Embryological implications of partial Müllerian agenesis with vaginal adenosis
A case report

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Summary
The finding of adenosis in the vaginal pouch of a patient with agenesis of the lower Müllerian tract suggests that not all cases of vaginal adenosis are Müllerian in origin.

The ovarian biopsy specimen showed no abnormality and contained a normal complement of primordial follicles. The vaginal biopsy specimen consisted of two small fragments which were embedded and serially sectioned. Microscopic examination showed the smaller fragment to be lined by a non-keratinized, stratified squamous epithelium, which on staining with periodic acid-Schiff (PAS) was found to be normally glycogenated. The surface lining of the second and larger biopsy fragment resembled an immature squamous metaplastic epithelium in which nuclear atypia was evident (Fig. 1). The source of this metaplasia was traced to the opening of a communicating gland-like structure lying within the underlying stroma (Fig. 2).

Diethylstilboestrol (DES)-induced vaginal adenosis has enjoyed prominence in the gynaecological literature of the past decade. We report a case of a previously undescribed association of non-DES-related vaginal adenosis with lower Müllerian agenesis as a contribution to the available evidence concerning the pathogenesis of vaginal adenosis.

Case report
An 18-year-old nulliparous Black patient with primary amenorrhoea presented at the Department of Obstetrics and Gynaecology, Baragwanath Hospital, Johannesburg, during June 1979. There was no history of prenatal exposure to drugs. Examination revealed normally developed secondary sexual characteristics. There was no acne, hirsutism or virilism. The vagina ended blindly a few centimetres from the introitus. A biopsy specimen was taken of a small, raised erythematous area noted on the left vaginal wall within this blind pouch. No uterus was palpable. The pituitary-ovarian hormonal profile was within normal limits, and the chromosome pattern was 46,XX.

At laparoscopy the ovaries appeared normal. A biopsy specimen of the left ovary was taken. The fimbrial ends of the Fallopian tubes appeared to arise from the lateral walls of the pelvis. The medial parts of the Fallopian tubes, the uterus and the upper vagina were absent.

A diagnosis of agenesis of the lower Müllerian tract was made.

Histological examination
The ovarian biopsy specimen showed no abnormality and contained a normal complement of primordial follicles. The gland demonstrated a simple branching pattern, and was lined by a double layer of non-ciliated cuboidal-columnar epithelium (Fig. 3). The cytoplasm of the component cells was intensely eosinophilic and the nuclei were regular and hypochromatic. Nucleoli were absent. Small PAS-positive, diastase-resistant mucin droplets were seen in the apical portions

Fig. 1. Vaginal biopsy specimen showing immature, squamous metaplastic epithelium with atypical nuclear forms near the surface (H and E x 400).

Fig. 2. Branching gland-like structure within the vaginal stroma. Note the inflammatory cell infiltrate surrounding the gland (H and E x 100).
adenosis may occur in a patient with agenesis of the lower Müllerian tract is relevant to the controversial question of the definition and pathogenesis of vaginal adenosis.

Vaginal adenosis is currently defined as the abnormal presence of glandular epithelium in the vagina.1 The lesion described in this case fulfils these criteria. The condition is most commonly found in, but not limited to, patients exposed to DES in utero. Of the theories concerning the pathogenesis of vaginal adenosis, i.e. mesonephric, prosoplastic or Müllerian,5-7 the last mentioned appears to enjoy the widest acceptance. On the basis of an autopsy study Kurman and Scully5 argue against a mesonephric origin for three reasons. Firstly, mesonephric remnants were not found in their cases of adenosis, and vice versa. Secondly, in all their patients with adenosis the process was located superficially, whereas mesonephric structures are primarily located deep in the wall. Thirdly, the epithelium lining the glands in their cases of adenosis, although sometimes nonspecific in appearance, in other instances clearly resembled Müllerian-derived epithelium. In opposition to the prosoplastic theory of origin, which implies that the lesion is acquired, the authors show that it is congenital in at least some cases by demonstrating its presence in five fetuses. They cite as further evidence its frequent association with other congenital abnormalities.

Vaginal adenosis is, therefore, generally thought to be Müllerian in origin. It is postulated that for some reason the process of displacement of the lining of the fused Müllerian ducts, which form the upper vagina, by an upgrowth of urogenital sinus epithelium, is erratic or interrupted.8

Adenosis occurring in the vaginal pouch of a patient with agenesis of the lower Müllerian tract cannot be accounted for by the above mechanism, since Müllerian agenesis is accepted to represent primary failure of the Müllerian ducts to develop below a particular level, the vaginal pouch developing exclusively from the urogenital sinus. The adenosis in the patient described could, therefore, not have originated from Müllerian tissue. The most likely source is mesonephric tissue.

We would suggest that the reason for the conflicting reports in the gynaecological literature concerning the pathogenesis of vaginal adenosis has been the attempt to account for all cases by means of a single pathogenetic process, and that certain cases, such as the one described, differ from the majority in that they are derived from tissues other than the Müllerian tract.

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REFERENCES