

EXTRAMAMMARY PAGET DISEASE OF THE VULVA: AN ANALYSIS OF 24 CASES

Obiettivo: Paget's disease can arise in the breast (Mammary Paget disease MPD) or in other locations (Extramammary Paget Disease EMPD) such as anogenital skin in both males and females (Extramammary Paget Disease of the Vulva EPDV). Underlying adenocarcinoma can be found in some cases. This study aims to report clinical aspects, surgical procedures, outcomes, recurrences of patients with EMPDV.

Metodi: A retrospective chart review was conducted on patients with pathologically confirmed diagnosis of EMPDV managed at the Department of Obstetrics and Gynecology, University of Bari, and "Giovanni Paolo II" National Cancer Institute in Bari, between 1998 and 2018.

Risultati: Records of 24 cases of EMPDV were examined. Median age of the patients at diagnosis was 68.9 (range 38-84), diagnosis of synchronous cancer was made in 2 cases and in 2 other cases of metachronous cancer. 3 patients had previously been diagnosed with other oncological diseases. All patients underwent surgery including wide local excision (6), simple vulvectomy (8) and radical vulvectomy (10). Lymphadenectomy was performed in 2 cases and reconstructions with advancement flaps in 7 cases. 4 patients were found to have invasive disease and one had inguinal node involvement. Positive margins were found in 11 patients. Wound dehiscence and urethral stenosis were found respectively in 4 and in one case. 8 recurrences (33,33%) were observed, regardless of positive surgical margins.

Conclusioni: EMPDV has a low rate of malignancy but a high rate of recurrence. It should be diagnosed early to avoid repeated surgery and to reduce symptoms and morbidity.