A RARE CASE OF EXTRA OVARIAN SEX CORD STROMAL TUMOUR PRESENTING AS BROAD LIGAMENT MASS SYNCHRONOUS OVARIAN MASS; PATHOLOGIC PATTERN REVEALS MIX GRANULOSA FIBROMA, TYPE OF MIX MESONEPHRIC AND MULLERIAN ORIGIN

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PRESENTATION OF CASE

A 30 yrs. female presented to OPD with a mass abdomen. FNAC of the mass showed papillary-adenocarcinoma. She had undergone B/L excision of the mass, which was a broad ligament mass, revealed on laparotomy. HPS and IHC revealed it to be malignant high-grade mixed sex cord stromal tumour (granulosa and fibroma type).

DIFFERENTIAL DIAGNOSIS

These tumours are to be differentiated from other small cell carcinomas like undifferentiated sarcomas, endometrial stromal sarcoma and lymphoma, by a panel of IHC inhibin, CK, EMA, Chromogranin, CD 10. They have to be differentiated from primary broad ligament carcinoma, which has a papillary arrangement of cells, with foci of transitional cells. As the initial FNAC showed adenocarcinoma and absence of transitional cells, this excludes primary endometria broad ligament carcinoma.

CLINICAL DIAGNOSIS

High grade extra ovarian mix sex cord stromal, probably of Mullerian and mesonephric origin, associated with B/L synchronous ovarian tumour.

DISCUSSION OF MANAGEMENT

Hysterectomy and B/L Salpingoopherectomy, with Tumour Debulking.

Role of adjuvant chemotherapy and radiotherapy is unknown.

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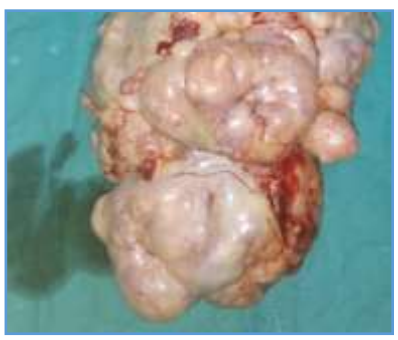

Figure 1. The Mass of Size 15x20 cm, Greyish Yellow, with Solid and Cystic, Haemorrhagic Areas

Figure 2. Synchronous Ovarian Mass

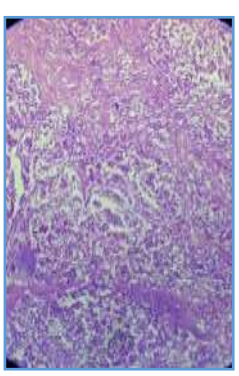

Figure 3. HPS-Granulose Cell Tumour of the Case

PATHOLOGICAL DISCUSSION

The patient presented as B/L broad ligament mass with normal uterus and B/L ovaries. There are number of tumour markers, like calretinin, inhibin +ve to confirm it to be a granulosa type. The positive stain for vimentin and spindle cells favours a stomal fibrous component. CD 99 +ve favours

sex cord tumour. The weakly positive WT1, and chromogranin, CK and SMA, favours mix origin Mullerian, coelomic and mesonephric origin. Rarely can develop from extraovarian site, broad ligament, retroperitoneum, mesentery, Liver adrenals[1] histogenetic origin from ectopic stromal tissue from mesonephros.[2]

GCTs vary in their gross appearance. Most are partly solid and partly cystic.[3] Microscopically, the tumour cells resemble normal granulose cells. they are small round or oval nuclei with fold longitudinal grooves and the folds they show a predominant trabecular and diffuse pattern, which was pattern in the above case.[4] A very interesting theory of ovulation and extra ovarian origin of ovarian cancer, as in this case, with a synchronous ovarian cancer, i.e ovulation providing and chemotactic environment for attraction of tumour elsewhere.[5] SDF-1 secreted by the granulose cells aids in chemotaxis of, embryonic germs cells, other tissue specific cells outside, like the broad ligament in this case. This has been proved in animal models.

The interaction of SDF-1 and CXCR-4 activates downstream signaling pathways that can result in chemotaxis, cell proliferation and survival, migration and gene transcription.[6] After ovulation ovarian stroma collagen IV provides a scaffold for adhesion of extra ovarian malignant cells.[7,8] The above theory could explain the synchronicity of ovarian tumour in the case.

A possible dual origin of extra ovarian GCT, i.e. from the coelomic and mesonephric origin has also been proposed.[9] Mesonephros or its influence seems to be necessary for creating the sex cord. This may also explain the origin of sex cord stromal tumours being limited to the broad ligament, the retroperitoneum and the adrenal, all of which differentiate close to mesonephros and mesonephric duct.[9]

The morphological differential diagnoses of GCT includes undifferentiated carcinoma, small cell carcinoma and endometrial stromal sarcoma. The characteristic immune-stains and histology has been described above. Extra-ovarian sex cord stromal in the broad ligament is a rare entity. The histogenetic origin of sex cord is thought to be from the ectopic gonadal stromal tissue, with sex cord originating from the mesonephros. A possible dual origin from both the coelomic and mesonephros has been proposed. Review literature reveals cases of extraovarian GCT in broad ligament, retroperitoneum. Cases of GCT from a Mullerian cyst in broad ligament has been reported.

Prognosis- high chances of recurrence and relapses. 17% relapses occur in more than 10 years of diagnosis.[5]

This case showed a resistance to first line of adjuvant chemotherapy (etoposide+carboplatin), there was progression of disease. The case is reported for its rarity and to describe its relevance to histogenetic origin and clinical practice.

Abbreviations

GCT – Granulosa Cell Tumour.

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