A Rare Case of Primary Neuroectodermal Tumor of the Ovary-Ependymoma

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Summary

Primary ependymoma of the ovary is a rare entity, very few cases of which have been described in the literature. There is still no uniform consensus on the management of these cases. Here we are presenting a case of an 18-year-old girl, who came with a history of abdominal distension for 2 months and loss of appetite. Imaging revealed the presence of a large adnexal mass. She underwent fertility sparing surgery with the removal of the adnexal mass with surgical staging. On gross examination, there was a well encapsulated cystic mass with papillary projections. On microscopic examination, perivascular pseudo rosettes were seen. On immunohistochemistry, GFAP was positive. Hence a final diagnosis of ependymoma was made. She received postoperative adjuvant chemotherapy withbleomycin,etoposide and cisplatin. The patient is alive and her menstrual cycles have resumed. Despite being a rare entity, ependymoma should always be included in the differential diagnosis of adnexal masses in young women.

Keywords: Primaryneuroectodermaltumor, primary ependymoma, ovarian ependymoma

Introduction

Ovarian ependymoma is an extremely rare gynaecological malignancy, classified under the category of neuroectodermal tumors of the ovary.Given its extreme rarity, it poses numerous diagnostic and treatment challenges. The clinical behaviour, molecular profile, and optimal therapy are incompletely characterised.[1]

Ependymomas, usually arise from the central nervous system and are classified under gliomas. It can easily be misdiagnosed as a case of mature cystic teratoma. Although histogenesis has not been fully elucidated, they are thought to represent monodermal teratomas of neural type.[2] Kleinman was the first person to describe a case of ovarian ependymoma in 1984.[2] Since then, 36 cases have been reported in the literature, two of which have been described during pregnancy.

Here we describe a case of an18-year-old girl with ovarian ependymoma diagnosed and managed at our institute, GCRI (Gujarat Cancer & Research Institute)

Case Report

An 18-year-old, unmarried girl was referred witha CT scan report showing a right adnexal mass of 10x16x17cm.

She complained of abdominal distension for 2 months and loss of appetite and indigestion for 15days.She had attained menarche at the age of 14 years and her menstrual cycles were regular since then with no history of excessive bleeding/scanty menses. She had no urinary or bowel disturbances. Her medical and family history was unremarkable. On clinical examination, there was a 26-28weeks size mobile cystic mass, that felt separate from the uterus.

An outside CT scan was reviewed at our centre, which revealed a well-defined multiloculated cystic lesion of 10x16.8x17.3cm arising from the right adnexa. The lesion showed heterogeneously enhancing solid components within. The right ovary was not seen separately from the lesion. Minimal free fluid present. The left kidney could not be visualised, which was an incidental finding (Figure 1). An ultrasound done at our institute showed the same findings and characteristics of the lesion.

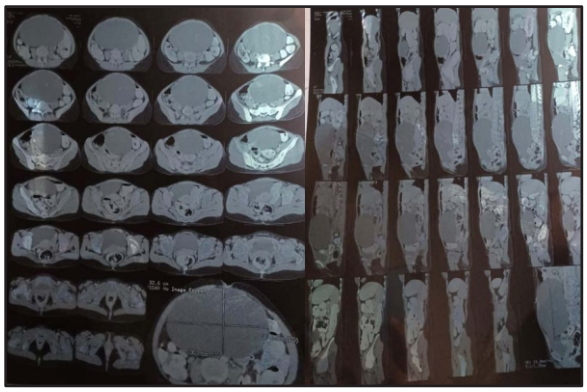

Figure 1: CT scan images showing a large predominantly cystic adnexal mass occupying the whole pelvis in axial and sagittal sections

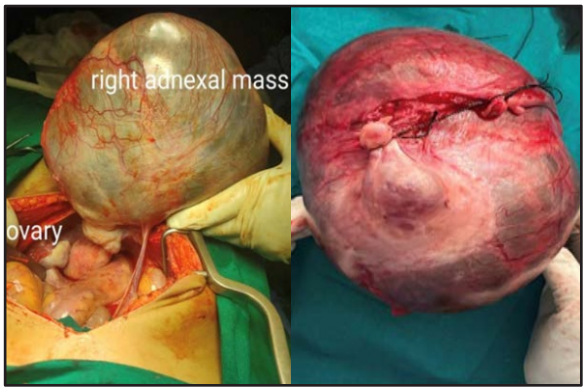

Figure 2: Intraoperative finding: Cystic adnexal mass of 17x14cm with few papillary projections

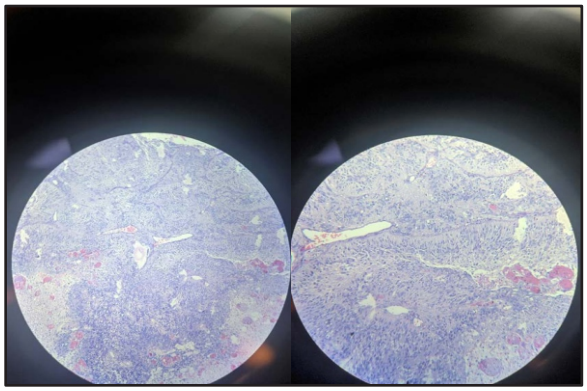

Figure 3: Microscopy findings: Perivascular pseudorosettes

Her tumor markers were as normal(CA-125=24 U/ml, HE-4= 52pmol/L, CEA- 1.8 ng/ml, alpha-fetoprotein= 1.68ng/ml, beta HCG<0.2 IU/ml, LDH=176 U/L).

She underwent a staging laparotomy in June 2022.Intraoperatively a large cystic mass of 10x12cm was noted in the right adnexa. Peritoneal washings were obtained. The mass was removed and sent for a frozen section. The opposite ovary and uterus were inspected for disease. On exploration, there was no evidence of disease in the abdomen and pelvic cavity, under surface of the diaphragm, omentum and mesentery. The frozen report turned out to be a benign ovarian lesion with a probable diagnosis of cystic teratoma. There was no evidence of disease apart from the right ovary. Fertility sparing surgery was planned for her. She was treated with unilateral salpingo-oophorectomy and a large omental biopsy and peritoneal washings were taken. The final histopathology report was ependymoma of the ovary

Gross Examination

There was a well-encapsulated right adnexal mass measuring 17x13.5x10cm. The external surface of the tumor showed nodularity and papillary projections. The largest papillary projection measured about 4x4x2cm. Ovarian surface involvement was present. The omentum was free of tumor (Figure 2). On the cut section, it was solid, friable, soft, and dull grey in colour, with areas of haemorrhage and necrosis. The peritoneal cytology was negative for malignant cells.

Microscopy

Perivascular pseudo rosettes, characteristically described for ependymomas were seen. These pseudorosettes were formed by tumor cells surrounding blood vessels like the"spokes of a wheel". Ependymal rosettes comprising tumor cells encircling a lumen were also seen. There was no evidence of lympho vascular space invasion (Figure 3).

Immunohistochemistry (IHC)

IHC staining by the immunoperoxidase method showed strong positive staining for glial fibrillary acidic protein(GFAP)
The detailed report was as follows:
AE1-Occasional focal positive
EMA- dot like positive
GFAP-positive
S100-positive
Inhibin- occasional cell positive
Synaptophysin-negative
AFP-equivocal
OCT ¾- negative
CK7-few cells focal positive
M1B1- 8-10%

The patient was diagnosed to have Stage 1c ovarian ependymoma. The patient received 3 cycles of adjuvant therapy with Bleomycin, Etoposide and Cisplatin, the last cycle concluded in November 2022. She has been advised regular follow up.

Discussion

Extra-cranial ependymomas have been described in the extraspinal, and sacrococcygeal regions, in pre-sacral tissues and the sacrum as well. Other locations include the ovary, Para ovarian tissues, broad ligament, omentum, mediastinum and the lung. The World Health Organization's (WHO) Histological Classification of Ovarian Tumor has classified ovarian ependymomasunder ''Neuroectodermal Tumors", which are monodermal teratomas. Monodermal teratomas are defined as germ cell tumors derived from a single germ layer. It may comprise neuroectodermal, vascular, sebaceous and mucinous tissues.[2]

Primary neuroectodermal tumors of the ovary are further classified under 3 groups: (1) Differentiated group: ependymoma, astrocytoma, and oligodendroglioma; (2) Primitive tumors: neuroectodermal tumors (PNETs), neuroblastoma, ependymoblastoma, medulloblastoma, and medulloepithelioma; (3) Anaplastic group: glioblastoma multiforme.[2]

The differentiated type– ependymoma of the ovary usually presents at a younger age but can occur in a wide age range (6–69 years). It usually presents with unilateral mass and without the extra-ovarian disease.Usually arising from the central nervous system, ependymoma is a glioma with differentiation toward ependymal cells. These cells are located in the spinal canal or the wall of the ventricles. In very rare cases, they arise from an ovarian teratoma.[2]

Ovarian ependymomas may often cause diagnostic dilemmas because of highly variable histology and hence mimicking other common conditions. The histological picture may harbour papillary areas with psammoma bodies, pseudo follicles, trabeculae, and microcysts, and may mimic other ovarian tumors such as struma ovarii, granulosa cell, Sertoli-Leydig cell, serous, and Wolffian tumors. Ependymomas can be falsely reported as mature teratomas as occasionally glial and ependymal elements have been observed in them.[3]

Many surface epithelial and stromal tumors, specifically serous and endometrioid borderline tumors and carcinomas, sex cord-stromal tumors, including Sertoli-Leydig cell tumors and granulosa cell tumors and certain tumors of Wolffian origin can resemble ependymomas and lead to diagnostic errors. The combination of long fibrillary cytoplasmic processes, the perivascular rosettes, and GFAP-immunopositivity helps the pathologist in clinching the diagnosis of ependymoma.[3]

IHC plays a very important role in the identification of ependymoma of the ovary. The classical marker is GFAP. In our case, GFAP and S100 were positive and EMA was dot positive which helped in the diagnosis. Hence the final histopathology in our case was reported as ependymoma.

The treatment of malignant germ cell tumors serves as a guide for the management of ovarian ependymomas as very few cases have been reported in literature. The treatment consists of surgical debulking followed by adjuvant chemotherapy. In cases of fertility preservation, surgical debulking includes removal of the adnexal mass, peritoneal washings, omentectomy, exploration and excision of all the disease while preserving the opposite ovary and the uterus. In those women who do not desire future fertility, surgical debulking involves total abdominal hysterectomy, bilateral salpingo-oophorectomy, comprehensive staging and removal of all the macroscopic disease.

In the cases reported so far in the literature, the traditional first-line chemotherapy regimen consisting of bleomycin, etoposide and cisplatin(BEP) for germ cell tumors has also been proven to be effective for ovarian ependymomas. Hinoe et al. have suggested an effective second-line therapy incorporating paclitaxel, ifosfamide, and cisplatin therapy in cases resistant to BEP therapy.[4] Our patient received BEP as adjuvant therapy.

A case reported by Takano et al illustrated that even in Stage 3c ovarian ependymoma, fertility sparing approach followed by adjuvant chemotherapy can be employed for the successful management of these tumors. After 16 months of follow up, the patient reported regular menstrual cycles and no evidence of recurrence during follow up in their study.[5]

Sevil Sayhan et al also reported a study in which a fertility sparing surgery was done on a 33-year-old woman diagnosed with ependymoma on a background of monodermal ovarian teratoma followed by adjuvant chemotherapy. The patient was alive with no signs of recurrence on 3 years follow up.[6] This case also illustrated the diagnosis and management of ependymoma in a young woman of reproductive age group utilising a fertility sparing approach in early-stage cases.

Other options such as hormone therapy including aromatase inhibitors and the use of GnRH analogues are also being explored as alternatives to chemotherapy in these tumors.

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