CASE REPORT

Mucinous metaplasia of the vulva in a case of lichen sclerosus. A case report

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Mucinous metaplasia of the genital area is a rare condition characterised by the emergence of mucin containing cells in stratified squamous epithelium. This report describes a unique case of benign mucinous metaplasia of the vulva associated with lichen sclerosus in a 60 year old woman. Histopathology revealed cervical type metaplasia with otherwise typical lichen sclerosus. This report discusses the pathogenesis and differential diagnosis of mucinous epithelium.

A 60 year old woman presented with a few months history of postcoital bleeding and pruritis vulvae. Physical examination revealed a white raw area 12 mm in diameter located on the left labium major and extending into the vaginal introitus. A biopsy was taken and histological examination showed lichen sclerosus. However, the most unusual aspect of this case was that the stratified squamous epithelium was replaced by a stratified epithelium, the superficial layers of which were represented by columnar cells with multivacuolated mucin containing cytoplasm. There was no nuclear atypia (fig 1). Mild acute inflammatory changes were also present, probably secondary to excoriation. Mucin laden cells in the epithelium stained violet with diastase periodic acid Schiff and Alcian blue at pH 2.5, indicating the presence of neutral and acid mucins (fig 2). These cells also stained positively for CA19.9, oestrogen receptor, and carcinoembryonic antigen. The cells also showed focal positivity for cytokeratin 7 and negative staining for cytokeratin 20, indicating endocervical type mucinous metaplasia.

DISCUSSION

Mucinous cells are not a normal component of the skin. The differential diagnosis of mucin containing cells in the epidermis of the vulva includes the following: cutaneous in situ and invasive squamous cell carcinoma with mucinous metaplasia, extramammary Paget’s disease,1 mucinous syringometaplasia,2 mucinous papulosis,3 and epidermotropic metastasis. These must, in their turn, be distinguished from superficial spreading melanoma.

Lichen sclerosus is a well recognised common inflammatory dermatosis of unknown aetiology, which affects both sexes, but is most commonly seen in the vulva in women of any age.4 There seems to be a higher risk that lichen sclerosus may be associated with vulval carcinoma.5 An association with mucinous metaplasia has not been reported to date.

Figure 1 Lichen sclerosus of the vulva with surface columnar epithelium (haematoxylin and eosin staining).

Figure 2 Mucin in the cytoplasm of the columnar cells (stained with Alcian blue/periodic acid Schiff with diastase).

Genital mucinous metaplasia is an uncommon disorder. A case of mucinous metaplasia of the vulva7 and vestibule and lower vagina,8 in addition to a few cases of mucinous metaplasia of the prepuce and glans penis,9 have been reported. However, the histogenesis of mucinous metaplasia of the genital area remains unknown. It probably represents a non-specific reactive process. The association with lichen sclerosus is probably fortuitous or may be a further indication of longterm external damage or chronic inflammation.10 The absence of nuclear atypia, preservation of nuclear polarity, confinement of mucin containing cells to the epidermis, the predominant location of the cells in the upper layers of the epithelium, and the fact that the mucinous cells replace squamous epithelium rather than infiltrating it distinguish this condition from extramammary Paget’s disease.
Take home messages

- We report a rare case of benign mucinous metaplasia of the vulva associated with lichen sclerosus in a 60 year old woman
- Histopathology revealed endocervical type metaplasia with otherwise typical lichen sclerosus
- The histogenesis of mucinous metaplasia of the genital area is unknown, but it probably represents a non-specific reactive process
- The association with lichen sclerosus is probably fortuitous or may be a further indication of long-term external damage or chronic inflammation

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