breast ultrasonography, the tumor is isoechoic relative to the fat surrounding the breast tissue and it has often a well-defined margins. Some authors suggest that sonographic appearance of a homogeneous mass with cystic and solid components, vascularity and distal enhancement should raise strong suspicion of pure mucinous carcinoma but the differentiation of this lesion from benign one is usually challenging when based only on radiologic findings [8]. In fact, in our case, the ultra-sound examination revealed an ill-defined mass with speculated margins. On macroscopic presentation, the typical glistening gelatinous appearance with bosselated, pushing margins and a soft consistency make the lesion readily recognizable. The tumours range in size from 1 cm to 20 cm with an average of 2.8 cm [4].

The majority of the cases of male breast cancer are infiltrating duct carcinoma (70-95%). Mucinous or colloid carcinoma is a less common variant of breast cancer, and probably slightly more uncommon in men than in women [10]. Microscopically, it is subdivided into pure and mixed forms. The pure form is characterized by variable amounts of extracellular mucin surrounding the invasive tumor cells while mucinous tumors with invasive areas not surrounded by mucin are considered mixed. Significant differences have been noted between male and female breast cancers with respect to the expression of a variety of biologic factors, including hormone receptors, c-erbB-2, proteins related to basement membrane and extra-cellular matrix degradation [11, 12]. The majority of cancers arising in the male breast are estrogen and progeteron receptors positive and this finding does not correlate with a better prognosis. For instance, cases of male breast cancer are less likely to overexpress p53 and erb-2, which are correlated with a worse survival and an increased cell proliferative activity.
There is no clear consensus regarding optimal treatment. Three studies have suggested that mucinous carcinoma may be associated with a low risk of local recurrence after breast conservation [4, 13, 14]. Moreover, patients with pure mucinous carcinoma differ from that of mixed carcinoma for which the rate is similar to that of infiltrating ductal carcinoma not otherwise specified so defined that the treatment strategy including the type of surgery would be different [4]. Mucinous carcinoma of the breast has pathological features that are of minimal risk for local recurrence after breast-conserving therapy. Lymphatic vessel invasion and nodal involvement rarely occurred in pure mucinous carcinoma [4]. The incidence of axillary lymph-node metastases increases as a function of primary tumour size [4]. This finding supports the use of lumpectomy without axillary node dissection for patients with small pure mucinous carcinoma. Patients with pure mucinous carcinoma except those invading the local skin are suitable candidates for breast-conserving therapy. Most pure mucinous carcinoma can be treated with this therapy probably even in large tumours up to 5 cm in diameter. A mixed mucinous carcinoma should be treated in the same manner as an infiltrating duct carcinoma would be [4]. According to these findings, our patient underwent a breast-conserving therapy. In fact, the tumour measured 3 cm and there was no lymph node extension. Criteria for adjuvant systemic treatment are identical for men and women, although hormonal therapy (tamoxifen) has a more prominent place in the adjuvant setting because of the high percentage of positive hormone receptors in men. The prognosis of male breast carcinoma compared to the female one isn’t clearly established. In fact, it appears in some studies that male breast cancer has a more aggressive clinical behaviour than female breast cancer with a worse outcome when compared stage for stage. Some studies referred to worse prognosis in men mainly due to anatomic factors consisting in the paucity of breast tissue and close tumor proximity to the skin and the nipple enabling dermal spread and early regional and distant metastasis. Other authors suggest that men and women have similar prognosis when they have similar age and stage [16]. As with other types of breast cancers, the detection of metastasis in the axillary lymph nodes and the size of the tumor at the time of diagnosis are the major prognostic factors in patients with mucinous breast cancers. For pure mucinous carcinoma, it has been documented that size does not correlate with the incidence of metastatic involvement, although smaller tumors usually carry an excellent prognosis. Some authors have reported that prognosis is improved when the cellularity decreases and the amount of the extra-cellular mucin increases. The presence of the abundant extra-cellular mucin may act as a biological barrier [8].

4. Conclusion

In general, pure mucinous carcinoma has a favorable prognosis. Numerous studies have shown that carcinoma as in male behave more aggressively than those in females but further research is needed on male breast cancer as it becomes more frequently observed.

5. References