Uterus like ovarian mass in a young female clinically mimicking Carcinoma ovary

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Abstract
Uterus – like ovarian mass is a rare entity of uncertain histogenesis. A 30 year old female had a Left ovarian mass. Her preoperative serum CA 125 was 700U/ml. So a clinical diagnosis of Carcinoma ovary was made. We received left salpingo-opherectomy specimen for intraoperative frozen consultation. On gross examination, left ovarian mass measured 10x8x6.5cm. External surface was smooth and showed congested blood vessels. On cutting, mass was partly solid and partly cystic showing small cystic cavities filled with blood mixed fluid. The cyst wall surrounding the cavities was remarkably thickened. Microscopic examination of frozen section revealed a lesion predominantly composed of benign spindle cells. So we gave a diagnosis of benign cystic non neoplastic lesion on frozen section. Formalin fixed paraffin embedded sections followed. Microscopic examination of formalin fixed paraffin embedded sections revealed a cystic mass lined by endometrial type of lining and stroma surrounded by remarkable smooth muscle hyperplasia. In areas, haemorrhage, infiltration by hemosiderophages and hemorrhagic corpus luteum were also seen. So we gave a final diagnosis of uterus like ovarian mass.

To the best of our knowledge, only 15 cases of extrauterine uterus like masses have been reported in the literature. Of these, 5 cases were of ovarian origin. This report documents a sixth case of uterus-like ovarian mass.

Keywords: Uterus like ovarian mass in a young female clinically mimicking Carcinoma ovary.

Introduction
Uterus – like ovarian mass is a rare entity of uncertain histogenesis. Uterus like extra uterine-masses within the pelvis may arise in the broad ligament, fallopian tube, or ovary⁵. It represents a unique arrangement of a cavity lined by endometrial mucosa surrounded by bundles of smooth muscle fibres, replacing the normal structure of ovary. To the best of our knowledge, only 15 cases of extrauterine uterus like masses have been reported in the literature⁶. Of these, 5 cases were of ovarian origin.³-⁷ This report documents a sixth case of uterus-like ovarian mass.

Case Report
A 30 year old female had a Left ovarian mass. Her preoperative serum CA 125 was 700U/ml. So a clinical diagnosis of Carcinoma ovary was made. We received left salpingo-opherectomy specimen for intraoperative frozen consultation. On gross examination, left ovarian mass measured 10x8x6.5cm. External surface was smooth and showed congested blood vessels. On cutting, mass was partly solid and partly cystic showing small cystic cavities filled with blood mixed fluid. The cyst wall surrounding the cavities was remarkably thickened (Fig. 1). Microscopic examination of frozen section revealed a lesion predominantly composed of benign spindle cells. So we gave a diagnosis of benign cystic non neoplastic lesion on frozen section. Formalin fixed paraffin embedded sections followed. Microscopic examination of formalin fixed paraffin embedded sections revealed a cystic mass lined by endometrial type of lining and stroma surrounded by remarkable smooth muscle hyperplasia (Fig. 2). In areas, haemorrhage, infiltration by hemosiderophages and hemorrhagic corpus luteum were also seen. So we gave a final diagnosis of uterus like ovarian mass.

Fig. 1: Gross specimen of ovarian mass showing small cystic cavities containing hemorrhagic fluid. Cyst wall surrounding the cavities is remarkably thickened

Fig. 2: Cyst wall lined by endometrial glands and stroma. Wall shows marked smooth muscle proliferation
Discussion

The Uterus like ovarian masses are extremely rare. The first uterus-like mass was described by Cozzutto in 1981. Since then only 15 cases have been published in the literature. Of these 12 cases were located within pelvis and 5 cases arising in the ovaries. Rare cases originating from broad ligament, liver and mesentry have been reported. Two extraovarian cases were reported at pelvic location after hysterectomy.

Sizes ranged from 6 to 16 cm. The gross appearance, including cystic and solid areas, varied between cases. In our case, size was 10x8x6.5cm and on gross examination of the mass was solid and cystic in appearance.

The microscopic descriptions of each mass included functional endometrial glands and stroma within a muscular walled structure resembling a uterus. In our case microscopic appearance also revealed same thing.

Ovarian endometriosis is associated with elevated serum CA 125 levels. In our case raised CA 125 levels were probably due to ovarian endometriosis.

The pathogenesis of uterus-like masses remains uncertain. When we reviewed literature, we found out four theories to explain this occurrence. The first theory is mullerian duct fusion defect theory is based on a developmental abnormality occurring during the formation of the female genital tract. The second theory is sub-coelomic mesenchyme transformation. Third is mullerianiosis theory defined by Batt as a heterotopic organoid structure composed of Mullerian rests that were incorporated within other normal organs during organogenesis.

Endometriosis with prominent smooth muscle hyperplasia or metaplasia, which has also been called endomyometriosis, is the fourth theory to explain pathogenesis. Pathogenesis in our case is best explained by the fourth theory.

Conclusion

In conclusion, these rare lesions present with ovarian mass having raised CA 125 levels. Hence, they are clinically mistaken for Ca ovary. Therefore, one must diagnose such lesions precisely, as they are benign and just require surgical excision. In the coming years, we are likely to encounter such lesions during intra-operative frozen sections as in our case.

References