

Case Presentation

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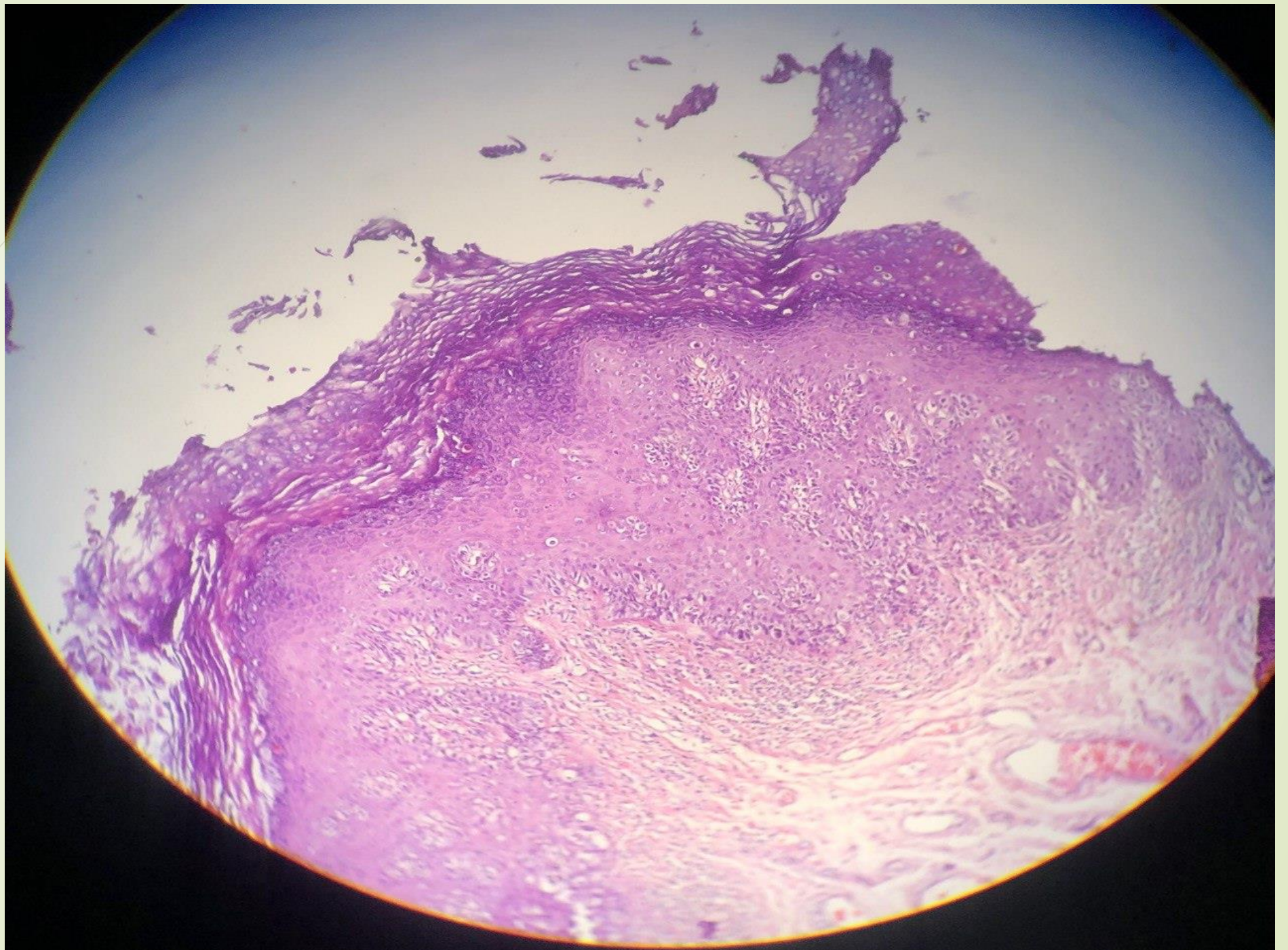


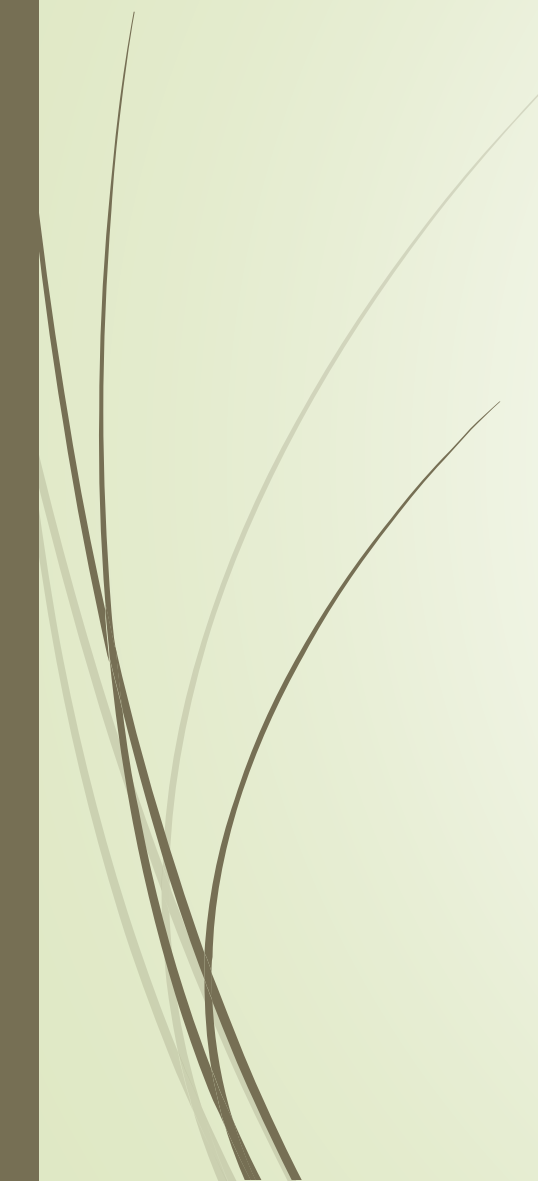
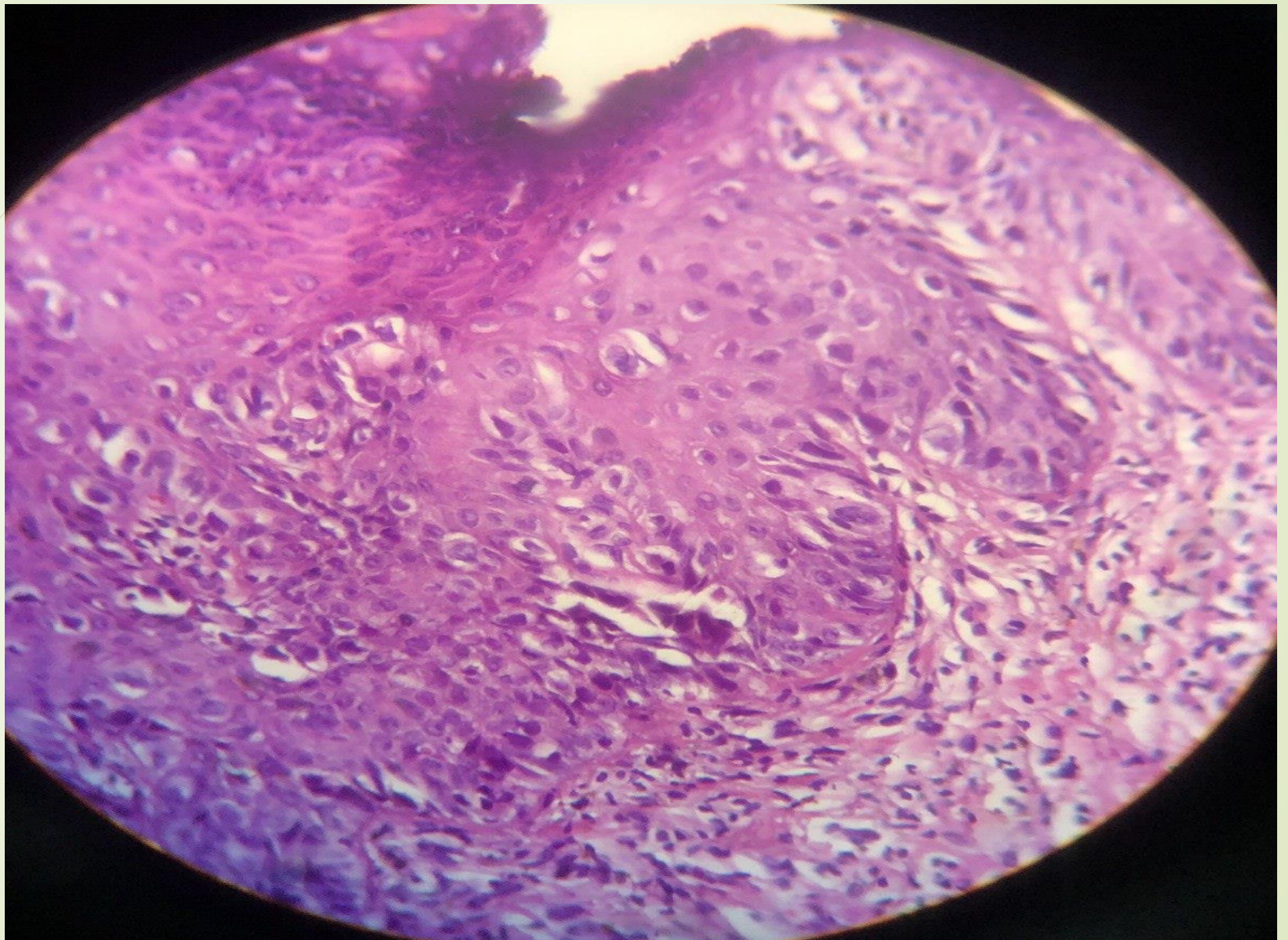
Macroscopy:

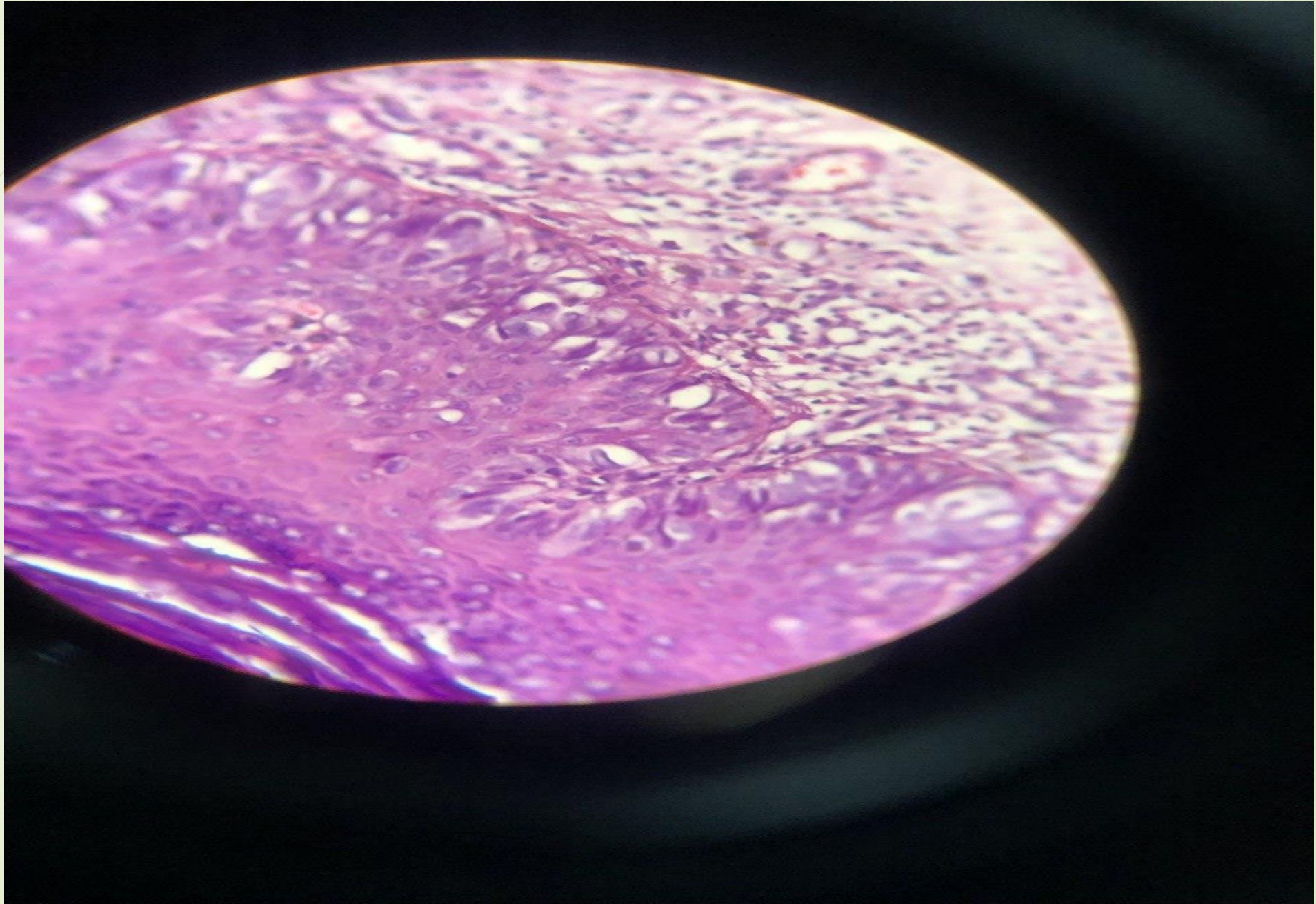
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- Specimen labeled as **Vulvar lesion**: Consists of six soft cream tissue fragments measuring 1.5 * 0.7 * 0.5 cm.

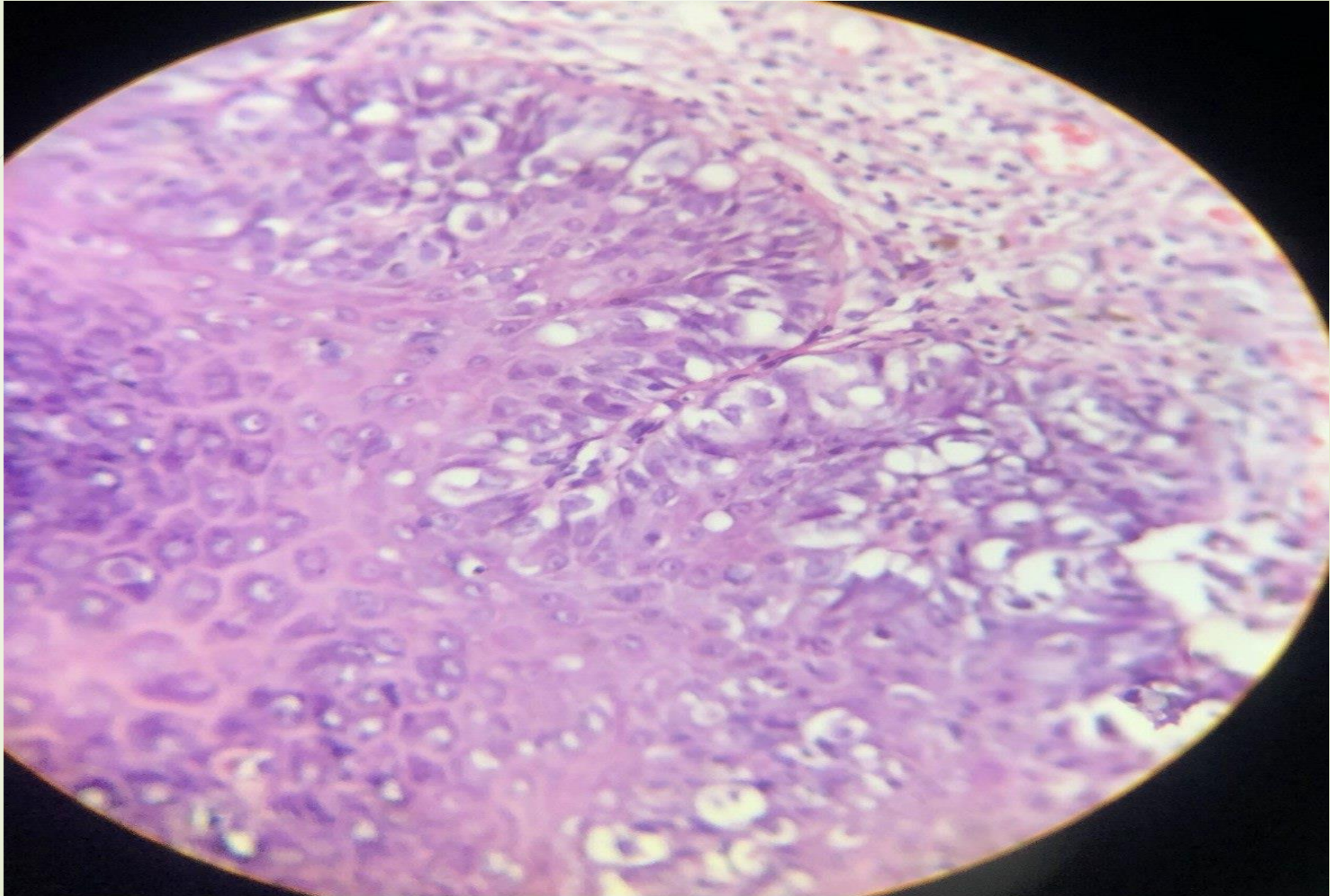


Microscopy:













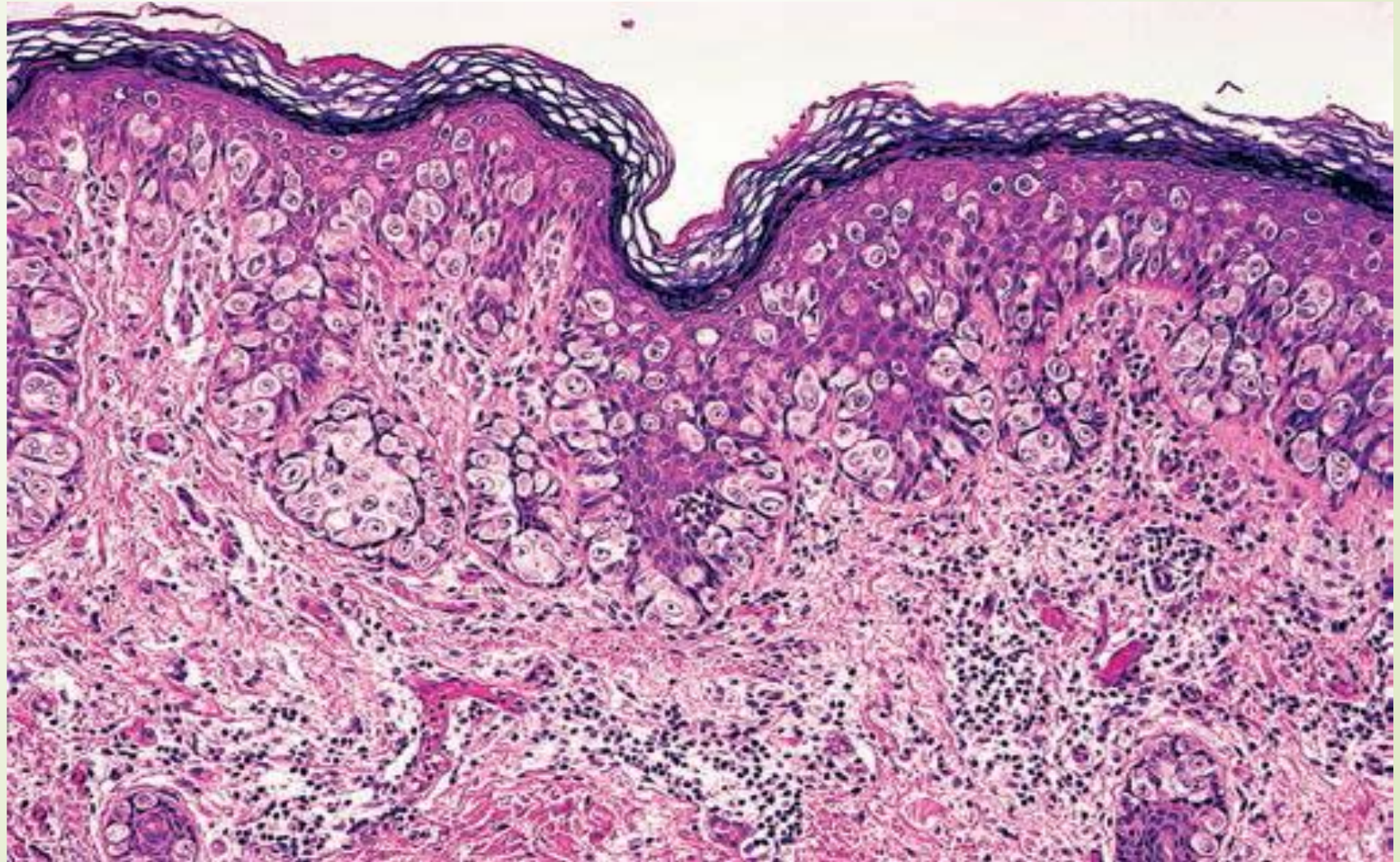
What's your diagnosis?

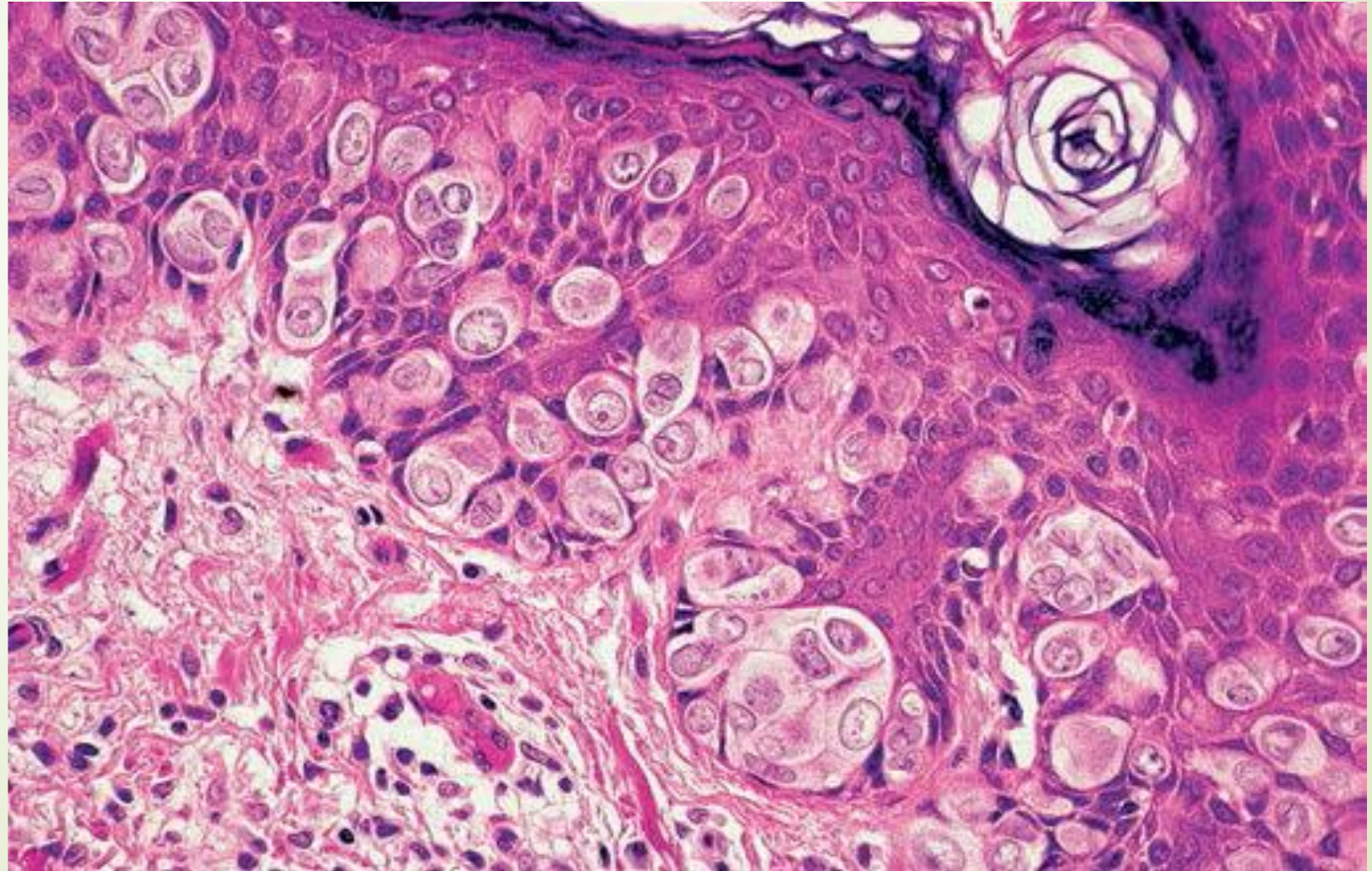



Paget disease

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- ▶ Paget disease is a malignant tumor of the vulva that can be subclassified as primary Paget disease, or secondary Paget disease, when it represents spread from an underlying carcinoma of rectum, bladder or endocervix.
 - ▶ Paget disease is a vulvar adenocarcinoma of uncertain histogenesis. It could be viewed as:
 - ✓ a sweat gland carcinoma arising primarily from the intraepidermal portion of the glands (acrosyringium)
 - ✓ a carcinoma of multipotential (adnexal stem cells) residing in the infundibulo-sebaceous unit of the hair follicles and other adnexal structures that differentiate along glandular (sweat gland) lines
 - ✓ as being derived from a population of CK7-positive clear cells associated with the openings of the ducts of the mammary-like glands of the vulva, known as Toker cells

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- Clinically, Paget disease presents as a crusting, elevated scaling erythematous rash in the labia majora , labia minora , and/or perineal skin of adult and elderly patients
 - Microscopically, the epidermis contains large pale tumor cells that form solid nests , glandular spaces, or a continuous layer along the epidermal basement membrane and also in pilosebaceous structures and sweat ducts
 - A cleft often develops between the row of malignant cells and the overlying keratinocytes, resulting in a low-power appearance sometimes reminiscent of an acantholytic suprabasal bulla
 - Paget disease can also be misinterpreted as **malignant melanoma**. It should be noted that the presence of melanin granules in some tumor cells does not rule out the diagnosis of Paget disease





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- ▶ **Histochemically**, some or all of the tumor cells contain acidic mucin, as evidenced by their positivity for Mayer mucicarmine and aldehyde fuchsin stains.
 - ▶ **Immunohistochemically**, these mucins are positive for **MUC1** and **MUC5AC**, the latter in striking contrast with Paget disease of the breast. They are also reactive for **HGM-45**, a marker associated with gastric surface mucous cells

▶ Other positive marker:

✓ **Pankeratin**

✓ **Cytokeratin 7 (CK7)**

✓ **Epithelial membrane antigen (EMA)**

✓ **Carcinoembryonic antigen (CEA)**

✓ **B72.3**

✓ **GCDFP-15 (a marker of apocrine differentiation)**

✓ **S-100 protein** is positive in up to one-third of the cases but **HMB-45** and **Melan-A** are negative

✓ **CDX2** negative (in contrast to a secondary Paget disease associated with rectal carcinoma)

Variably expresses **ER** and **PR** and frequently expresses androgen receptor



✓ **CK7 + / CK20 –**

✓ **GATA 3**



The difference between paget disease of the breast and vulva:

- ▶ **Paget disease of the breast** is nearly always associated with an underlying carcinoma, which may be intraductal or invasive and the intraepidermal malignant cells are more often than not mucin-negative.
- ▶ In contrast, the majority of the cases (about 90%) of vulvar Paget disease are not associated with an invasive carcinoma and are usually (although not always) positive for mucin stains

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- Unfortunately, the microscopic extent of the disease is often greater than that suspected from clinical examination and this should be taken into account at the time of surgery
 - The status of resection margins is only minimally related to the incidence of local recurrence
 - Cases of Paget disease with an invasive component beyond the microinvasive stage have a high likelihood of nodal involvement

تاریخ جواب ۱۳۰۰/۱۲/۰۱

مرکز آموزشی درمانی شهید بهشتی

صفحه ۱

گزارش آزمایشگاه پاتولوژی

درخواست ۱۳۰۰/۱۲/۰۱

تاریخ خدمت ۰۰/۱۲/۰۱

نام ملیحه احمدی نام پدر: ش کد ملی: ۱۱۴۱۱۴۷۴۷۵ سن بیمار: ۴۴ سال
 ش ورود ۲۰۴۴۵۴۹ پرونده: پذیرش: سرپایی صدور: سایر جنسیت: مونث پزشک: دکتر فهیمه ثابت
 وضعیت تاهل: ملیت: ایرانی شغل: بیمه: تامین اجتماعی سرپایی [۱۱۴۱۱۴۷۴۷۵]
 آدرس و شماره تلفن: ۰۹۱۴۹۱۷۱۴۵۲ - سلیمان - شیخ بهئی - عرجا ۲۱ بن بست جابجایی
 پزشک تایید کننده: دکتر آقر تعیمی
 لام شماره 2673

Macroscopic description :

با برجست ضایعه و لو : شامل ۶ قطعه بافت کرم خاکست : جمعاً به ابعاد ۱/۵*۰/۷*۰/۵ س م

Microscopic description :

در بررسی میکروسکوپی نمونه ضایعه و لو : مقاطعی از اپی درم با تغییرات کراتوز و هیپرکراتوز و باپیلوماتوز خفیف مشاهده می شود در ناحیه تحتانی اپی درم پرولیفراسیون سلولهای اپی تلیالی با نمای آشیانه های سلولی دارای سیتوبلاسم های روشن و هسته های با آتیبیسم و پلنومورفیسم مشاهده می شود نهاجم به استرومای زیرین دیده نمی شود

IHC study on block 2673-2 results as follow:
CK-7: positive
CEA: Positive
HMB-45: Negative

Diagnosis : Vulvar lesion biopsy :
- Paget disease
- Invasion to stroma is not seen
- Margin evaluation is not possible (due to fragmentation)

C:51.9 M:8542/3
پاتولوژیست :

