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RESEARCH ARTICLE

BILATERAL MALIGNANT OVARIAN SERTOLI-LEYDIG CELLTUMOR :A CASE REPORT

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Abstract

Ovarian stromal and or sex cord tumors develop from the supportive tissue of the ovaries. Within this group of tumors, we distinguish granulosa tumors, androblastomas(Sertoli Leydig tumors), sex cord tumors with annular tubules, steroid cell tumors and fibrosarcomas. Most of these tumors synthesize ovarian hormones: estrogens, androgens and corticosteroids. Sertoli Leydig tumors are almost always benign, while others are malignant with locoregional recurrences. Their prognosis is uncertain. The poorly differentiated forms of Sertoli-Leydig tumors have a significant potential for malignancy. We report the case of a 45-year-old patientwith a large non-secreting ovarian tumor, whose biopsy suggested a Sertoli-Leydig tumor, of intermediate differentiation. We decided to perform surgical excision, which revealed a tumor at the stage of peritoneal carcinomatosis. And a cytoreductive procedure was performed. Surgery is the mainstay of treatment, and should be supplemented by chemotherapy in case of malignancy.

Introduction:-

Ovarian tumors that develop from supporting tissue account for about 8% of ovarian tumors, of which 3% are granulosa cell tumors (1).

Sertoli Leydig tumors arise from the mesenchyme and sexual cord, which in a histological aspect include all phases of embryonic development of the testis.

They are mostly benign, but recurrences or metastasis occur in about 20% of cases(1).

We report the case of a bilateral ovarian tumor at the stage of peritoneal carcinomatosis.

Case report :
We report the case of a45 years old women, mother of two children,still of reproductive age.

With a history of recent metrorrhagia (two months prior to hospitalization), repeated post-prandial vomiting, and a weight loss of 6 kg.

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Physical examination revealed a largepalpable abdominopelvic mass with moderate ascites.

Clinical findings were very suggestive of an ovarian tumor. There was no clinical secretory syndrome (absence of virilization signs).

The patient's biological tests showed no abnormalities, and tumor marker tests were normal (CEA, CA19-9, AFP and CA125).

An abdominal ultrasound was performed, and revealed a large abdominal-pelvic tumor of approximately 30 cm with both a fluid and tissue component, well defined, with regular contours (Figure 1,2).

A CT scan was performed, whichshowed a bilateral ovarian tumor, measuring 13*11 cm on the left and 19*14 cm on the right, both were solid heterogeneous tumors, with large necrotic areas.There were some regional lymph nodes, and mild ascites. There was no bone, nor hepatic or pulmonary metastases (Figure3).

A suspicious thickening of thebulbo-duodenal junctionsuggests a Krukenberg tumor.Upper GI endoscopyshowedan extrinsic compression of the second portion of the duodenum, andno primary gastric tumor.

An Ultrasound-guided biopsy, confirmed by immunohistochemistrythe diagnosis of a Sertoli Leydig tumor.

The patient was discussed at the multidisciplinary team meeting and the decision was to proceed to surgery.

We performed a laparotomy through a midline suprapubic incision, Explorationrevealed a bilateral solid ovarian tumor,ascites and peritoneal carcinomatosis (Figure 4,6).

Total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and appendectomy was performed(Figure 5,7).

The postoperative course was uneventful.

The excised mass was sent to the pathology department,which they concluded an ovarian androblastoma (Sertoli-Leydig tumor) of intermediate differentiation, with a heterologous component of intestinal glandular type, extending to the omentum and the rest of the peritoneum. Immunohistochemistry showed that Sertoli and Leydig cells expressed, among others, inhibin, and the glandular structures expressed cytokeratin 7 and 20.

Adjuvant chemotherapy was decided on the multidisciplinary meeting.

After athreemonth follow-up,the patient improved and gained additional weight,and the radiological follow-up revealed theabsence of ascites and disappearance of peritoneal lesions.

The patient consulted eight month later, as she presented with jaundice. The abdominal CT scan showed peritoneal relapse. An external biliary drainage was performed and chemotherapy was pursued. But the patient died at one-year follow-up.

Discussion:-

Sertoli Leydig tumors are rare, they represent 0.2% of all ovarian tumors (**1-2).**

Their incidence is around the age of 25 (3). However, there are some cases observed beyond the age of 45 (**4).**

These tumors are classified into 3 groups: well-differentiated benign forms, which are secretory in 60% of cases and are less frequent.Forms with intermediate differentiation (immature Sertoli cell, associated with a stroma made up of Leydig cells, in a small proportion). And the poorly differentiated forms, composed of spindle cells with a pseudo-sarcomatous appearance **(4)**. Some tumors may also contain heterologous elements, such as bone tissue, gastrointestinal-type epithelium, skeletal muscle or hepatocytes (**4).**

The bilateral and synchronous involvement of both ovaries is rare (**5**). Bilateral tumors represent less than 5% of cases(**4).** Most patients present with non-specific symptoms, such as abdominal symptoms,or menstrual disorders. Signs of virilization (which are absent in our patient) are present in 35-50% of patients (**6).**

Cancer cells release the male sex hormone (testosterone),as a result the woman may develop signs of virilization.Three clinical signs are revealing, which are amenorrhea, deepening of the voice, and hirsutism, often accompanied by clitoromegaly.Exceptionally, these tumors can secrete estrogens, which sometimes results in precocious puberty.In the presence of virilization signs, it is advisable to measure the main major androgens levels in women. In general, only ovarian derived androgens are elevated. In the presence of amenorrhea, the dosage of androgens should be coupled with that of pituitary hormones (FSH, LH, Prolactin). However, it is important not to systematically perform hormonal assessment in case of an isolated tumor syndrome (**3-4)**.

Other markers have been proposed by some authors for biological monitoring, such as alpha-fetoprotein, inhibin A and inhibin B **(4-5).**

Diagnosis relies on ultrasound and abdominal CT.Pelvic MRI may be useful in case of a diagnostic uncertaintyregarding a uterine or a small ovarian tumor (**4).**Sertoli-Leydig tumors appears as solid and lobulated tumors, sometimes cystic, heterogeneous and vascularized tumors. Their size can vary, reaching up to twenty centimeters (**4-7)**.

Its management is not standardized (**8).**

Sertoli-Leydig tumors have a low malignant potential. Their treatment is mainly based on surgery, and excision of all lesions is the mainstay of treatment. In early stages and a unilateral ovarian tumor, conservative surgery (adnexectomy or oophorectomy)is recommended, to preserve the desire for pregnancy(**3).** The prognosis is excellent, especially if the tumor is well differentiated. This procedure can be performed by laparoscopy (**4).**

Poor prognostic forms are rare, and represent less than 20% of cases (**4)**.Malignant Sertoli-Leydig tumors should be treated by surgery, and radical excision should follow the same rules of surgical management of epithelial ovarian cancer (**4-6).**

Chemotherapy protocols used for Sertoli-Leydig tumors are similar to those used in malignant germ cell tumors. There have been some reports of partial and complete response to chemotherapy **(3-4).**

Prognosis is relatively good in case of early diagnosis with a lesion still confined to the ovary (**8)**. Early recurrences may occur in case of malignant Sertoli Leydig tumors (**1)**. Poor prognostic factors are large tumor volume, late stage (peritoneal carcinomatosis), histological grade (poorly differentiated form), tumor rupture, presence of heterologous elements, and the number of mitoses (**1).**

Clinical, biological and radiological follow-up is mandatory. Patients with signs of virilization, can be monitored by measuring DHEA level, estrogen, 17-hydroxyprogesterone and cortisol. Feminine characteristics usually return after surgery,but male characteristics resolve more slowly (**9).**

Abreviations :
Kg : kilogram
CEA :carcinoembryonic antigen
CA19 9 : carbohydrate antigen19-9
CA125 :cancer antigen125
AFP : alpha fetoprotein
FSH :follicle stimulating hormone
LH :luteinizing hormone

Figures :

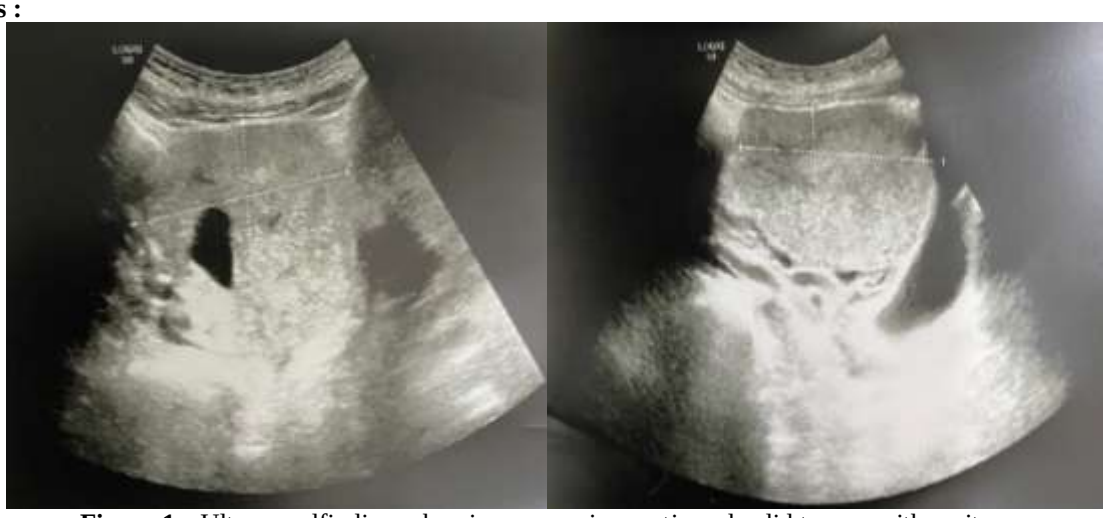


Figure 1:- Ultrasoundfindings showing an ovarian cystic and solid tumor, with ascites.

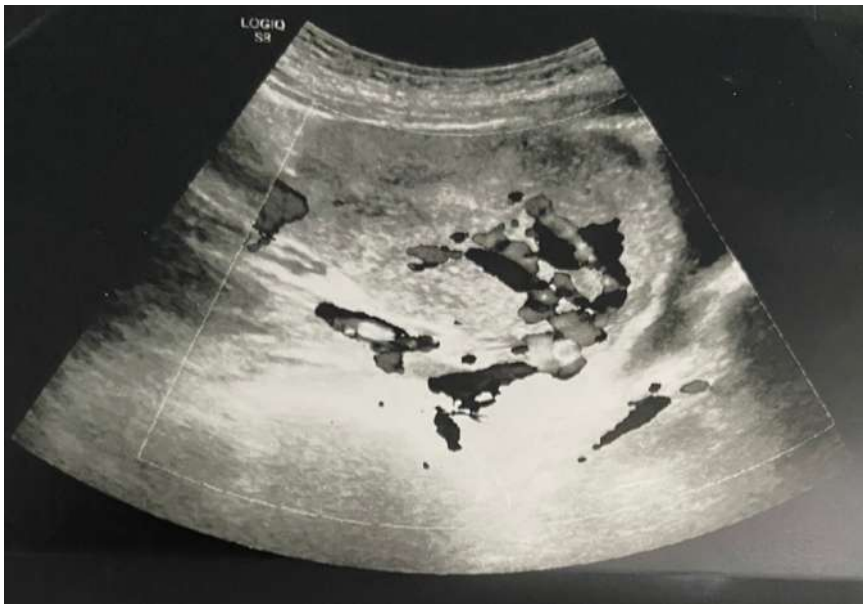
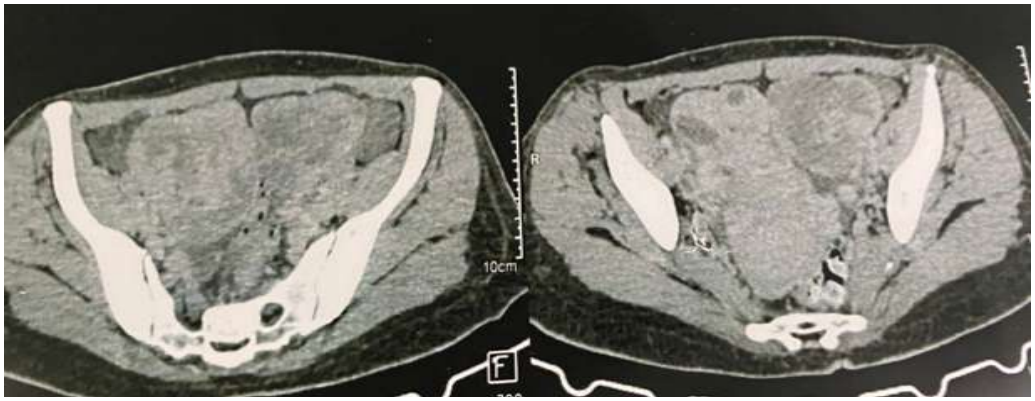


Figure 2:- Doppler ultrasound showing a hypervascularized ovarian tumor.



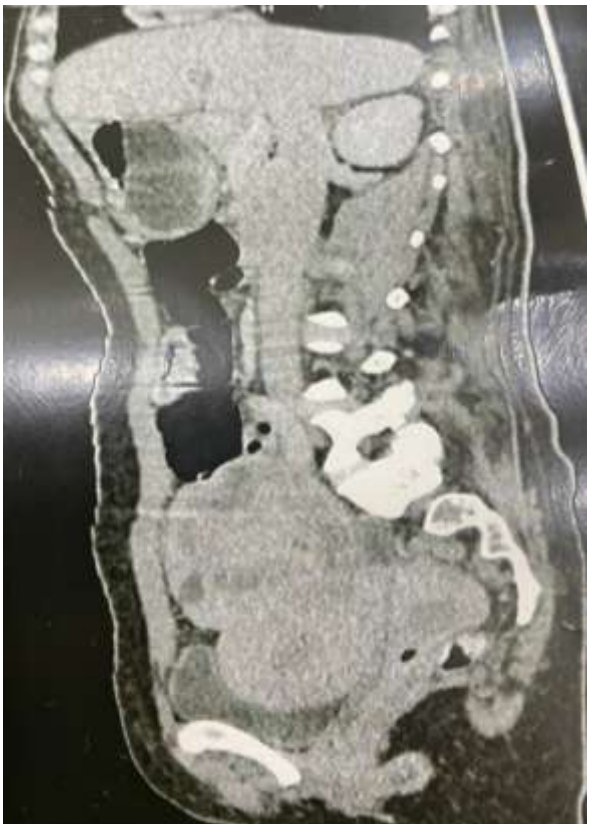


Figure 3:- Abdomino-pelvicCT scan,showing a bilateral cystic-solid ovarian tumor (axial and sagittal plane).

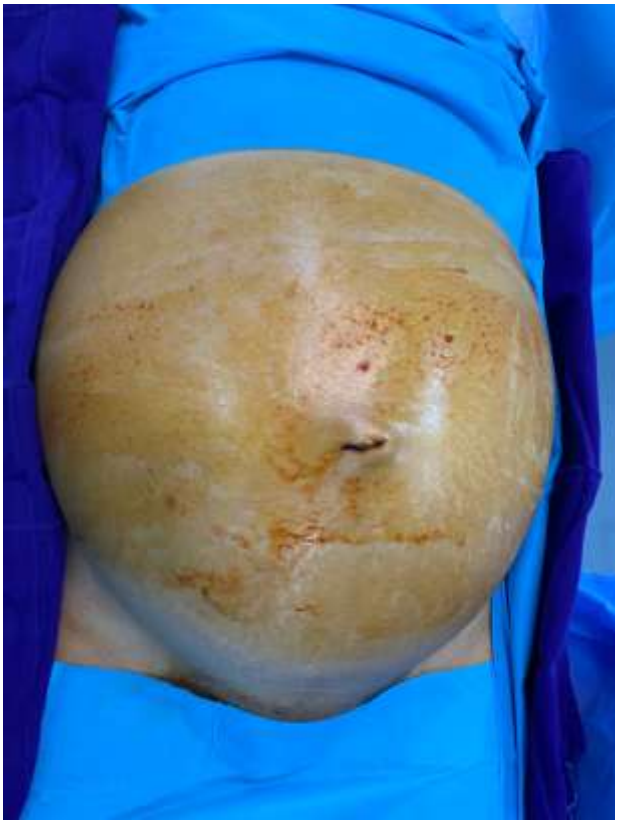


Figure 4:- Preoperative image of the abdomen of the patient.

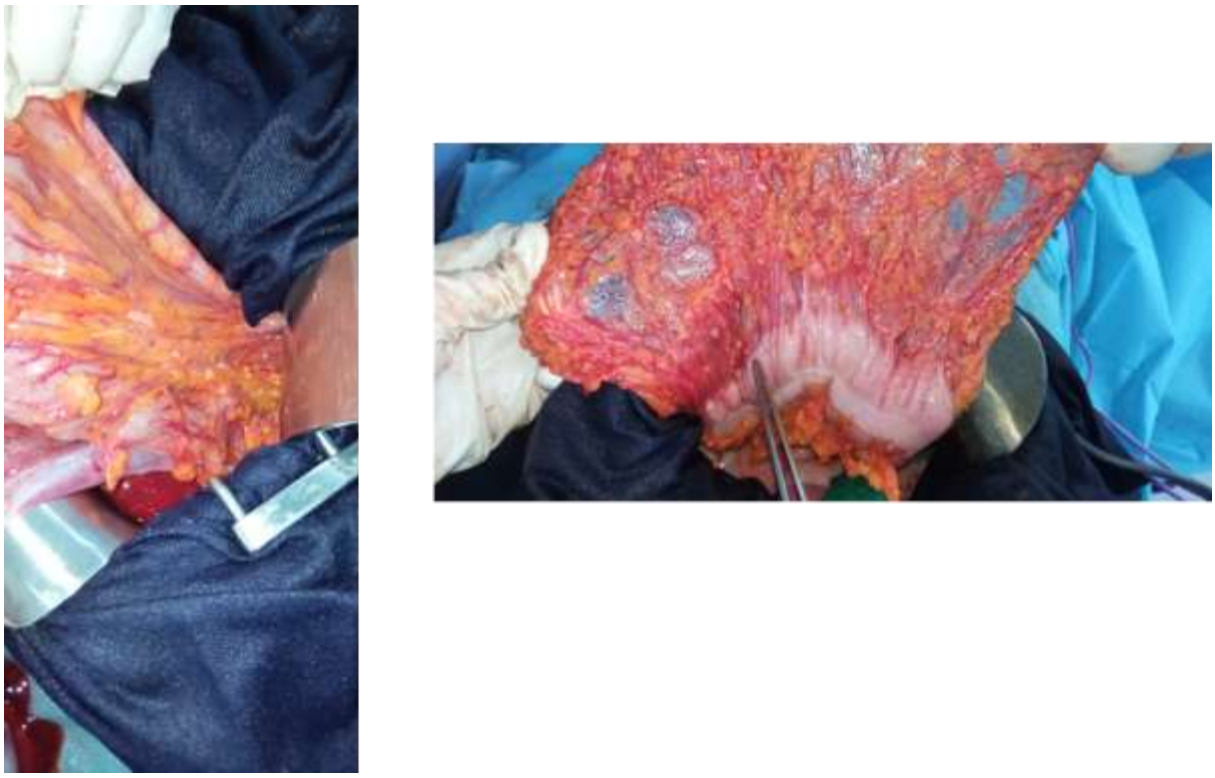


Figure 5:- Operative view showingperitoneal carcinomatosis.

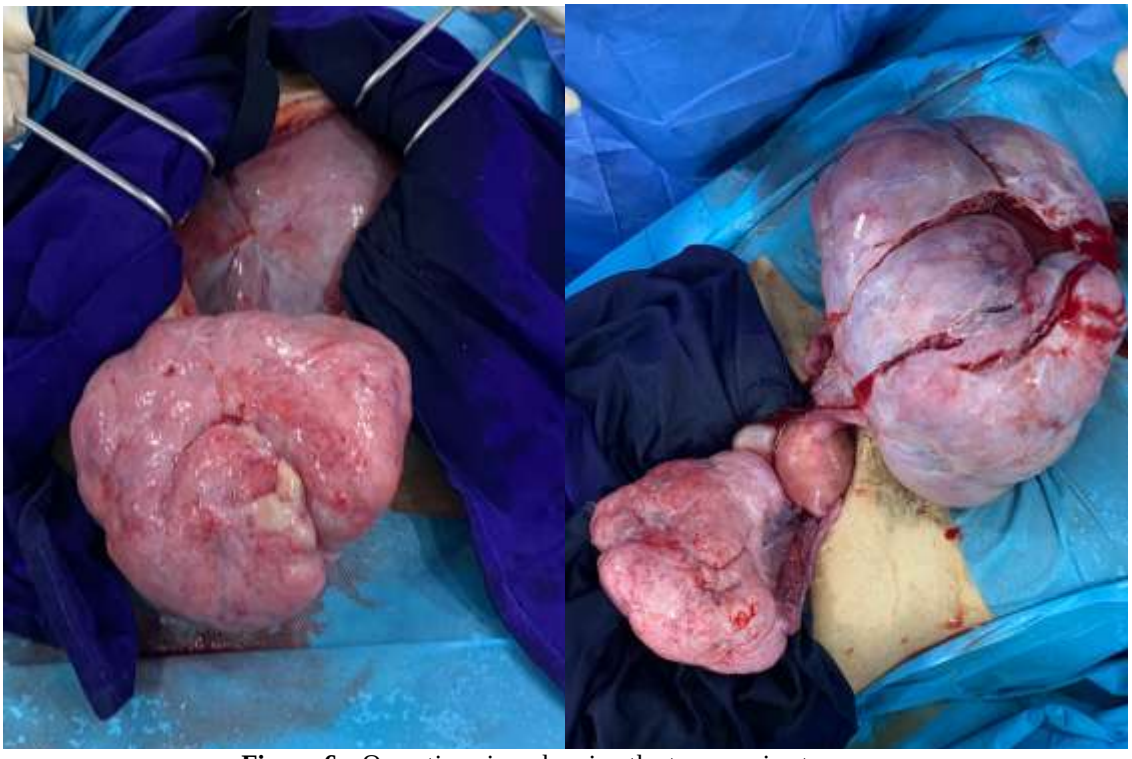


Figure 6:- Operative view showing the two ovarian tumors.

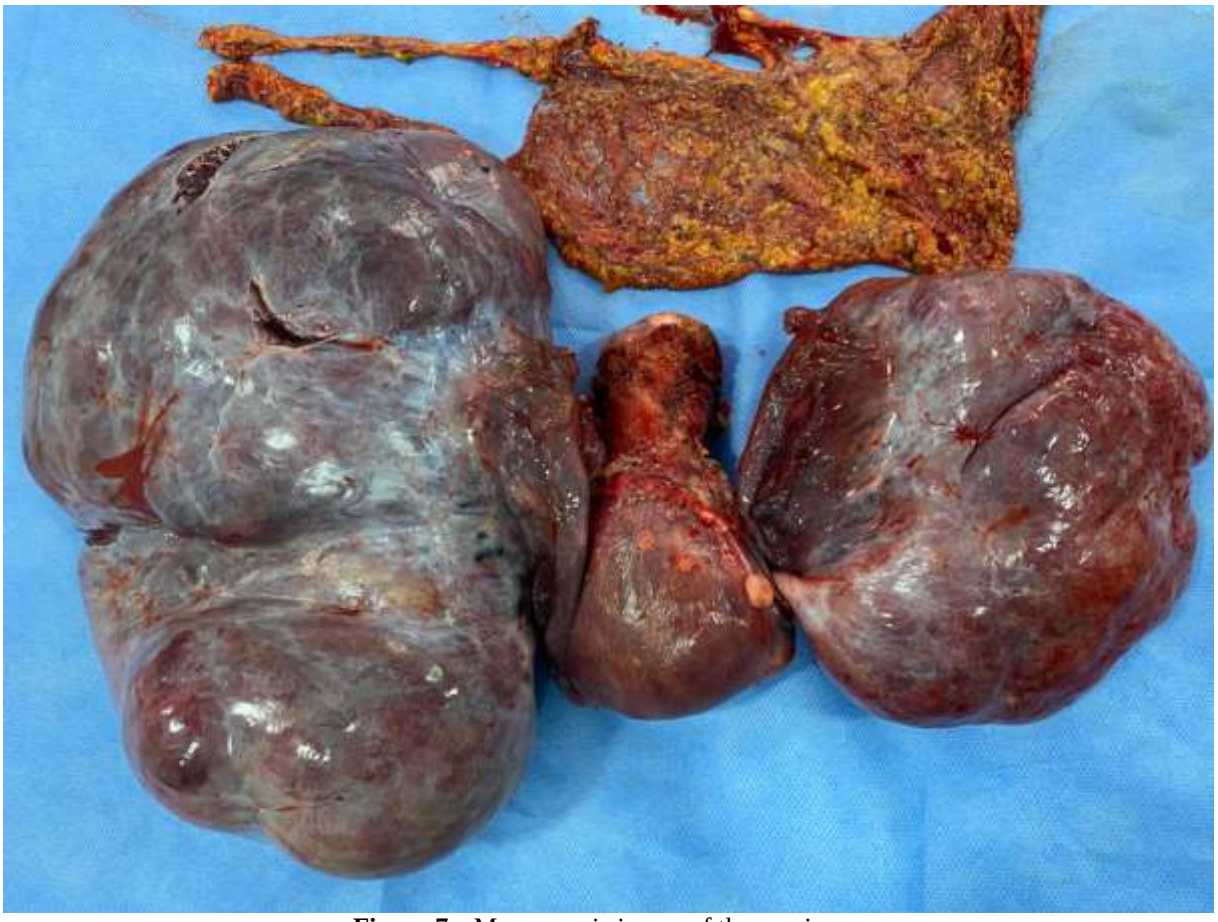


Figure 7:- Macroscopic image of the specimen.

Conclusion:-

Sertoli-Leydig cell tumors of the ovary are rare, especially when they are bilateral. They occur in young women, and often present with signs of virilizing (35-50%). Poorly differentiated forms have a significant malignant potential. Treatment relies on radical surgery combined with chemotherapy.

These germ cell tumors generally have a poor prognosis.

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