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Case Report

Immature teratoma with a rare finding of choroid plexus

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ABSTRACT

Mature cystic teratoma of the ovary is the most common ovarian tumour in young women. Immature teratoma is a very rare tumour, representing only 1% of all ovarian cancers. Paediatric ovarian tumours are rare, with a reported incidence rate of 2.2/100,000 in girls aged 0 to 15 years. In the case report presented here, was 12-year-old suspected to have serous cystadenoma of ovary based on CT scan, which was ultimately diagnosed with immature teratoma grade 3 after histopathological examination was done.

Keywords: Immature teratoma, Ovary, Choroid plexus

INTRODUCTION

Immature teratoma of the ovary is a rare tumor, representing 1–3% of all germ cell tumors and 20% of malignant ovarian germ cell tumors. It is more common in younger patients.[1] The tumor is composed of tissue derived from the three germ layers-ectoderm, mesoderm and endoderm. Contrary to the much more frequent mature teratoma, it contains immature or embryonal structures.[2] In adults, they are classified under malignant germ cell tumors. The histological grading of immature teratomas is based on the proportion of immature neuroepithelial component. It ranges from 1 to 3. The grade and stage of immature teratomas correlate with prognosis.[3] In the past, surgery alone was the accepted treatment, and despite spontaneous maturation in selected cases, overall prognosis was poor.[4] Higher-grade immature teratomas in adults require adjuvant chemotherapy.[3]

CASE REPORT

A 12-year-old girl presented to the Gynaecology and Obstetrics department with pain abdomen since 2 months. Per abdomen examination revealed mass palpable corresponding upto 22-24 weeks size of uterus. It was mobile, non-tender, cystic mass and had smooth surface.

Pre-operative examination

Hemoglobin-12.8g%, total leucocyte count-4900/cub mm, platelets-2.2 lakhs/cub mm. LDH-230 IU/l, AFP-2.7 ng/ml, beta HCG-0.2 mIU/ml, CA-125-19.7U/ml. On USG abdomen it was found large abdomino-pelvic mass measuring approx. 15.6 x 6.4 cm in size with fine internal septations and compressing upon the uterus and bilateral ureters likely ovarian origin with possibility of cystadenoma. On further evaluation on CECT abdomen-large complex abdomino-pelvic mass with minimally enhancing thin internal septations likely benign ovarian (serous cystadenoma) cyst, however the possibility of mesenteric cyst could not be ruled out. After routine examination, laparotomy followed by right salpingo-oopherectomy with left ovarian wedge biopsy with omental and peritoneal biopsy was done and specimens were sent to histopathology lab.

Pathological examination

Gross-container I labelled as left ovary wedge biopsy-received a cream white soft tissue piece measuring 1.5 x 0.5 x 0.4 cm. Container II labelled as omental biopsy-received fibrofatty tissue measuring 5 x 2 x 0.5 cm. Container III labelled as peritoneal biopsy-received a linear grey brown soft tissue piece measuring 1 cm in length. Container IV labelled as right ovarian cyst-received already cut open specimen of right ovarian cyst altogether measuring 9 x 6 x 3.5 cm. Outer surface was smooth, shining and firm. Inner surface showed multiloculation. Contents had been drained out. No solid areas, necrosis, haemorrhage was seen. Attached fallopian tube measured 4 cm in length. Extensive sectioning was done.

Microscopic examination

Right tubo-ovarian mass showed two germ cell components comprising of multiple cysts lined by mature skin epithelium containing keratinous material, skin based appendageal glands and neuroglial tissue with abundant choroid plexus. Also seen was a single focus of mature cartilaginous tissue. Immature neuroepithelial tissue composed of rosettes and tubules which were lined by overlapping cells having hyperchromatic nuclei with scant cytoplasm, and occupied >3 low power fields in any slide. Adjacent area to neuroepithelium showed stellate cells in myxoid background.

Left ovary wedge biopsy showed follicular cyst and graafian follicles.

Biopsy sent as peritoneal biopsy showed mature adipose tissue fragments along with areas of fibrosis.

Biopsy sent as omental biopsy showed mature fibrofatty tissue with dilated and congested blood vessels and occasional lymphoid follicles. In addition, abundant papillae and nodular proliferation of bland mesothelial cells also seen.

Impression was given as immature teratoma-grade III with large areas of choroid plexus.

Left ovarian wedge biopsy, peritoneum and omentum were free from tumor deposits.

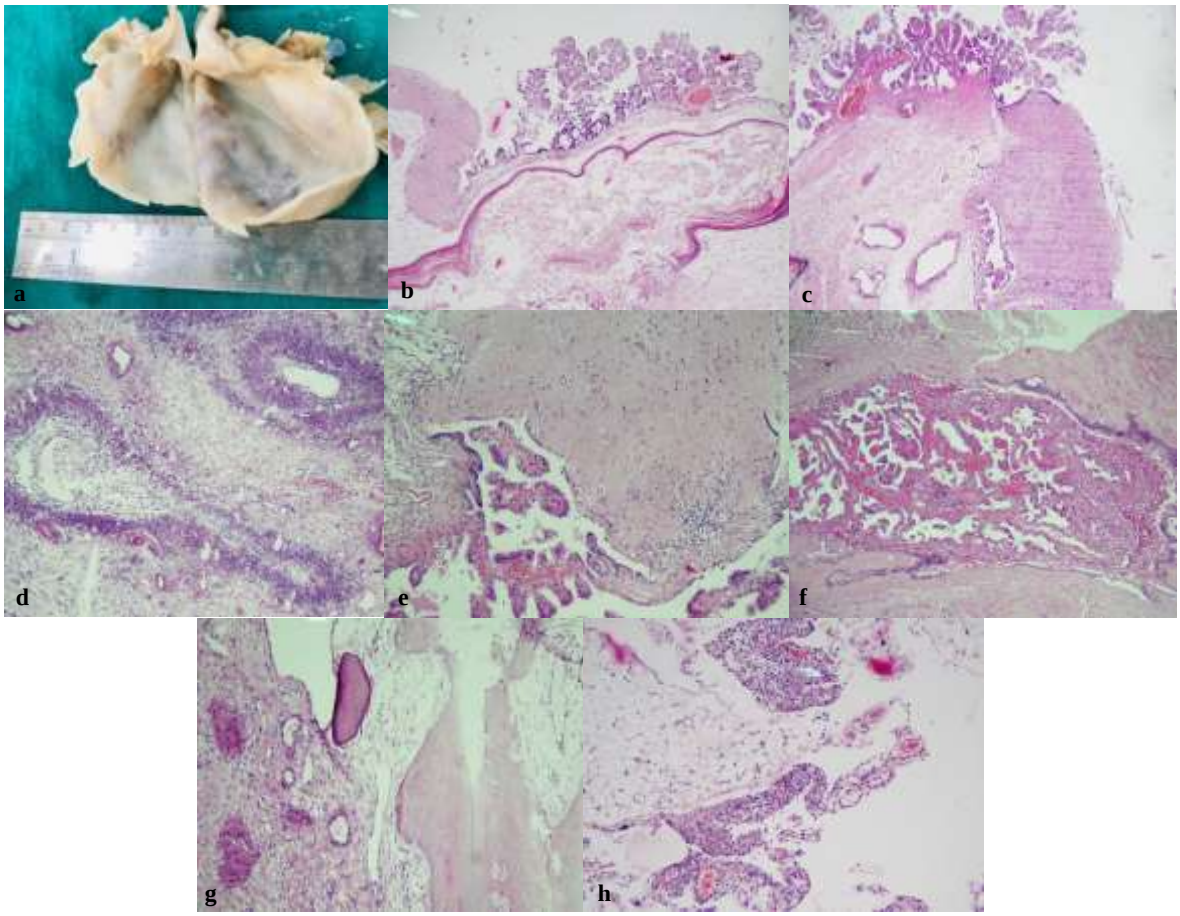

Figure 1: (a) Gross appearance shows multiloculated cysts. (b) Tumor showing glial tissue, stratified keratinized squamous epithelium, keratin flakes and choroid plexus. (c) Tumor showing choroid plexus. (d) Primitive neural tubules and immature rosettes. (e) High power view of choroid plexus and glial tissue. (f) Cyst showing choroid plexus. (g) Foci of mature cartilaginous tissue. (h) Omentum-Papillae and nodular proliferation of bland mesothelial cells.

DISCUSSION

The word 'teratoma' is derived from Greek work 'teraton' meaning monster. Immature teratoma of the ovary is a malignant germ cell tumor containing variable amounts of immature tissue, typically primitive neuroectodermal tissue. Its etiology is usually unknown. Although benign cystic teratomas of the ovary are relatively common tumor that can occur at any age, the incidence of malignant elements in a teratoma is low (~1–2%). The tumor occurs in all ages but usually affects mostly young females.[2] Diagnosis of immature teratoma of the ovary can be challenging especially in small biopsies. Immunohistochemistry has been reported to have limited role in diagnosis of immature teratomas. With resection specimens, extensive sampling may be needed to include areas of immature teratoma at the time during grossing or with frozen section in cases where a preoperative diagnosis is suspected.[2]

Immature teratoma is the only ovarian germ cell neoplasm that is histologically graded.[5] Morphological spectrum of immature teratomas of ovary is varied, complex and offers diagnostic challenges. Presence of immature neuroepithelium is essential to make a diagnosis of immature teratoma (mere presence of immature mesenchyme is not sufficient). However, presence of immature mesenchyme should prompt a careful search for immature neuroepithelium. For grading immature teratomas, primitive neural tubes and immature rosettes are counted.[6]

Choroid plexus is a relatively common component of mature ovarian teratoma; however, it is ill defined in immature teratoma. Immature teratoma mainly composed of choroid plexus is extremely rare.[7] Histological grade in these tumors is considered an important prognostic factor that predicts extra ovarian spread and overall survival.[5] The optimal treatment for immature teratoma in a patient still hoping to bear a child is a unilateral salpingooophorectomy in stage I (as determined macroscopically). Surgery alone is sufficient if the tumor is grade 1 pathologically, while chemotherapy with vincristine, actinomycin D, and cyclophosphamide (VAC or P) is required for a grade 2 or 3 tumor.[8] Combination of surgery and chemotherapy can give longer survival even in recurrent disease.[6]

CONCLUSION

In our case, tumor showed large amount of immature neuroepithelial tissue occupying >3 low power fields (40X) i.e., grade 3 (high-grade) accompanied by large areas of choroid plexus. Thus, our case suggested that extensive choroid plexus proliferation can be seen in immature teratomas.

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