

CASE REPORT

BILATERAL OVARIAN NEOPLASM IN ADOLESCENT GIRL: A RARE ENTITY

N. Gopal, Prashant S. Joshi, Vinayachandra Singh.

1. Professor. Department of Obstetrics & Gynaecology, Adichunchanagiri Institute of Medical Sciences,
2. Associate Professor. Department of Obstetrics & Gynaecology, Adichunchanagiri Institute of Medical Sciences,
3. Post Graduate Resident. Department of Obstetrics & Gynaecology, Adichunchanagiri Institute of Medical Sciences,

CORRESPONDING AUTHOR:

Dr. Prashanth S. Joshi,
Flat No. 201, Nanar Sorrento,
36th main, 3rd Cross, BJM I Stage,
Madivala Dollars Colony, Bangalore-560068.
E-mail: joshpj@rediffmail.com.

ABSTRACT: A 20 year old girl presented with mass per abdomen and failure to attain menarche. She had undergone previously left salpingo-oophorectomy surgery for torsion of left ovary, histopathology of which showed dysgerminoma of left ovary. After a thorough investigation, she underwent staging laparotomy followed by hysterectomy with right salpingo-oophorectomy for stage 1 C dysgerminoma

KEY WORDS: Dysgerminoma, Torsion, Salpingo-oophorectomy

INTRODUCTION: Dysgerminomas are the most common malignant germ cell ovarian carcinoma. Most dysgerminomas occur in adolescence and early adult life; 5% occur in pre-pubertal children, and they are extremely rare after 50 years of age¹. Abnormal gonads (due to gonadal dysgenesis and androgen insensitivity syndrome) have a high risk of developing a dysgerminoma. Most dysgerminomas are associated with elevated serum Lactate Dehydrogenase (LDH), which is sometimes used as a tumour marker. Dysgerminomas present as bilateral tumours in 10% of patients and in a further 10% there is microscopic tumour in the other ovary. Dysgerminomas can be located in the brain, usually arising in the hypothalamic or epiphysial regions.

CASE REPORT: A 20 year old presented with failure to attain menarche, Pain abdomen since 2 months, Mass per abdomen since 2 months, Mild dull aching diffuse lower abdominal pain. She gives history of having an abdominal surgery at the age of 13 yrs in a private hospital for acute pain abdomen and vomiting. As per hospital records, she underwent diagnostic laparoscopy followed by laparotomy which showed torsion of left ovarian tumour and she underwent left salpingo-oophorectomy. The histopathology showed dysgerminoma of left ovary.

On examination, she was pale and ill looking with no significant lymphadenopathy, no acne, Breasts-Tanner stage I, nipples not widely spaced & no galactorrhoea, no hirsutism and no webbed neck. External Genitalia examination revealed that pubic hair was Tanner stage I, labia majora and minora appeared normal (Figure1). Vagina admitted one finger with difficulty and Cervix was not felt. Routine blood & Biochemistry investigations were normal. Serum marker was negative for HCG and Positive for LDH. Buccal smear was Positive for Barr body (>18%),

CASE REPORT

Karyotyping was 45X0. Abdominal Ultrasound showed that uterus was compressed due to right sided adnexal mass extending from pelvis to epigastrium. Mass was solid lobulated with cystic components in the centre, showing septations and loculations (Figure2). Bilateral hydronephrosis was present. CT scan revealed a well defined soft-tissue-density mass lesion of 20x12x10 cm occupying predominantly right iliac region with superior extension up to right hypochondriac region medially crossing the midline. Few areas of cystic hypo-dense and necrotic areas noted within the mass (Figure3). There was mass effect on bilateral ureters causing mild bilateral hydronephrosis. There was no evidence of paraaortic, retroperitoneal or pelvic lymphadenopathy, and ascites. Both ovaries could not be visualized. Uterus was hypoplastic 2x3x2 cm. There was no evidence of intraperitoneal/retroperitoneal/solid organ metastasis. Mass appears to displace bowel loops to left abdominal quadrants.

TREATMENT: Staging laparotomy was done followed by TAH with right salpingo-oophorectomy. Per operatively right ovarian mass of 20x15x10cm with bosselated surface with intact capsule was found(Figure4). Uterus appeared infantile. Under surface of the diaphragm, liver, omentum were free of metastatic growth. Para aortic nodes were not enlarged. Peritoneal fluid sample and omental biopsy were taken. Final Diagnosis was Stage I C Dysgerminoma.

On histopathological examination, macroscopic appearance showed Irregular grey white lobulated mass of 20x12x11cm. External surface showed stretched out fallopian tube measuring 11 cm. On Cross Section, grey white lobulated homogenous and yellowish solid solitary cyst with areas of necrosis. Microscopic appearance showed monomorphic cellular tumour composed of round to polygonal cells with large vesicular and prominent nucleoli. Cells arranged in sheets, cords, pseudoalveolar formation and peritheliomatous pattern. Fibrovascular septae were seen separating sheets and groups of cells with lymphocytic infiltration (Figure5). Capsule showed partial thickness infiltration. Mitotic activity was not significant. Peritoneal fluid analysis was positive for malignant cells. Omental biopsy was normal.

DISCUSSION: Dysgerminoma represents the ovarian counterpart of testicular seminoma. These two tumors are histologically identical and the term germinoma has been proposed for both. It usually occurs in normally developed females but is the most frequent ovarian malignant tumour found in dysgenetic females, testicular feminization, hermaphrodites and ambiguous sex. As in our case, Pierga et al. have also described a case of dysgerminoma in a pure 45, XO Turner syndrome².

It is bilateral in 10% of cases. Dysgerminoma complicating pregnancy, though rare, has been reported in 60 cases in literature worldwide. Ultrasound is the first investigation to get the clue. According to Mainz et al, 10 sonographic parameters are assessed and scored on a scale of 0-2, a score of less than 9 is rated as benign; the sensitivity of this scoring is 96.3%. The conventional USG is not diagnostic. CT scan is the most common imaging modality used to stage ovarian tumours; MRI has been shown to be equally accurate⁴. Many germ cell tumours produce biochemical markers like Beta HCG, Alfa Fetoprotein which help in monitoring the response to treatment. Dysgerminoma is commonly devoid of hormonal product.

Small percentage of tumours produces low levels of HCG. LDH is frequently seen in a patient of dysgerminoma. CA-125 level, though elevated, is not specific. FNAC is not recommended due to the risk of dissemination⁵. The sensitivity is only 25%, specificity being 90%. Metastatic spread is mostly via the lymphatic system, does not occur early in the course of

CASE REPORT

disease, and because of that a 5-year survival is 75-90%. Unfavourable prognostic parameters include presence of metastases at the time of diagnosis, presence of adhesions and spread into adjacent structures, presence of bilateral tumour and presence of other neoplastic germ cell elements. Surgical staging is essential for determining the extent of disease.

In Dysgerminoma stage 1A, unilateral salpingo-oophorectomy with proper follow-up is usually sufficient⁶. The 10 year survival rate following conservative surgery is 88.6%. Uneventful pregnancy following conservative surgery is also reported¹. About 15-20% of dysgerminoma recurs but can be successfully treated with combined chemotherapy at the time of recurrence with a high likelihood of cure. This is one of the success 10 years back diagnostic laparotomy was done, with removal of mass, unilateral salpingo-oophorectomy conserving the uterus and opposite ovary, as suggested for the dysgerminoma of stage 1 without any ascites. Then patient was lost for follow up. 10 years later she came back with mass per abdomen. On high index of suspicion and proper investigations a probable diagnosis of dysgerminoma of the contralateral ovary was made. Staging laparotomy with total abdominal hysterectomy with oophorectomy was done. Histopathology report showed DYSGERMINOMA -STAGE 1A. She was again lost for follow up because of financial constraints.

CONCLUSION: The case is interesting because of its heterogenous behaviour.

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CASE REPORT



Figure-1



Figure-2



Figure-3

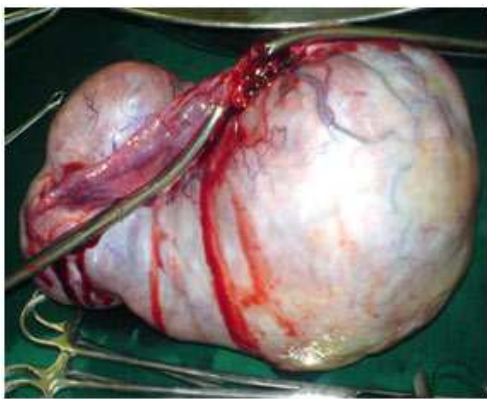


Figure-4

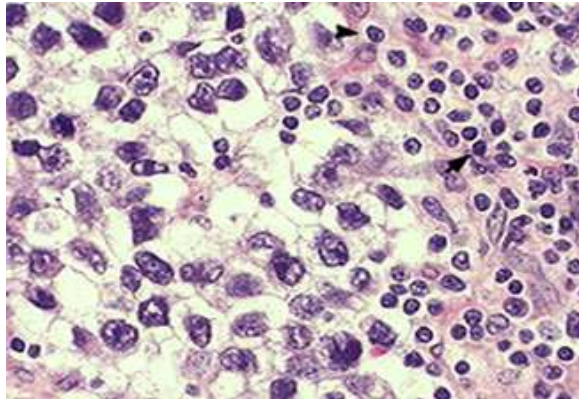


Figure-5