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Short Communication

Paget's Disease of the Vulva: A Review

Powell JL*

Department of Obstetrics & Gynecology, University of North Carolina School of Medicine ,Wrightsville Beach, N. C. 28480, USA

***Corresponding author:** John L Powell, Department of Obstetrics & Gynecology, University of North Carolina School of Medicine, Wrightsville Beach, N. C. 28480, USA

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Paget's disease is classified, according to location, as mammary or extramammary disease. The original lesion was described by James Paget (1814-1889) in 1874, involved the nipple and areola of the breast in 13 women, who developed underlying ductal adenocarcinoma within three years of presentation [1]. Extramammary Paget's disease most commonly involves the vulva and anus and was first described in 1901, by William Auguste Dubreuilh (1857-1935), professor of dermatology at the University of Bordeaux [2]. Paget's disease appears as a patchy, reddish and whitish, velvety, and eczematous skin lesion. Women with Paget's disease of the vulva are usually white and postmenopausal and complain of localized itching and burning. Paget's disease of the vulva is of apocrine origin and is confined to the epithelium in most cases. However, invasive disease is present in 13 to 25% of cases, either, as a result of direct invasion through the basement membrane or, less commonly, because of the presence of an underlying apocrine gland adenocarcinoma. Histologically, intraepithelial Paget's disease appears as large, pale cells, often in nests at the tips of the rete ridges. The cells are often seen infiltrating upward in the epithelium, which is hyperkeratotic. The Paget's cells can be located within any of the skin adnexa. Vulvar Paget's disease occurs with other malignancies in about 35% of patients; the most common of these is breast carcinoma. Other commonly associated cancers are basal cell, rectal, and genitourinary carcinomas. When Paget's disease involves the anus, there is a very high incidence of coexisting rectal cancer. Part of the preoperative work-up should be directed at screening for these cancers. When a patient is diagnosed with Paget's disease of the vulva, the area of involvement should be carefully inspected and palpated to detect areas suspicious for invasive cancer. If the disease clinically appears to be intraepithelial, then a wide local excision is performed, including a small amount of subcutaneous tissue. A well-known characteristic of intraepithelial Paget's disease is histologic extension far beyond that which is clinically apparent. Some authors advocate resection up to 1 to 2 cm beyond the clinically visible lesion. However, they report recurrence rates of 29.6% to 42% [3,4]. Theyalso report up to a 52% incidence of positive margins on permanent sections [4]. Some authors advocate the use of intra-operative frozen sections to define the extent of surgery necessary in order to reduce the incidence of positive margins and recurrences to as low as 11%. [5,6]. Frozen sections will not guarantee a 0% recurrence rate, but can significantly reduce them. Intraoperative mapping involves excising strips of tissue along the initially planned margins and sending them for frozen section. While waiting for the frozen section reports, the central lesion is excised. Other methods that have been reported to be useful in ensuring clear margins include Mons micrographic surgery [7] and fluorescein dye with ultraviolet light [8]. Colposcopy and toluidine blue staining are not helpful [9]. An experimental technique reported to be potentially useful in evaluating histologically negative margins is the application of a panel of monoclonal antibodies that may detect occult Paget cells [10]. When intraepithelial Paget's disease extends far beyond that which is clinically apparent, very extensive excision is necessary to obtain clear margins [6]. Primary closure of vulvar wounds is desirable, but may not be possible in such cases. Possible means of dealing with such cases include skin grafting, laser vaporization, and pedicle flap reconstruction [6,11,12]. Paget's disease can recur in a skin graft. Topical 5-fluorouracil or bleomycin, administered either pre- or postoperatively to treat recurrent disease [13]. Topical imiquimod 5% cream has also been used [14]. Management of recurrent intraepithelial Paget's disease of the vulva is similar to that of the primary lesion, with excision of at least the clinically evident disease. When vulvar Paget's disease is found to contain an invasive component, then the treatment is radical surgery [6]. Radiotherapy has been used as an alternative therapeutic approach for patients with extensive inoperable disease or medical contraindications for the surgical approach [15-18]. Salvage radiotherapy can be given for regional lymph node metastases, inoperable loco-regional recurrence, and in those who refuse additional surgery. Extramammary Paget's disease affecting the scrotum and penis was first described by Henry Radcliffe Crocker (1845-1909) in 1888 [19]. Crocker was a dermatologist at the University College Hospital and Middlesex School of Medicine in London. The mainstay of treatment is wide surgical excision [20,21]. In a series of 36 patients, 13 (36.1%) underwent local wide excision with frozen section evaluation of the margins and only 1 patient (7.7%) had a positive surgical margin on permanent sections [21]. However, [23] (63.9%) underwent local wide excision with margins of 1 to 2 cm only by gross examination, but 17 (73.9%) of them had positive margins on permanent sections. Topical 5-fluorouracil has been used for intra-epithelial disease [22] and combined chemo radiotherapy has been successfully used for regionally advanced unresectable disease [23].

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