CASE REPORT

Extra mammary Paget's disease of the vulva

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ABSTRACT

Extra mammary Paget's disease (EMPD) is a rare condition, which affects postmenopausal women. Wide local excision may not be appropriate in elderly patients with extensive disease. It is an uncommon malignant neoplasia with a high local recurrence rate. The standard treatment is local excision of the affected area with adequate margins; however, 40~45% of cases recur after surgery within 4 years. Although surgery is currently considered the preferred primary treatment for EMPD, it has a high relapse rate due to the multifocal nature of the disease. Hence, RT in selected cases of EMPD of vulva may be beneficial.

Key Words: Extra mammary Paget's disease of vulva, extra mammary Paget's disease, radiotherapy

INTRODUCTION

James Paget first described mammary Paget's disease in 1874, and EMPD was first described by Crocker in 1888.[1] Extra mammary Paget's disease (EMPD) of the vulva mainly affects aged postmenopausal women.It is an uncommon malignant neoplasia with a high local recurrence rate. The standard treatment is local excision of the affected area with adequate margins; however, 40~45% of cases recur after surgery within 4 years.[2] Mohs micrographic surgery has been tried in many advanced centers to tackle with the problem of adequate margin resection. Cryosurgery, topical 5-fluorouracil (5-FU) alone and systemic chemotherapy (mitomycin C and 5-FU) have also been employed as primary treatments.[3,4] Imiquimod cream applied locally helps in the prevention of recurrence. EMPD usually affects the elderly who may be medically unfit for surgery, and the treatment of genital lesions may involve mutilating surgery. However, more recently radiation therapy (RT) has been considered as an appropriate primary or adjuvant treatment with curative intent, in those who are medically unfit for surgery or in whom other modalities are inappropriate.

CASE REPORT

A patient 65 years came to our OPD on 20th April 2010, with h/o local excision of a vulval lesion on the right labial fold, which was 1×1 cm in size, she had severe itching,erythematous plaque with ulceration for the last 4 years, she consulted a gynecologist following which excision biopsy was done on 10th April 2010. Her biopsy report was Paget's disease of Vulva with margin positive. She was evaluated in our OPD and on examination her general condition was good, she was hypertensive controlled with medication, mild pallor, bilateral breast was normal. Per-abdominally no abnormalities detected, bilateral groin normal, no palpable nodes. On local examination of the vulva there was a fibrous scar with nodularity [Figure 1], there was pruritus and tenderness present. Per rectal examination was normal. Genitourinary and gastrointestinal neoplasms were excluded by abdominopelvic CT scan and endoscopy.It was decided to perform right sided radical vulvectomy with widesurgical margin [Figures 2 and 3] along with bilateral groin node dissection which was done under general anesthesia on 25th April 2010. Her post-operative histo-pathological examination showed epidermis with keratinization and pagetoid infiltration of epidermis by the malignant cells with clear cytoplasm confirming it to be a case of extramammary Paget's disease of vulva [Figures 4 and 5], margin again turned out to be positive, inspite of taking a wide margin. Histo-pathological

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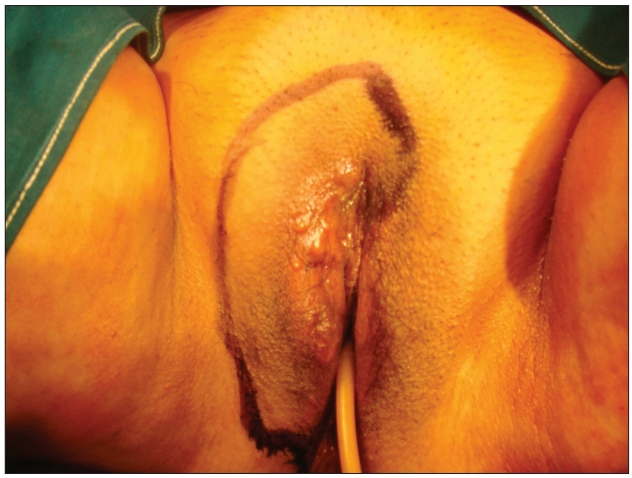

Figure 1: Pre-operative photograph of the lesion

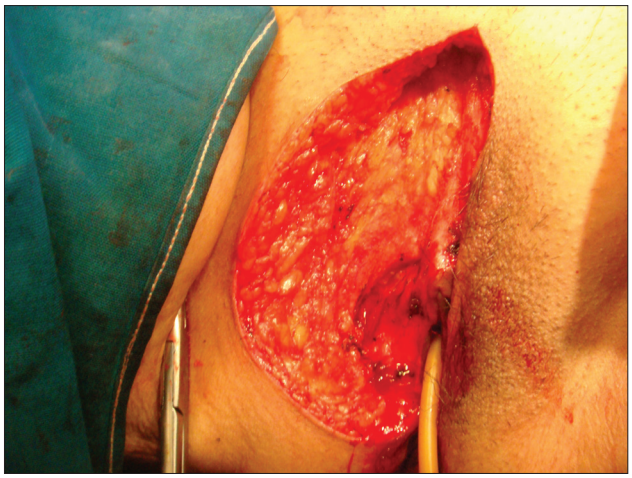

Figure 2: Intra operative photograph post resection showing wide resection margin

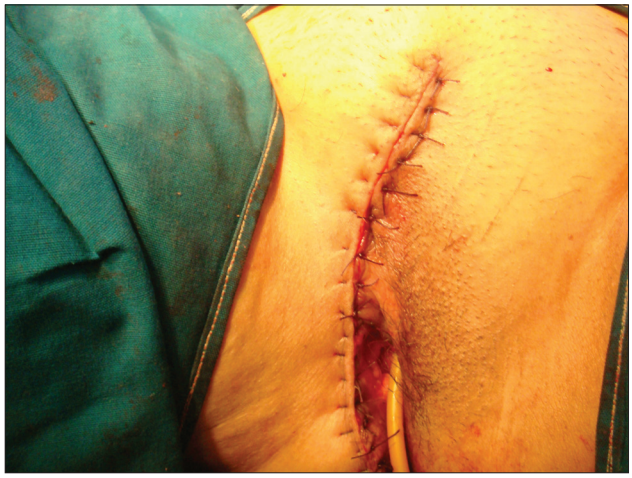

Figure 3: Showing post-operative closure of the wound

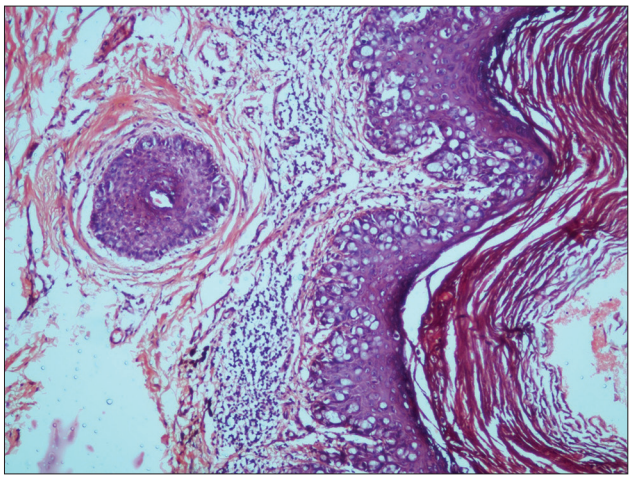

Figure 4: Showing epidermis with keratinization and pagetoid infiltration of epidermis by the malignant cells with clear cytoplasm

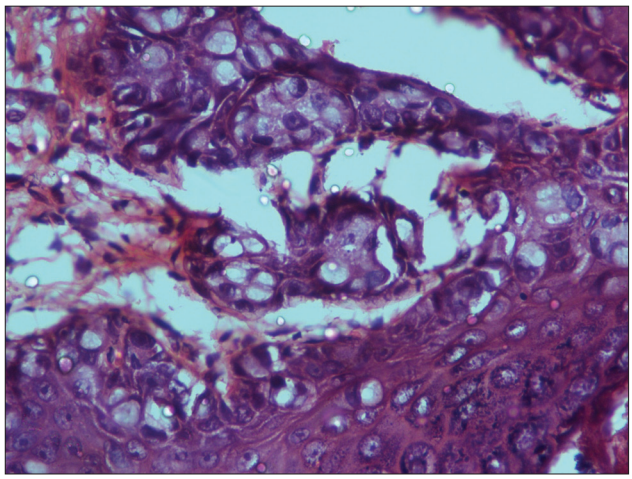

Figure 5: High power view of Paget's cells in the epidermis

differential diagnosis includes Bowen's disease and superficial spreading melanoma, IHC studies showed positivity for CK7 and negative for CK20, a typical immune-phenotype of vulvar Paget's disease. HMB 45 was negative in our case further ruling out melanoma.So it was decided to take her up for adjuvant radiotherapy. She received 40 Gy/20 # from 26.05.10 to 22.06.10 (6MV) F.S 20 × 18 cms, 9.5 cm depth by AP/PA with shielding, + 10 Gy/5# from 23.06.10 to 29.06.10 (12 MeV) F.S10X10 cms, at 2.5 cms, with Frog Leg position, wet cotton bolus over right inguinal scar region. Following RT, she showed an almost normal skin and mucosal appearance in the genital area, although she had experienced temporary radiation-induced skin reactions toward the end of the RT, which completely resolved within six weeks after RT. She is undergoing regular followup for the last twenty months, she last attended our OPD on 20th February, 2012, and she is doing fine with no evidence of disease.

DISCUSSION

EMPD is a rare disorder which occurs more commonly in women than in men, and more frequently between

the ages of 50 and 80 years. Because of its rarity, no randomized clinical trials have been performed, and thus it is difficult to compare the efficacies of surgical resection and RT. So to determine the efficacy of RT long-term follow-up and randomized study is mandatory. However, primary or postoperative adjuvant RT in selected patients with EMPD should be viewed as highly effective treatment modalities. Though, optimal radiation doses have not been definitively determined, many authors recommend 40~50 Gy in intraepithelial Paget's disease and 55~65 Gy in invasive Paget's disease or EMPD with associated adenocarcinoma. Although surgery is currently considered the preferred primary treatment for EMPD, it has a high relapse rate due to the multifocal nature of the disease. Hence, RT in selected cases of EMPD of vulva may be beneficial.

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