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BILATERAL ADNEXAL AGENESIS WITH AN ECTOPIC OVARY — CASE REPORT AND REVIEW OF THE LITERATURE

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Bilateral adnexal agenesis (both ovaries and tubes) is not a common finding. Ectopic ovaries are also very rare. A report of the combination of the two, as far as we know, has never been published.

Such a case is presented here with its endocrinological evaluation. The literature concerning these entities is reviewed.

adnexal agenesis; ectopic ovary

CASE REPORT

A 27-yr-old gravida 0, para 0 woman was referred to our out-patient infertility clinic, to further evaluate the cause of her primary sterility.

Apart from appendectomy (the appendix was not inflammed) 20 yr earlier, she had no other relevant past medical history. Menarche was at age 11, and for about a year she recalled having quite regular menses. Then she became oligomenorrheic, and, at the age of 15, she became completely amenorrheic. She was given cyclic progesterone therapy and had withdrawal bleeding. She failed to respond to progesterone 18 mth later and was put on contraceptive pills. She recalled quite vividly that whenever she stopped the pills, she missed her periods altogether. Her breasts and other secondary sexual characteristics developed well, equal to those of other girls in her class. She married at the age of 21, and about a year later ceased taking the pills, planning to become pregnant. Failing to conceive and being amenorrheic, she sought gynecological help. She was given clomiphene citrate therapy for about 6 mth, but the basal body temperature curve remained monophasic. Cervical mucus was practically nil, and she had no response-bleeding to progesterone. She was then treated by her physician with gonadotropins with no response. Endometrial biopsy 10 mth after last uterine bleeding showed atrophic endometrium. When directly asked, she admitted to having hot flushes now and

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then, but attributed them to 'migranes'. Physical examination was completely normal, in so far as sexual characteristics were concerned (normally developed breasts, feminine hair distribution, unenlarged clitoris, no galactorrhea, normally developed vagina although somewhat 'dry', hypoplastic anteflexed uterus, no adnexal masses, no acromegaly, and no goiter).

Her hormonal profile was in the postmenopausal range with very high gonadotropins: FSH = 130 mIU/ml (N = 10.5 mIU/ml), LH = 107 mIU/ml (N = 10 mIU/ml), prolactin = 7.6 ng/ml (N = 5—15), testosterone = 0.57 ng/ml (N = 0.4—1.2), total urinary estrogens were 16 ng/24 h (N = 35—55). Thyroid function was normal, ACTH and dexamethasone tests were normal, X-ray tomography of her sella turcica and visual fields were all within normal range.

Again she responded only once to progesterone therapy. She did, however, bleed after cyclic estrogen and progesterone therapy (positive E + P test). Chromosomal evaluation (both buccal smear and skin biopsy) were normal feminine (46-XX) 18% Barr bodies.

In order to differentiate between 'early menopausal syndrome' and gonadotropin-resistant ovary syndrome (Koninx and Brosens, 1977), a gonadotropin loading test was done (during 5 consecutive days FSH + LH 225 LI Units = 3 amps) were injected intramuscularly with monitoring of blood 17β-estradiol, cervical mucus and vaginal cornification index. The results were unsatisfactory (minimal rise in 17β-estradiol, no estrogenic effects on cervical mucus, and a slight estrogenic effect on the cornification vaginal index). Hence we proceeded with the same test, this time under adrenal suppression by corticosteroids. The slight estrogenic effect we had seen the first time now disappeared completely, suggesting the adrenals to be the source of the estrogens. Laparoscopy was then performed. Since no ovaries could be seen, we proceeded with an explorative laparotomy. During the laparotomy, the following findings were observed.

The uterus was hypoplastic, measuring 3 × 3 × 2 cm. There were no tubes attached to the uterus and no ovaries, or remnants of ovaries (e.g. streak ovaries) were found. The pelvis was free from adhesions, suggesting that there had been no pelvic inflammatory diseases in the past. Since the lack of ovaries did not correlate with her past history of spontaneous menses, we proceeded to look for the ovaries. The retroperitoneum was explored at both sides ascending alongside the ureters, but no abnormality was found.

The peritoneal cavity was thoroughly explored. At the level of the umbilicus, on the right flank, adhered to the parietal peritoneum and buried in the omentum 10 to 15 cm above the pelvic rim, a white hard small mass, 1 × 1 × 1/2 cm in size was found. This mass, which looked like an ovary, had a small bluish cyst on its surface. The mass was excised with the adherent omentum and was sent to histopathological examinations. No other abnormality was detected. The postoperative course was uneventful.

PATHOLOGICAL FINDINGS

Microscopical examination revealed a white—yellow hard elastic mass measuring 1 × 1 × 1/2 cm and embedded in soft fatty tissue. Adjacent to this nodule we saw a cyst measuring 3 × 4 cm with a watery bluish content. The cut surface of the specimen showed ovarian-like tissue with an ovarian cyst. The inner and outer lining was smooth and there were no papillary structures.

The microscopical examination showed ovarian tissue embedded in fat. The ovarian cortex was thin but normal and contained a few primordial follicles, one growing follicle (Fig. 1), lined with several layers of granulosa cells; there was also a corpus albicans and some athretic follicles (Fig. 2). The cystic wall was lined with one layer of flat cuboidal cells almost without cilia and was a serous benign cyst (Fig. 3).

The ovary was surrounded by fatty tissue with congested blood vessels.

The pathological diagnosis was ectopic omental ovary with a serous benign cyst.



Fig. 1. Ovarian tissue with small athretic follicle embedded in omental fat (× 40).



Fig. 2. Ovarian tissue with corpus albicans within omental fat (×40).

DISCUSSION

Ovarian agenesis is a very rare phenomenon. Usually it is connected to genetic abnormality — more often than not to Turner syndrome. Other genetical abnormalities associated with ovarian agenesis are trisomy 13 and trisomy 18 (Kennedy et al., 1977), ataxia-telangiectasia (Miller and Chatten, 1967) and Di George's syndrome (Nishizuha and Aakakura, 1971). An exception is Swyer's syndrome (Swyer, 1955; Sarto and Opitz, 1973) ('pure gonadal dysgenesis'). Usually these females have eunochoidale habitus with normal external genitalia and streak ovaries (usually without follicles).

Adnexal agenesis (lack of both ovary and tube) which can be unilateral or bilateral, has also been described in the past. It can be the result of an inflammatory process or necrosis and absorption following torsion (Demopoulos et al., 1978; Sirisena, 1978).

It is curious to note that in this case only the uterine part of the Mullerian ducts developed with absence of the rest of the Mullerian system. We could not explain this finding with the above mentioned citations. However, in Rokitanski syndrome we can find a partially developed Mullerian system.

The distinction between two ovarian abnormalities, namely 'accessory ovary' and 'supernumerary ovaries' was clarified in Wharton's review in 1959. The term 'accessory ovary' refers to cases in which excess ovarian tissue is situated near the normally placed ovary and connected to it.

Supernumerary ovary includes those rare cases in which the third ovary is



Fig. 3. The wall of the ovarian serous cyst (×40).

entirely separated from the normally situated ovary. This third ovary may be situated in the greater omentum or retroperitoneally (Printz et al., 1973). This last entity is synonymous with the term 'ectopic ovary' in the literature.

In the above described case there was bilateral adnexal agenesis. The only ovary was situated in the omentum. Hence this is not a case of supernumerary ovary but a case of a true ectopic ovary. In such a case the ovary may or may not ovulate, but due to lack of the tubes, pregnancy is, of course, impossible.

In this case, the ectopic ovary ovulated for some time, and this explains the spontaneous menses the patient had experienced in the past. This is also proved histologically by the athretic bodies found in the specimen (see Fig. 1). Estrogen secretion from the follicles of the ectopic ovary explains the development of the secondary sexual characteristics.

It may be suggested that the blood supply to such an ectopic ovary is less than normal, and this shortens its life span. When estrogen secretion ceased, the patient started having hot flushes and became amenorrheic.

CONCLUSION

Since, in the literature that we reviewed, we could not find a case with bilateral adnexal agenesis and an ovary outside the pelvis, it seems to us that in this case the use of the term 'true ectopic ovary' is justified.

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