

CASE REPORT**Uterus Like Mass of the Uterus: Report of a Case and Review of the Literature**

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Abstract:

We describe a case of 32 years old lady who presented with a uterine polypoid mass. Exploration revealed a uterus like mass i.e. a cavity lined by endometrial like tissue surrounded by smooth muscle layer resembling myometrium. We favour the mullerian duct fusion defect theory to explain the origin of the mass.

Key words: Uterus-like mass, Uterus

Introduction:

Uterus like mass is a rare entity proposed by Cozzuto in 1981 [1]. It represents a cavity lined by endometrium type mucosa surrounded by bundles of smooth muscle cells with a striking macroscopic and microscopic resemblance to the uterus. Very few cases involving ovary [2-4], broad ligament [5], mesentery [6], scrotum [7] have been reported, but involvement of uterus itself by uterus like mass is extremely rare. We report the case for its rarity.

Case:

A 32 years old lady was admitted with pain in lower abdomen and white discharge per vaginum of 15 days duration. Patient's general condition was good. Routine hematological investigations were within normal limits. Per speculum and per vaginal examinations showed a greyish mass protruding through the cervix. Ultrasonography showed a polypoid, solid cystic mass measuring 4x3x3 cm. arising from the fundus and projecting into the endometrial cavity.

Pathology :

We received hysterectomy specimen which on opening revealed a pedunculated mass measuring 4.5x 3.8x3.5 cm. attached to the fundus. Cut

Fig. 1: Gross photograph of hysterectomy specimen showing a polypoidal mass arising from fundus.



Fig. 2: Cut section showing a well formed cavity within the mass.



section of the mass showed a central, round cystic cavity measuring 2.5 cm. in diameter containing brownish fluid. (Fig. 1)

Microscopically, the mass was composed of thick muscular cyst wall lined by endometrial stroma covered with endometrial type mucosa resembling endometrium (Fig. 2).

Collections of hemosiderophages were seen in the endometrial stroma. The cyst wall consisted of thick smooth muscle bundles resembling

Fig. 3: The cavity lined by endometrial type mucosa surrounded by smooth muscle bundles resembling myometrium. (H&E, x40)

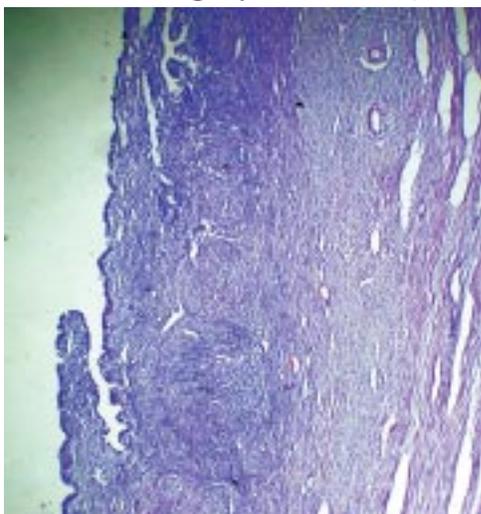
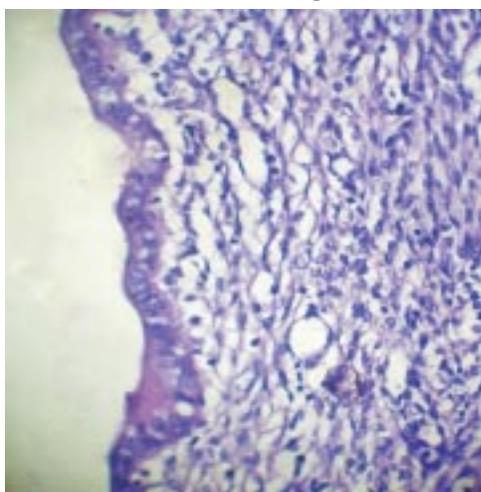


Fig. 4: Endometrial lining. (H&E, x400)



myometrium (Fig. 3).

Discussion:

Uterus like mass composed of cavities lined by endometrium type mucosa is a rare entity. The lesions have been reported in small intestine, small bowel mesentery, scrotum and extrauterine sites, [8-10] but involvement of the uterus is extremely rare. To the best of our knowledge, only 3 cases have been reported so far [11, 12].

Review of these cases showed a wide range of ages of 11-59 years. Clinical presentation was variable depending on the site of involvement and included lower abdominal pain with or without bleeding, dysuria, paresthesia and weakness of legs. Size of the mass ranged from 2.5 to 16 cm. All the cases shared similar gross appearance of solid cystic mass which on microscopy revealed uterus like organization *i.e.* outer thick myometrium composed of smooth muscle bundles and inner endometrium type mucosa lining the cystic space. Although the mass may simulate adenomyoma, uterus like organization differentiates it.

Histogenesis of this entity is uncertain. Three theories have been proposed.

- i. Uterine/mullerian duct fusion defect theory.
- ii. Subcoelomic mesenchyme transformation theory.
- iii. Heterotopia.

Cozzutto, who has documented the first case of uterus like mass in ovary, has postulated that the mass originated from ovarian stromal cells that underwent smooth muscle cell metaplasia in a focus of endometriosis. For the same case, Rosai has offered an alternative explanation and suggested mullerian duct fusion defect as

the origin of the mass.

The mullerian duct fusion defect theory is based on developmental abnormality occurring during formation of female genital tract. Embryologically, the female genital tract develops from the paramesonephric (mullerian) ducts, the unfused portions developing into fallopian tubes and fused portions into the uterus and cervix [13]. Lack of fusion of the mullerian ducts explains various duplications of the uterus. Rosai has proposed that a well formed uterus with a single horn, the uterus unicornis can be associated with a rudimentary detached uterine horn in the contralateral adnexal region, providing the most plausible explanation for the mass reported by Cozzuto. Ipsilateral renal agenesis in the same patient noted retrospectively also favours the mullerian duct fusion defect theory.

The subcoelomic mesenchyme theory challenges the fusion defect theory. Subcoelomic mesenchyme, the tissue layer beneath the mesothelial surface of the peritoneum embryologically gives rise to the mesenchyme of the urogenital ridges and forms the mullerian ducts by trapping the overlying mesothelium. Ahmed et al have reported uterus like mass in the broad ligament and have hypothesized that the mass could have originated from the subcoelomic mesenchyme which retains the ability to duplicate mullerian duct structures [5].

Redman et al have reported uterus like mass with features of an extrauterine adenomyoma presenting 22 years after total abdominal hysterectomy with bilateral salpingo-oophorectomy in a 50 year old lady. They have also supported metaplasia of subcoelomic mesenchyme theory as the patient had been receiving regular estro-

gen therapy because of premature menopause [8].

Pai et al have described 3 cases of uterus like mass of ovary, out of which 2 had elevated CA 125 levels giving a clinical suspicion of ovarian malignancy. Absence of any anatomic abnormalities and residual ovarian stroma suggested metaplasia rather than congenital anomaly as the cause of these masses [4].

Occurrence of uterus like mass in the scrotum of men receiving estrogen therapy for prostate carcinoma as described by Scully also supports the hormonal responsiveness of secondary mullerian system [7].

Peterson et al reported a 12 years old girl with uterus like mass in the ileum with multiple lower intestinal & urogenital tract abnormalities. She has had a past history of sacrocooccygeal teratoma. These authors have favoured the theory of heterotopia since neither the metaplastic nor the malformation theories could explain the case [14].

Conclusion:

We feel that uterus like masses involving the uterus, including our case can be best explained on the basis of congenital malformation i.e. mullerian duct fusion defect theory and should be differentiated from adenomyoma.

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