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Case Report
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A Rare Case of Extraovarian Mixed Sex Cord Stromal Tumor Presented as Broad Ligament Mass, Pathologic Pattern Reveals a Mix Granulosa, Fibroma Type of a Mix Origin of Mesonephric and Coelomic Origin

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Abstract

A 30 yrs female presented to opd with a mass abdomen. Fnac of the mass showed pappilary-adenocarcinoma. She had undergone b/l excision of the mass. Which was a broad ligament mass, revealed on laparotomy.HPS and IHC revealed to be malignant high grade mixed sex cord stromal tumor (granulosa and fibroma type).

Abbreviations

GCT: Granulosa Cell Tumor

Introduction

Extra-Ovarian sexcord stromal in the broad ligament is a rare entity. The histogenetic origin of sexcord is thought to be from the ectopic gonada 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 muellerian cyst in broad ligament has been reported.

Case Presentation

A 30 yrs female presented to opd with mass abdomen .o/e patient was of normal built with no pallor and lymphadenopathy .on p/a the mass suprapubic firm to cystic non tender with restricted mobility .p/vp/r the mass was felt separate from uterus firm to cystic in consistency, uterus retroverted.

Investigations

Usg: A well defined in homogenous hypoechoic mass of approx 12 x10x10 cm is seen in right lower abdomen. The mass is close to ovary and free from uterus, right kidney, liver and gall bladder. No calcification/echo free area is seen within the mass.

Fnac well differentiated papillary adenocarcinoma

Cect: large well defined heterogeneously enhanced mass lesion in right lumbar and pelvic region (broad ligament fibroid/ gist). b/l ovaries and uterus normal. With minimal fluid collection.

Surgery: it was broad ligament mass (b/l) +omental nodules.b/l ovaries and tubes and uterus normal.

b/l excision of mass + omentectomy

hps: high grade malignant tumor epithelial tumor. With focal areas of spindle cells.

IHC: the discordance between clinical and pathological context we prefer a confirmation by IHC.

EMA was positive suggestive of mullerian origin. Calretinin and in hibin strongly suggestive of granulosa cell tumor. Broad ligament mass vimentin positive, ema, calretinin positive. CK -7 +, WT1+, CD99+ favours mullerian origin. Calretinin, CD99 is positive in sex cord stromal tumor.

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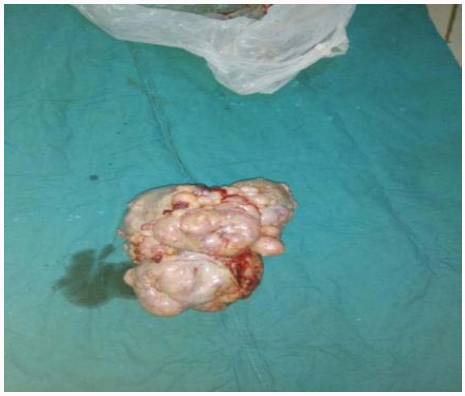

Figure 1: Description: the mass of size 15x20 cm, greyish yellow, with solid and cystic, haemoragic areas.

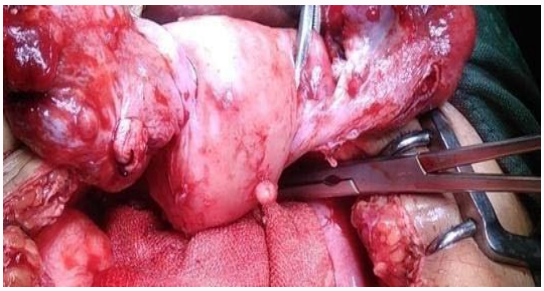

Figure 2: Synchronous ovarian mass.

Vimentin positive in favour of mesenchymal component.

- Received etoposide + carboplatin paclitaxel cisplatin

During treatment the mass reappeared and did not respond to chemotherapy. She presented again with mass abdomen.

o/e – moderate pallor , no icterus no lymphadenopathy , no pedal oedema p/a - 28 wks size mass variagated consistency ill-defined borders restricted mobility non tender, ascites +

- p/v p/r - uterus ns , pod nodules, the lower limit of mass felt and the fornics were full .
- CA 125 - 18.3iu/ml
- Inhibin A - 2.8 iu/ml
- CECT ON 8 /5 /18 –CECT 15 x12 CM abdomino pelvic mass
- omentum involved, pod deposits
- Left lower lobe of lung 4.2 x 3 mm
- 10 x 12 mm lesion in liver.
- Upper gi endoscopy normal
- Inhibin A -27.2iu/ml
- Afp -1.97, bhcg - .23IU, CEA - 6.5IU
- Ldh - 492IU
- Plan - laparotomy , TAH +BSO and excision of the mass
- Iop findings - haemoragic ascites, solid irregular varieagated mass 20 x15 cm in the pelvis was found adherent to bowel. sigmoid adherent to tumor
- b/l ovaries surface irregular.

Multiple peritoneal, diaphragm and liver surface tumor deposits

- Specimen sent for hps and ihc
- Hps - gross – abdominal tumor 20 x 15x 10 cm. surface nodular cut section partly solid and partly cystic with haemorragic areas. uterus and cervix unremarkable
- Right ovary 4x2.5x2.5cut section solid and cystic containing clear fluid left ovary 3x2.5 1.5 cm cut section solid and grey
- Microscopic examination –malignant undifferentiated tumor(similar to the first biopsy)in the abdominal mass
- Presence of tumor in both ovaries
- Absence of surface deposits
- b/l tubes endomyo and cervix free of tumor
- IMP-malignant mix sex cord stromal tumor
- IHC panel negative for – CK 7, CK 20, CDX2, TTF-1,WT-1,EMA,CD10,Inhibin, synaptophysin and melanin
- Focal strong positive punctuate positive in tumor cells-chromogranin and SMA, CK.
- Chromogranin positive is suggestive of mesonephric component. Calretinin -positive
- Imp - adult granulosa tumor of extra ovarian origin but mix muellerian, coelomic and mesonephric origin.

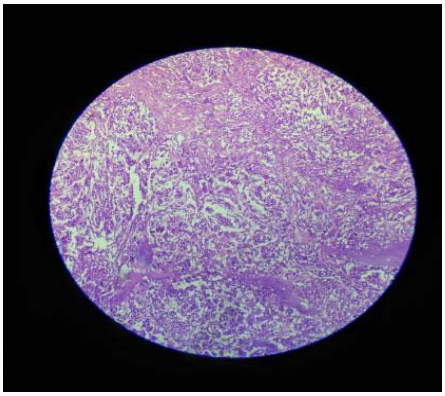

Figure 3: Slide image.

Clinical Diagnosis

Interpretation of the hps and ihc and anatomic location, and the initial presentation of the tumor and its recurrence pattern, reveal it to be an high grade extra ovarian mix sex cord stromal, propably of mullerian and mesonephric origin, associated with b/l synchronous ovarian tumor (Figure 1). Extraovarian mix sexcord stromal can arise in locations other than ovary, and is said to derive from mesenchyme of genital bridge.

Differential Diagnosis

These tumors are to be differentiated from other small cell carcinomas. Undifferentiated sarcomas endometrial stromal sarcoma, lymphoma, by a panel of ihc inhibin, ck, ema chromogranin cd 10.

They are to be differentiated from primary broad ligament carcinoma, which has a pappilary arrangement of cells, with foci of transistional cells (Figure 2 and 3). As the initial fnac showed an adenocarcinoma, but absence of transistional cells, this excludes primary endometroid broad ligament carcinoma.

Management

Hysterectomy and b/l salpingo-oophorectomy; eith tumor debulking

Role of adjuvant chemotherapy and radiotherapy is unknown.

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.

Discussion

The patient presented as b/l broad ligament mass with normal uerus and b/l ovaries. There are number of tumor markers, like calretinin, inhibin +ve to confirm it to be a granulosa type. The positive stain for vimentin and spindle cells favour a stomal fibrous component. CD 99 +ve favours sexcord tumor. The weakly positive WT1, and chromagranin, ck and sma, favours mix origin muellerian, coelomic and mesonephric origin. Rarely can develop from extra ovarian site, they, broad ligament, retroperitoneum, mesentery, liveradrenals [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 tumor cells resemble normal granulose cells. they are small round or oval nuclei with fold longitudinal grooves and the folds they show apredominate 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 tumor 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 cxcr4 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 synchronocity of ovarian tumor 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 sexcord. This may also explain the origin of sex cord stromal tumors being limited to the broad ligament, the retroperitoneum and the adrenal, all of which differentiate close to mesonephros and mesonephric duct [10]. The morphological differential diagnoses of GCT includes undifferentiated carcinoma, small cell carcinoma and endometrial stromal sarcoma. The characteristic immunostains and histology has been described above text. The case is reported for its rarity and to describe its relevance to histogenetic origin and clinical practice.

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