

Paget's Disease of the Breast

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Introduction

In 1874 Sir JAMES PAGET described a series of 15 women, in each of whom chronic eczema or a weeping eruption of the skin of the nipple or areola was followed by the development of breast cancer. He was supported in this view by BUTLIN who made the first microscopic studies and CHEATLE, INGULIS postulated that the lesion begins in an intraductal carcinoma and spreads within the epithelium of the nipple. Most present-day pathologists support the view that the lesion of the nipple is an intraepithelial extension of an underlying intraductal carcinoma.

Case Material

During the period 1966 through 1973, three cases of histologically proven PAGET's disease of the breast were seen by General Surgical Division at Tenri Hospital. At this period, there were 182 cases of surgically treated breast cancers. An incidence of PAGET's disease was 1.6%. This compares to the 0.7% reported by DOCHERTY and the 3.2% reported by R. ASHIKARI. Preoperatively, all of the three patients were suspected as Paget's disease.

Case I. The patients (A.J.), a seventy-two year old female, had noted erosion and crusting of the right nipple for one year and a half prior to admission for retraction of the nipple. (Fig. 1) On physical examination no lump was noted in the breast. Biopsy showed PAGET cells and a radical mastectomy was carried out with axillary lymph nodes being negative histologically. Figure 2 demonstrates PAGET's disease of the skin of the areola in the microscopic sections. In the lower layer of the epidermis, large and hyperchromic nuclear PAGET cells



Fig. 1 Erosion and crusting of the right nipple (Case I)

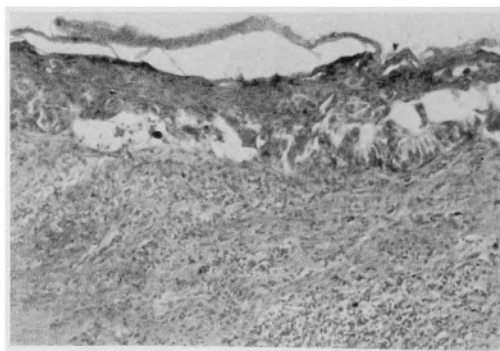
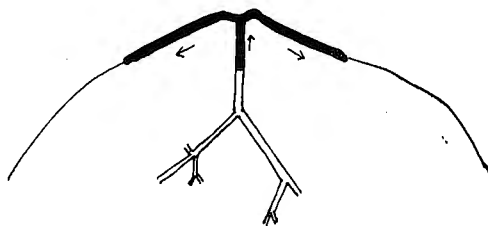


Fig. 2 Paget cells are seen in the lower layer of the epidermis. (H-E $\times 100$)

72 Y. F.



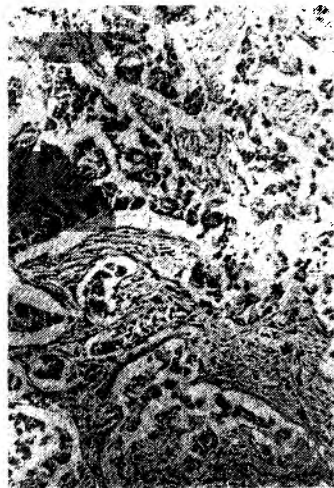
Noninfiltr. D. Cancer

Ax. Meta. (-)

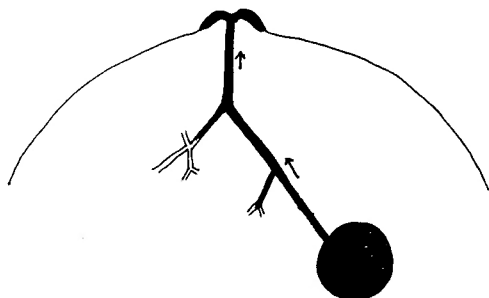
Fig. 3 Intraepidermal extension of PAGET cells (Case I)

are seen in clumps, but do not invade the cornium. On histological examination with serial microscopic sections, we found non-infiltrative duct cancer limited to the ostia of the major ducts. And then it is considered that the extension of the duct cancer is as described the schema (Fig.3). It's a typical extension of PAGET's disease of the breast without mass.

Case II. The patient (S.H.), a fifty-five year postmenopausal female, was admitted to the hospital because of eczema of the left nipple with a lump which measured 2 by 3 cm. (Fig. 4). After positive biopsy a radical mastectomy was performed and the nodes examined were free of metastatic disease. Figure 5 shows infiltrative duct cancer at the major ducts and Figure 6 demonstrates underlying massive cancer (medullary tubular carcinoma). And therefore this is thought to be infiltrating carcinoma of common type with intraepidermal extension, so called Pagetoid carcinoma. (Fig. 7)

**Fig. 4** Erosion of the left nipple (Case II)**Fig. 5** Infiltrative duct cancer at the major duct (H-E×40)**Fig. 6** underlying massive cancer (medullary tubular carcinoma) (H-E×100)

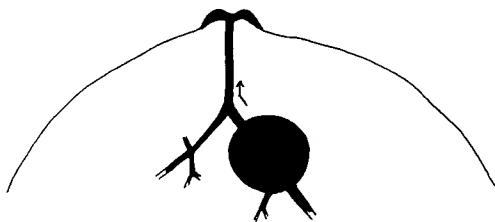
55 Y. F.



Infilt. D. Cancer
Ax. Meta. (-)

Fig. 7 Intraepidermal extension of the underlying infiltrative duct cancer (Case II)

52 Y. F.



Infilt. D. Cancer
Ax. Meta. (+)

Fig. 8 Spreading type of cancer cells (Case III)

Case III. The patient (Y.I.), a fifty-two postmenopausal female, had exzema of the left nipple and underlying lump. A radical mastectomy was underwent and axillary lymph nodes were positive for tumour. On histological examination, the spreading type of cancer cells is also cleared as like Case II. (Fig. 8)

Comments

The nature of PAGET's disease of the breast has been the subject over the years of considerable debate in the literature. PAGET himself gave no histologic description of the entity in his original paper. There is not yet complete agreement on the pathogenesis, the origin of the PAGET cells and their relationship to the underlying breast carcinoma. Although the recent ultrastructural observation suggests an in-situ transformation epidermal cells to PAGET cells, the most commonly accepted view is that the nipple and skin changes are due to extension to the epidermis of cancer cells migrating from an underlying intraductal cancer by way of the lactiferous ducts.

After HAAGENSEN's classification patients with PAGET's disease can be divided into two groups: those with nipple changes with or without palpable masses. It has been show by ASHIKALI and others that PAGET's disease in the breast without a palpable mass carries a far better prognosis than that in breasts with a palpable mass. But MAIER illustrated that survival in the group with no mass versus the group with a palpable mass...without lymph node involvement...are not significantly different and that involvement of axillary nodes is the single, most important, prognostic factor. Clinically the most significant aspectes of PAGET's disease of the breast are related to the presence or absence of a mass and the presence of lymph node metastasis.

On the surgical treatment, it had been shown by many authors that radical mastectomy is not significantly better than other modes of therapy. It is concluded that simple mastecto-

my and irradiation were as effective as radical mastectomy in patients who had no breast mass, and patients with an associated underlying mass had a lesion which behaved much like ordinary carcinoma of the breast, therefore extended radical or radical mastectomy to be appropriate therapy.

Summary

A review of three patients with PAGET's disease of the breast seen at Tenri Hospital from 1966 to 1973 has been made. All three cases are postmenopausal, two associated with underlying masses and the other without. On careful serial histological examinations, it is considered in all three cases that nipple and skin changes are due to extension to the epidermis of underlying intraductal cancer by way of the lactiferous ducts.

Radical mastectomies with irradiation were carried out of to them.

The abstract of this article was reported in part before the 113th Kinki Local Meeting of Surgery.

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和文抄録

乳房 ペー ジ ャ ッ ト 氏 病

天理病院腹部一般外科

谷川 允彦, 柏原 貞夫, 倉本 信二
前谷 俊三, 殿塚 健司, 佐藤 守

1966年より1973年まで三例の乳房ページェット氏病を経験した。これは乳癌183例中の1.6%にあたり、全例閉経女性である。第1例は72才で乳輪を越える湿疹様皮膚病変及び乳頭陥凹があり、乳腫腫瘤は触れず、腋窩淋巴腺転移は認めなかった。標本を連続切片にて検索することにより、乳管洞付近の輸出管に於ける非浸潤型腺管癌の扁平上皮内進展と考えられた。

第Ⅱ、Ⅲ例は共に、近傍に腫瘤を伴うページェット氏病で、年齢は52、55才であり、52才の例には、腋窩淋巴腺転移を認め、55才の例は、これを認めなかつ

た。共に通常型管外浸潤型乳癌の経導管的表皮内蔓延と考えられた。

以上のように、三例はすべて輸出管上皮に由来する乳癌の乳頭表皮内蔓延であると考えられた。

乳房ページェット氏病は、その臨床像、組織像、罹患年齢、予後等により、腫瘤の有無をもって二型に区別される。

治療については、我々は、腫瘤の有無を問わず、標準型根治的乳房切断術兼術後照射療法を施行した。