



## Multidisciplinary management of mammary Paget's disease: recommendations to optimize oncological and aesthetic outcomes.

We read with interest the article of Francesca Maria Plutino entitled "A peculiar case of Paget's disease of the breast" and would like to make some considerations about this particular topic <sup>1</sup>.

Mammary Paget's Disease (MPD) or Paget's disease of the breast is an uncommon pathology which accounts for less than 5% of breast cancers <sup>1-3</sup>.

MPD occurs with alterations of the nipple-areolar complex (NAC) such as redness, eczema, bleeding ulceration and usually itching <sup>1-3</sup>.

In the era of personalized care, a careful multidisciplinary management is mandatory to optimize the results and minimize the risk of overtreatment <sup>4</sup>; an adequate knowledge of the MPD, surgical skills and use of appropriate adjuvant therapies allow to reduce the risk of local recurrence and improve the aesthetic outcomes and patient's quality of life; however, a successful work can be more easily achieved thanks to the repetitive performance of some standardized tasks, such as <sup>5</sup>:

- an accurate radiological preoperative assessment with mammography and ultrasonography is important to identify associated glandular lesions; an underlying breast carcinoma (in situ and/or invasive) may be present up to about 80% to 90% of MPD although often without an evident breast mass or mammographic abnormality (2,5); therefore, all patients with MPD should also perform a magnetic resonance imaging to detect possible underlying occult breast carcinoma and define the true extent of disease <sup>5,6</sup>;

- a pathological diagnosis should be early established by nipple scrape cytology when a MPD is clinically suspected; full-thickness punch or wedge biopsy of the NAC may be necessary to accurately diagnose MPD; the histological examination must detect malignant intraepithelial carcinoma cells, also known as "Paget cells", in the epidermis of the NAC <sup>2,3</sup>; a needle biopsy is also required for any suspicious glandular lesion identified by imaging and associated with MPD <sup>5</sup>;

- a multidisciplinary "Surgery Board" is mandatory to select the more adequate local treatment for the patient: breast-conserving surgery (BCS) followed by radiothera-

py (RT) is the optimal local treatment when a NAC resection and wide local excision of any underlying cancer allows to achieve tumour-free margins and appropriate aesthetic outcomes <sup>4,5,7</sup>; the oncoplastic techniques with the remodelling of breast tissue and placement of clips within the excision cavity as a landmark to guide adjuvant RT should always be used in BCS in order to optimize oncological and cosmetic results <sup>5,8,9,10</sup>. Instead, skin-sparing mastectomy with immediate breast reconstruction is indicated for MPD associated to multicentric or extensive carcinoma, inadequate margins after BCS, contraindications to adjuvant RT and patient preference <sup>5,8</sup>. Staging and surgical treatment of the axilla in MPD is based on the possible presence of underlying cancer; sentinel lymph node biopsy (SLB) is not necessary when BCS is used to treat pure MPD or MPD associated with ductal carcinoma in situ; SLB must be performed when MPD is associated with underlying invasive cancer and treated with breast-conserving surgery; SLB is always recommended when a mastectomy is performed in order to avoid complete axillary lymph node dissection in case an invasive component is revealed at final pathology of the gland (mastectomy precludes subsequent use of SLB) <sup>5,7,8</sup>;

- a multidisciplinary "Tumor Board" is crucial to choose the adjuvant treatment; whole breast radiation should be always performed after BCS and a radiation boost should be considered for the site of the resected NAC and any associated resected cancer site <sup>2,4,5</sup>; adjuvant systemic therapies in patients with MPD should be based on biological features and the stage of the underlying cancer; no data are available to support the use of endocrine therapy in the MPD without an associated DCIS or invasive carcinoma <sup>5</sup>.

In conclusion, a dedicated multidisciplinary pathway with meticulous repetitive performance of some specific tasks could help to perform a successful work while optimizing oncological and aesthetic outcomes in patients with MPD.

## References

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