FUNCTIONING OVARIES IN ASSOCIATION WITH RUDIMENTARY UTERUS AND ABSENT VAGINA

by

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Every case of primary amenorrhoea associated with a poorly developed vagina should have a buccal smear read for nuclear sex chromatin. This might prove to be proper counsel in the light of recent knowledge on intersexuality. In other words, one should endeavour to identify such intersexual conditions with female configuration as testicular feminizing syndrome and gonadal dysgenesis before deciding on any plastic surgery. But, on the other hand, a case is presented here as an example of yet another condition in which functioning ovaries are found in association with rudimentary uterus and absent vagina.

Case Report.

M. B., (3993/62) was admitted on 27th March 1962, complaining of pain in the right side of the lower abdomen since a month. She was a well-built married woman of 30 years. She presented normal feminine habitus and well-developed breasts. There was no evidence of hirsutism. Hair line of the scalp conformed to the female type. The axillary and pubic hair appeared adequate. External genitalia were normal and the vagina was of good length and capacity though no cervix or body of uterus could be made out. A rectal examination revealed a firm irregular mass, size of 2 c.cm. on the right side, vaguely suggestive of a gonad.

Investigations showed a buccal smear-positive for nuclear sex chromatin, an oestrogenic vaginal smear, and a negative Galli Mannini test. Facilities for estimating oestrogens and pregnandiol were, however, not available.

Previous History

She had been married for two years when she came to the hospital in 1949 for dyspareunia. An artificial cagina had successfully been constructed for her by the method of dissection of the recto-vaginal space and retaining a mould of balsa wood covered with a condom. She had been leading a successful married life ever since.

Her present complaint of pain in the abdomen was intermittent, low down on the right side and a laparotomy was undertaken to exclude a pelvic appendix.

On 18th April 1962, under general anaesthesia when the abdomen was opened by a sub-umbilical midline incision, normal looking ovaries presented themselves prominently (Fig. 1). They were found lying against the side walls of the pelvis. The tubes also appeared normally developed but when traced medially they ended in two rudimentary unicornuate uteri which were connected by a cord-like band about 4 cm. long. Further, two knob-like thickenings

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were noted about the middle of the transverse cord lying between the bladder and the rectum. The left horn was very small (1.5 cm. x 1.0 cm.). But the right one was larger and almost cubical, about 1.5 cm. in thickness and 2 cm. in length. An attempt to pass a probe into this from the abdominal ostium of the right tube proved futile. And when this cornu wos bisected a reddish brown velvety central area of 5 sq. mm. was noted. For the sake of biopsy a disc-like portion, 0.5 cm. in diameter, was resected from the larger cornu.

The ovaries were over 3 cm. long and 2 cm. in diameter. The surface was studded with follicles of varying sizes. On the left ovary was found an yellowish oval body about 1 cm. long with a **punctuate** body about 1 cm. long with a punctuate stoma in its centre, from which could be seem fine radiating blood vessels. This body, which was suggestive of a normal corpus luteum was shelled out.

No abnormalities could be appreciated in the anatomy of the ureters. The appendix appeared innocent and realising that the pain was probably ovarian in origin, the abdomen was closed and the patient made an uneventful recovery.

Fig. 2 shows the uterine musculature to be normal, but studded with embryonic endometrial glands and stroma. Fig. 3 is a microscopic section confirming the yellow mass to be corpus luteum in a stage of maturity. Fig. 4 is that of a frozen section of the same stained with the Sudan III showing lipoid-rich areas.

Comment

Although this condition was reported only once from Britain by Bender (1956), there is every reason to realise that it would frequently be noted by gynaecologists who study cases of primary amenorrhoea. A case of rudimentary uterus with normally functioning ovaries has to be differentiated from other cases of primary amenorrhoea possessing feminine secondary sex characters and external genitalia, such as testicular feminizing syndrome (Morris, 1953) and gonadal dysgenesia (Naidu and Mehdi, 1959).

The findings worthy of comment are the presence of functioning ovaries associated with hypoplastic non-functioning endometrium. The presence of a normal looking ovary and a corpus luteum prove also the existence of cyclic hormonal function. This also points to the possibility that ovaries would ordinarily carry on their function for several years even after hysterectomy. Grogan (1958) has reviewed the condition of residual ovaries after visualising them in a series of post-hysterectomy cases and found evidence of ovarian activity lasting from 8 to 16 years. The other important evidence of continued ovarian function is the finding of physiological status of the secondary sex characteristics both after hysterectomy as well as in the case presented.

Summary and Conclusion

1. Exclusion of intersexual conditions in cases of agenesis of vagina would be advisable before appropriate treatment is undertaken in the given case. While cases of testicular feminizing syndrome would require oestrogen therapy after gonadectomy, the type of case narrated here would not need any.

2. Since actively functioning ovaries may be found both with congenitally defective uterus as well as after hysterectomy, conservatism towards healthy ovaries should be the rule at the time of hysterctomy in the younger age groups.

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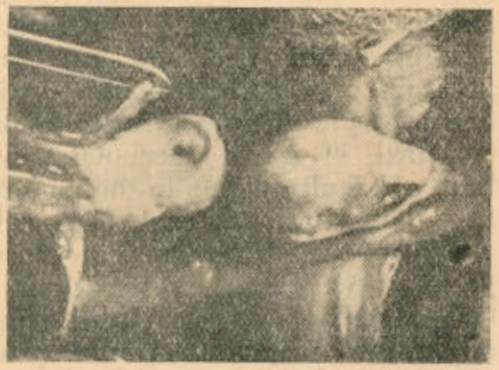

Fig. 1
Normal looking ovaries presenting themselves prominently.

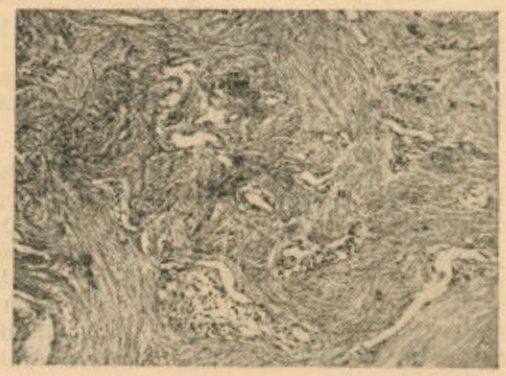

Fig. 2
Normal uterine musculature studded with embryonic endometrial glands and stroma (x 60 H & E).