

CASE REPORT

Paget's Disease and Apocrine Carcinoma of Vulva

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ABSTRACT

A case of Paget's disease and apocrine carcinoma of vulva in an 82 years old woman was diagnosed and treated surgically. Three years after the operation the patient was alive and well. The role of recognition and long term follow up are emphasized.

Extramammary Paget's disease is a rare intraepithelial neoplasm. Common sites of occurrence include the vulva, perianal region and perineum. Vulvar Paget's disease usually occurs in women in her 7th decade of life or older. The lesion is usually noninvasive which can be cured by local wide excision. The recurrence is common and a regular long term follow up is mandatory.

A case report

A 82 years old woman, Para3, last 50 years, was seen at the outpatient department of Rayong Hospital because of an ulcer at her vulva. Her past history was essentially normal. She was always in good health. She had never seen any physician or visited any hospital. Three months before this visit, a mass was found on her right labium majus, neither pain nor tenderness was reported by the patient. She had experienced pruritus now and then for years. Ulceration and inflammation around the mass brought her to the hospital.

Physical examination revealed a healthy old woman, not anemic, not icteric. Her heart and lung were normal on auscultation. Abdomen was soft, no mass could be felt. Breast examination showed no abnormal mass, ulcer or discharge. Her blood

pressure was 160/90 mmHg, pulse rate was 80/min, respiratory rate was 21/min. An inflammatory papule 1.5 cm extending from the mass. The rest of the labia was atrophic as the result of aging. Vagina, cervix, uterus and adnexa were all atrophic. Rectal examination showed neither mass nor abnormal rectal shelf. No lymph node enlargement was found in both inguinal and femoral areas. Her PAP test was class 1. Biopsy was taken from the mass. Pathological result was apocrine carcinoma and Paget's disease.

Diagnosis of Paget's disease and apocrine carcinoma was made. The patient was advised to have the lesion removed. The patient refused to have any operation. She stated that she was too old to have any operation. Two weeks later, she returned and agreed to have surgical treatment for the disease.

Preoperative evaluation of the patient showed a normal complete blood count and normal urinalysis. Her EKG and chest film were within normal limit. Her fasting blood sugar was 90 mg/dl. BUN was 14 mg/dl. Creatinine was 0.9 mg/dl. VDRL was non-reactive.

Under general anesthesia, the patient was in lithotomy position, prepped and draped in usual manner. Wide excision was made around the lesion. The margin of excision was 1 cm clear of the

inflammatory area. Total skin thickness and underlying fat were removed. Ipsilateral groin nodes were explored and removed. The border of surgical wound, as expected, could not be approximated without undue tension. Bleeding was checked and the wound was partially closed. The area that could not be easily approximated are left open. Wet dressing was done on daily basis. Claforan 1 gm three times a day was given before the incision was made and continued postoperatively for 1 week. It took 18 days for the wound to heal by secondary intention. The stitches was removed and she was discharged on 24th postoperative day.

The excised tissue was submitted for pathological examination. Pathological report confirmed the diagnosis of Paget's disease and apocrine carcinoma of vulva without nodal involvement. The surgical margin was free from Paget's cell.



Fig. 1. Arrows indicate typical Paget's cell.

One week, four weeks, three months postoperative follow up showed no evidence of recurrence. She was scheduled for follow up every 6 months. Three years after the operation the patient was still healthy and showed no evidence of recurrence.

Discussion

Twenty seven years after Sir James Paget

described Paget's disease of breast, extramammary Paget's disease was described. Vulvar Paget's is one of extramammary Paget's disease. Despite the fact that vulvar Paget's disease contribute only 1-5 % of vulva malignancy,^(1,2) the significant of recognition and proper management of the lesion is of growing importance, due to increase in life span of women in this era. Paget's disease of vulva is probably has different origin and behaves differently from Paget's disease of breast⁽¹⁾ which is believed to be cutaneous manifestation of intraductal carcinoma. It is widely accepted that Paget's disease of vulva originated from apocrine cell.^(3,4,5) The evidence was substantiated by using monoclonal antibody specific to cytokeratin.^(6,7) This evidence also described the fact that Paget's disease are occasionally multifocal.

The disease is usually affected woman older than 60 years old.^(1,2) In this report, she was 82 years old. In some cases adenocarcinoma of breast, colon, cervix and skin were reported to occur concurrently.⁽⁸⁾ But this patient had normal pelvic, rectal and breast examination. The treatment was wide excision and ipsilateral removal of groin nodes. As with squamous cell carcinoma of vulva, which usually affect the aged woman, healing by primary intention is very difficult. In order to prevent necrosis of surgical edge, healing by secondary intention was attempted in all of these kinds of vulvar operation in our hospital with satisfactory result. In some cases, skin graft has been used to cover the wound with good result. Recurrence at the graft were reported on some occasions.^(9,10) The recurrence of Paget's disease had been reported to occur as long as 10 years after treatment. Long term follow up is mandatory.^(6,7,11) The occurrence of apocrine carcinoma is not unexpected. The presence of the tumor need more aggressive treatment than Paget's disease per se, ie, the ipsilateral groin nodes had to be removed. If the patient's ipsilateral nodes were involved, the contralateral nodes should have been removed. The role of frozen section in management of Paget's disease is controversial.^(1,2,12) In our situation thorough clinical evaluation is probably the best way to handle it. Not to mention laser

treatment which is also controversial at best.⁽¹⁾ Her prognosis would be good as she had no risk factors, ie, invasive Paget's disease, underlying adenocarcinoma, intraepithelial Paget's disease with coexisting cancer or clitoral and urethral location.^(8,9)

In conclusion, an 82 years old woman with Paget's disease and apocrine carcinoma was diagnosed clinically and management was given accordingly. The relation between apocrine gland and Paget's disease of vulva was reviewed. The diagnosis and optimum treatment for our situation was suggested. Three years after the surgical treatment, the patient showed no evidence of recurrence .

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