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Case Report

Nongestational primary choriocarcinoma of the ovary: A case report

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Abstract

Nongestational primary choriocarcinoma of the ovary is an extremely rare tumour which occurs usually in the reproductive age group. It is extremely difficult to diagnose as it often mimics torsion ovarian cyst, ectopic pregnancy etc. As choriocarcinoma alone is an extremely rare entity, a thorough search for uterine or tubal origin should be undertaken. We present a case of a 15yr old girl having nongestational primary ovarian choriocarcinoma.

Keywords: Nongestational, primary, ovarian choriocarcinoma

1. Introduction

Pure Primary ovarian choriocarcinoma is a rare entity. It may be either gestational or nongestational in origin. Most are gestational in origin and usually metastasize to ovary from a uterine or tubal choriocarcinoma[1]. Nongestational choriocarcinoma of the ovary usually presents as mixed germ cell tumour. Pure nongestational ovarian choriocarcinoma is rarer & difficult to diagnose and have worse prognosis[3]. We present this case because of its rarity.

2. Case Report

A 15 Yr female who had menarche at 13 yrs was brought to IPGME&R & SSKM Hospital, Kolkata with acute onset pain abdomen and bleeding p/v. She had no history of amenorrhoea/pregnancy. She was suspected to have a twisted ovarian cyst/tumour.
Pre –op workup showed- haemoglobin - 7.2 g/dl, total count – 18,500/cmm, neutrophil - 84%,platelets - 1.2 lakhs/cmm.
Emergency salpingo-oophorectomy was done at the Gynaecology Department and the respected specimen sent to the Pathology Department.
Post-operatively: Her β-hcg– 32,000 miu/ ml CA-125, 102.3 U/mL (<35); CA19-9, 16.6U/mL (<30); alpha fetoprotein (AFP), 12.8 ng/mL (<20); and carcinoembryonic antigen (CEA), 1.5 ng/mL (<5). There was no metastasis to other organs.

Histopathology

Grossly, the ovarian tumour with fallopian tube measured 15x9x6 cm (Fig.1), the cut section was brown in colour with areas of haemorrhage and necrosis. (Fig.2)

Fig.1-Gross specimen of ovarian tumour with fallopian tube.

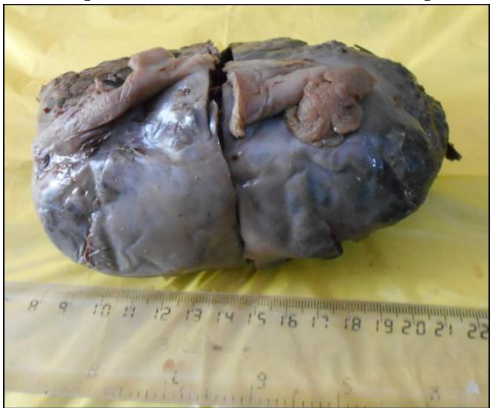
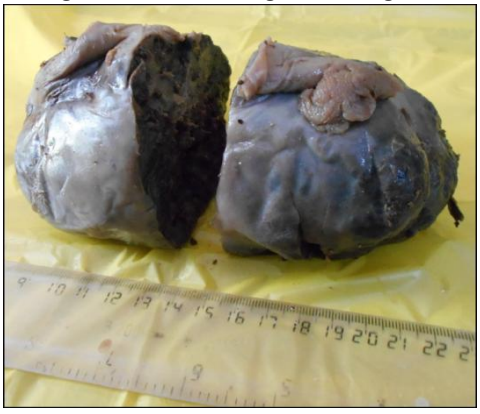


Fig.2 Cut section showing haemorrhage.



Microscopically, a tumour mass was seen with extensive areas of haemorrhage and necrosis. The tumour was composed of complex admixture of syncytiotrophoblasts, cytotrophoblasts and intermediate trophoblasts with absence of villous structure (Figure 3,4,5) At one end normal compressed ovary was seen.(Figure 6)The tube appears to be free of tumour.(Figure 7). The syncytiotrophoblastic cells have multiple smaller nuclei with basophilic cytoplasm. The cytotrophoblast cells have pale cytoplasm and vesicular nuclei with prominent chromatin clumps.

Fig.3 LP view showing extensive haemorrhage and necrosis.

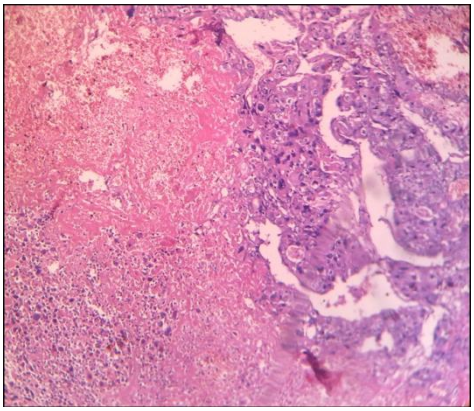


Fig.4 HP view showing cyto & syncytiotroblasts with plexiform arrangement.

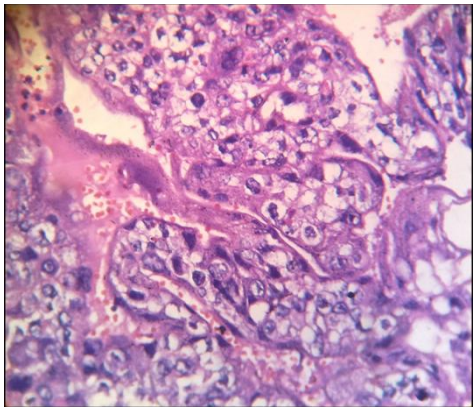


Fig.5 HP view showing cyto &syncytiotroblasts with areas of necrosis with adjacent ovarian stroma

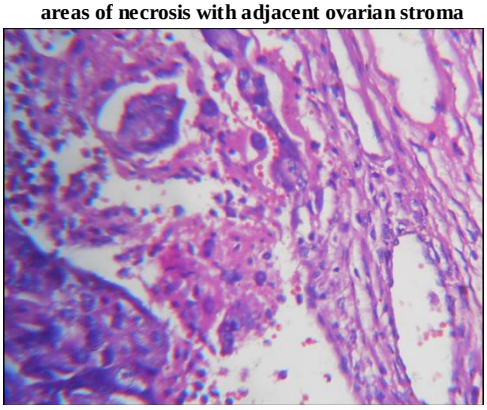


Fig.6 LP view showing ovarian stroma with adjacent tumour.

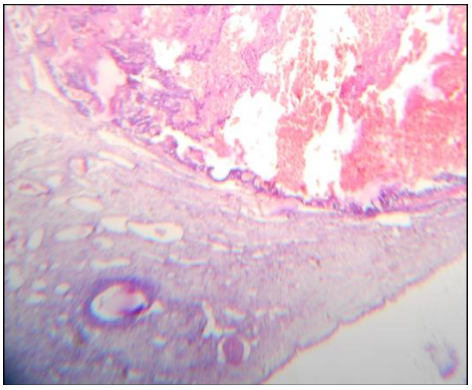


Fig.7 LP view of fallopian tube showing normal histology.

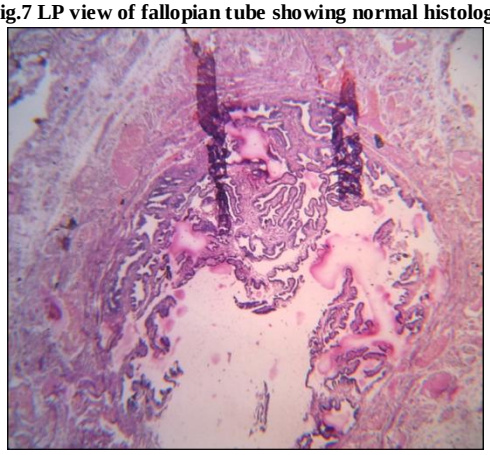
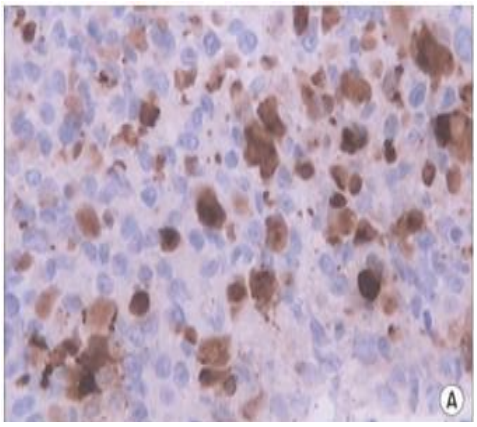


Fig.8 IHC shows β HCG staining taken by syncytiotrophoblasts.



3. Discussion

Pure ovarian choriocarcinoma is rare, with less than 40 cases described[2,5]. Pick (1904) first described a choriocarcinoma in an ovarian teratoma.[6] Choriocarcinoma usually presents as a component of mixed germ cell tumour[7].
Choriocarcinoma is present in 10 to 20 % of mixed germ cell tumours[5,8].

The nongestational type is an extremely rare germ cell neoplasm accounting for ≤0.6% of all ovarian neoplasms. Nongestational type choriocarcinoma usually involves females under 20, with an average age of 13 years (6, 10). Dysgerminoma is the commonest malignant germ cell tumour in this age group.

Saito *et al*, first described the diagnostic criteria for nongestational choriocarcinoma of the ovary in 1963. -Absence of disease in uterine cavity
-Pathological confirmation of the disease
-Exclusion of molar pregnancy & intrauterine pregnancy.[4]

Nongestational choricarcinoma is pathologically indistinguishable from gestational choricarcinoma. Ultrastructurally & Immuno histochemically also, both types are indistinguishable. β-HCG levels are usually lower in nongestational variants compared to gestational types.[5] Nongestational choriocarcinoma of the ovary can be distinguished from gestational choriocarcinoma by DNA polymorphism analysis.[6,7]

Nongestational choriocarcinoma has a poor prognosis & is resistant to Methotrexate.[8] The prognosis correlates with the bulk of tumor& the sites and number of metastases.[9] Monitoring of serum β-HCG can be useful in evaluating response to therapy.

Choriocarcinoma of the ovary is unilateral and treatment is done by salpingo-oopherectomy. Total abdominal hysterectomy with bilateral salpingo-oophorectomy is required only if contralateral ovary or uterus is involved. Surgery is followed by combination chemotherapy with platinum based regimen.

The response to chemotherapy and prognosis for patients with gestational choriocarcinoma differs from that for nongestational choriocarcinoma. So it is very necessary to distinguish whether the choriocarcinoma is of gestational/germ cell type. If the patient is premenarcheal, choriocarcinoma is of germ cell origin. In young and of child bearing age group, gestational choriocarcinoma and germ cell choriocarcinoma are morphologically indistinguishable because of their close similarity to each other. Clinical history of the patient helps in this regard. If corpus luteum of pregnancy is identified, it favours the diagnosis of gestational choriocarcinoma. Presence of other germ cell elements is indicative of germ cell choriocarcinoma. Choriocarcinoma of gestational origin can be established by identification of paternal component with the help of DNA analysis which is a reliable method for identifying the genetic origin of pure ovarian choriocarcinomas [10,13,14,15]. However, since such techniques are always expensive and not generally available in our institution,, the application is limited. For this very reason, we could not perform molecular genetic analysis on the tumor for our patient.

As a follow-up serial beta hcg level monitoring should be done to evaluate the therapeutic response[16].

In our case, the patient was 15 years old young female complained of severe pain abdomen and associated with vaginal bleeding. Emergency left sided salphingo-oophorectomy was done with the provisional diagnosis of left sided twisted ovarian tumour. Postoperative beta –hcg was high 32,000 miu/ ml. Grossly, the ovarian tumour with fallopian tube measured 15x9x6 cm and the cut section was brown in colour with areas of haemorrhage and necrosis with viable tumour cells at the periphery showing syncytiotrophoblasts, cytotrophoblasts in a plexiform manner .The histology of fallopian tube was normal. On multiple sectioning of the specimen, no germ cell elements other than choriocarcinoma were detected. In addition, no history of pregnancy/amenorrhoea was present and no corpus luteum was detected in the ovarian tissue section. So we diagnosed the case as nongestational pure choriocarcinoma .Postoperative chemotherapy was given Bleomycin, Etoposide & Cisplatin. Patient tolerated and responded which was indicated by lowering of serum beta hcg level (20,000miu/ml.

4. Conclusion

Extensive sectioning should be done to rule out a mixed germ cell tumour. It is important to determine the origin of pure extrauterine choriocarcinomas to select the most appropriate treatment.

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