

Uterus-like mass of the ovary

Ovaryumun uterus benzeri kitlesel lezyonu

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ABSTRACT

We report an incidental case of uterus-like mass located in the right ovary. A mass, measuring 7 mm in its largest dimension was noted in the right ovary of a 54-year-old woman. Histologically, the lesion was resembling a miniature uterus with a cavity lined by a tubal-like epithelium, and surrounded with circumscribed smooth muscle bundles. Since 1981, 8 cases of uterus-like mass arising from the ovaries have been reported and their pathogenetic mechanism has been explained by two basic theories; congenital anomaly and metaplasia. The present case was incidentally found. The uterus was unicornus secondary to fusion defect of müllerian duct during embriogenesis supporting the congenital anomaly theory.

Key words: Uterus-like mass, ovary, congenital anomaly

ÖZET

Elli dört yaşında kadın hastanın total abdominal histerektomi ve bilateral salpingooferektomi materyalinde, rastlantısal olarak saptanan sağ ovaryum yerleşimli uterus benzeri kitlesel lezyon sunulmaktadır. Histolojik incelemede lezyonun, tuba epiteline benzer epitel ile döşeli kaviter alanı çevreleyen sirküler yerleşimli kalın düz kas demetlerinden oluştuğu, minyatür bir uterusu benzediği, 7 mm çaplı ve iyi sınırlı olduğu saptanmıştır. Literatürde ovaryum kökenli uterus benzeri kitlesel lezyon olarak tanımlanan 8 olgu yayınlanmıştır. Bu lezyonların gelişiminde konjenital anomali ve metaplazi teorileri öne sürülmüştür. Olgumuzda, uterus kavitesinin tek kornulu oluşu nedeniyle, gelişiminin embriyogenez sürecinde müllerian duktusun füzyon bozukluğuna bağlı konjenital anomali şeklinde geliştiği düşünülmüştür.

Anahtar sözcükler: Uterus benzeri kitle, ovaryum, konjenital anomali

INTRODUCTION

An uterus-like mass is a very rare condition that was first described by Cazutto in 1981 (1). This lesion was characterized by a central cavity lined with endometrial and tubal type mucosa surrounded by thick bundles of smooth muscle cells. Many cases of uterus like mass were reported under various terms such as adenomyoma, endomyometriosis and tumor of müllerian type since 1981 (2-5). Although, histogenesis of this lesion remains unknown, several hypotheses have been proposed. We hereby report an incidental lesion arising from the right ovary mimicking a miniature uterus.

CASE REPORT

A 54-year-old woman who had pelvic pain and postmenstrual bleeding was admitted to Gynecology and Obstetrics Department of Dokuz Eylül University Medical Faculty hospital. The patient had undergone abdominal hysterectomy with bilateral salphingo-oophorectomy with a clinical diagnosis of myoma uteri. The uterus weighed 250 g and measured 10x6x5 cm. The cavity was well developed with a single horn extending to the left side. The thickness of endometrium was 2 mm. Both ovaries were within normal dimensions. The cut surfaces revealed white areas concordant with corpus albicans and both ovaries had cystic lesions measuring less than 10 mm. Both tubae uterina were normal in appearance. The myomectomy material

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was white to green with a whorled cut surface, measuring 7x7x6 cm. Histopathologically, the cervix showed non-specific inflammatory changes. The endometrium was in proliferative phase and myometrium contained an adenomyotic focus. The described solid mass revealed typical microscopic features of a leiomyoma. Both ovaries had cystic follicles and corpora albicantia. Besides these, a nodular lesion localized within the ovarian stroma in close proximity to corpora albicantia was observed in the right ovary. Microscopically, this lesion measured 7 mm in diameter and composed of tubal-like mucosa and circular, thick smooth muscle bundles (Figures 1 and 2).

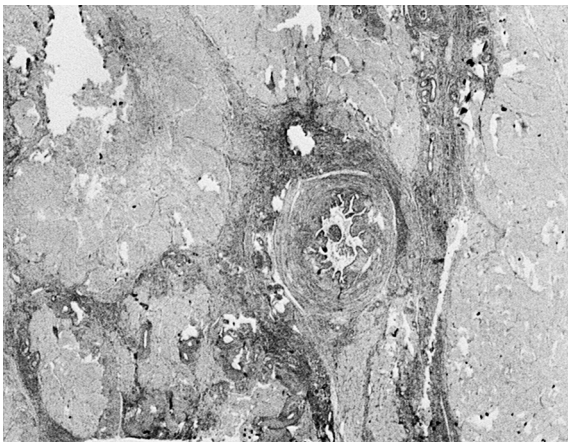


Figure 1. Microscopic appearance of uterus-like mass localized in the stroma of the ovary (HE x10).

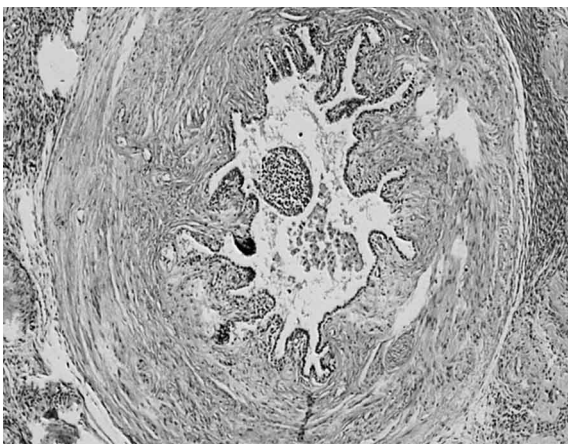


Figure 2. Cavity lined by tuba uterine like mucosa and surrounding thick and organized bundles of smooth muscle (HE x40).

DISCUSSION

Uterus-like mass of the ovary is a unique entity. Since 1981, 8 cases of ovarian uterus-like mass have been reported (1,4,6-9). In addition, extra-ovarian cases localized in the broad ligament, small bowel, small bowel mesentery, lumbosacral region, conus medullaris, lower uterine segment, uterosacral ligament, paratesticular region, left obturator lymph node, uterine cervix, and vaginal cuff wall were also described. Their sizes ranged from 4 to 16 cm with gross cystic and solid areas. Furthermore, an elevated CA125 level (4,10) (two cases), an associated breast carcinoma (4) (two cases), endometrioid carcinoma (7) (one case), uterine leiomyoma (11) (one case), lipoma (12) (one case), clonal chromosome deletion 2p21 (8) (one case), receiving estrogen therapy (10, 19) (2 cases,) and associated congenital anomalies (1,6,12) were also reported. Close relationship of this lesion with elevated serum levels of CA 125, associated neoplasms and estrogen therapy suggested the role of hormones in the pathogenesis of this lesion.

Histogenesis of uterus-like mass has not been clarified yet. However, two basic theories have been suggested. These are congenital anomaly and metaplasia theories (1,4,6-9,11,12,15). Also, heterotopia or choristoma theory was proposed by Peterson et al (18).

Congenital anomaly theory was favored by the presence of congenital organ abnormalities such as uterine or renal systems in several cases (1,3,9). Rosai et al. (13) suggested that the lesion reported by Cozzutto in 1981 as the first uterus-like mass, represented uterine tissue that was anatomically separate from the uterine corpus, not related with ovarian tissue. It was probably an example of uterine unicornis, associated with a rudimentary detached uterine horn in the contralateral adnexial region, representing Müllerian duct fusion defect or true partial duplication of the müllerian system (13). Formation of female genital tract depends on the müllerian

duct fusion of three separate portions from cranial to caudal during the embryonal development (14). Developmental anomaly of müllerian duct fusion in various sides or throughout the length of the duct may explain various duplications or atresias of the uterus. A case was suggested to be due to partial atresia of one of the müllerian ducts resulting in a uterine appendix whose central cavity did not connect with the vagina (6).

The metaplasia hypothesis attributes the histogenesis of the uterus like mass to endometriosis with subsequent smooth muscle metaplasia or to endomyometrial metaplasia resulting directly from the subcoelomic mesenchyme transformation. A case of uterine like mass arising in the broad ligament was suggested to arise from subperitoneal mesenchymal cells that retained the ability to duplicate müllerian duct (14,15). The subcoelomic mesenchyme (secondary müllerian system) is defined as the layer of tissue that lies underneath the mesothelial surface of the peritoneum (14,17). These tissues may proliferate in response to hormonal stimulation. A vital initiating role of the hormones in the pathogenesis of these lesions was suggested (4,17). A close relationship with breast cancer and elevated serum CA125 level was found. The essential role of hormones in the development of the lesions was supported by the development of uterus-like mass in the scrotum of men who received estrogen therapy for prostatic carcinoma (19). Also, abundant expression of estrogen and progesterone receptors in all cell types of the mass is reported (20). Furthermore, such masses in the ovary without any congenital anomalies of uterus or renal system were also described supporting the metaplasia theory (7,8).

Heterotopia theory was proposed by Peterson et al (18). They noted that neither the congenital nor the metaplasia theory was satisfactory for the explanation of some of the uterus-like masses. They reported an ileal uterine like mass that had morphological features of uterus and two smaller tubes like contiguous nodules

and various associated anomalies.

The present case supports the rudimentary horn concept. The uterus had a well formed cavity with a single horn extending to the left side and a small uterine like mass was localized to within the contralateral adnex. Also, a relatively smaller, well-organized mass lesion localized within ovarian tissue without any relation to subperitoneal mesenchyme might suggest the lesion to be a rudimentary horn secondary to localized fusion defect of müllerian ductal system.

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