Case Report

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Pathology Section

Pilocytic Astrocytoma Arising in Ovarian Mature Cystic Teratoma: An Unusual Neuroectodermal Malignant Transformation Occurring in Pregnant Female

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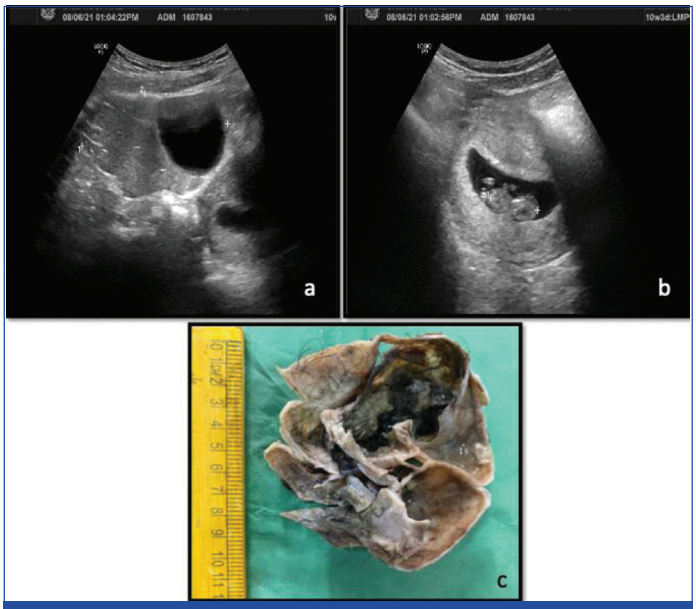
ABSTRACT

Mature cystic teratomas are usually benign, however malignant transformation occurs rarely. It is identified in only 0.17-02% of cases. Malignant cells can arise from cell of any type, but most common malignant transformation found to be squamous cell carcinoma (80% cases) followed by adenocarcinoma. While neural tissue is identified in nearly 80% cases of mature cystic teratomas, neuroectodermal tissue with malignant transformation is the most exceptional event. There is minimal data available on primary neuroectodermal tumours of ovary, majority are astrocytoma of different grades, only few cases were reported in the literature so far. Authors report a extremely rare and unique case of pilocytic astrocytoma arising from a mature cystic teratoma in a pregnant female. A 33-year-old pregnant female in her first trimester came for regular antenatal visit, Ultrasound (USG) abdomen and pelvis single live intrauterine gestation corresponding to 10 weeks and incidental complex right ovarian cyst likely to be mature cystic teratoma. Patient underwent right ovarian cystectomy five months after normal vaginal delivery and provisional diagnosis was given as complex right ovarian dermoid cyst. Final detailed histopathological examination revealed a tumour with glial tissue within the cyst, reported as pilocytic astrocytoma {World Health Organisation (WHO) grade 1} arising in mature cystic teratoma. To the best of authors' knowledge this is the third case showing pilocytic astrocytoma component in ovarian teratoma and the first case of this entity occurring in a pregnant female. Authors present an unusual case where radiology gave preliminary diagnosis however, extensive histopathological examination, histochemistry and immunohistochemistry helped in definite diagnosis.

Keywords: Dermoid cyst, Glioma, Mature teratoma, Rosenthal fibres

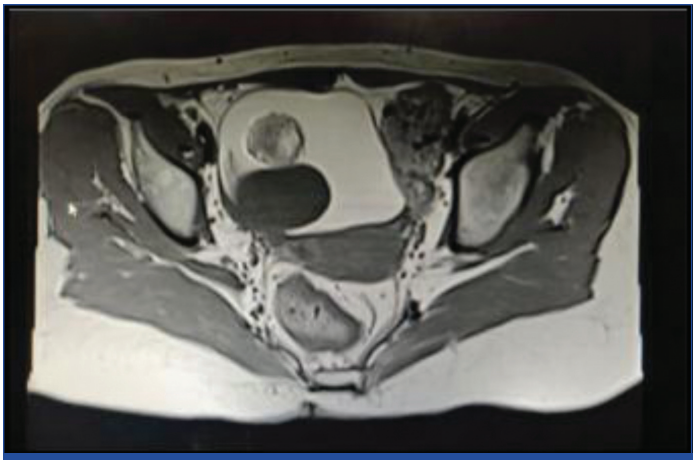
CASE REPORT

A 33-year-old pregnant female in her first trimester of second pregnancy, came to Obstetric Outpatient Department for regular antenatal check-up without any significant complaints. Ultrasound (USG) abdomen and pelvis revealed single live intrauterine gestation corresponding to 10 weeks and complex right adnexal cyst of size 6.5×5.8×6.0 cm [Table/Fig-1a,b]. She was advised Nuchal Translucency (NT) scan and follow-up at 11 to 13 weeks, however she revisited the Institute OPD for cystectomy only five months after normal vaginal delivery which was conducted outside. She was P2L2 with regular previous menstrual cycles, married four years back and last child birth was five months. No other associated co-morbidities present. On abdominal palpation, separate, non tender, mobile right-sided cystic mass was found, compatible with ovarian cyst, was sent to radiology department for further evaluation. Baseline investigations such as complete blood count, routine urin e examination were done and found to be within normal limits. Serum tumour markers were analysed, Cancer Antigen (CA)125 was 12.6 U/mL, Alfa fetoprotein was 3.10 ng/mL, and beta human Chorionic Gonadotropin (HCG) was not detectable. USG abdomen and pelvis showed a complex right adnexal cyst of size 7.8×6.7×7.2 cm with fluid fat level and calcific focus. Magnetic Resonance Imaging (MRI) revealed large cyst with heterogenous fluid and fat signaling complex ovarian cyst of size 7.9×7.9×6.2 cm and the impression was given as likey to be mature cystic teratoma [Table/Fig-2].



[Table/Fig-1]: a) Ultrasound abdomen and pelvis shows right adnexal cyst with fluid fat level and calcific focus; b) Along with single viable intrauterine pregnancy; c) Gross picture of multiloculated ovarian cyst with visible hair follicles admixed with fat and pultaceous material with few solid areas (c).

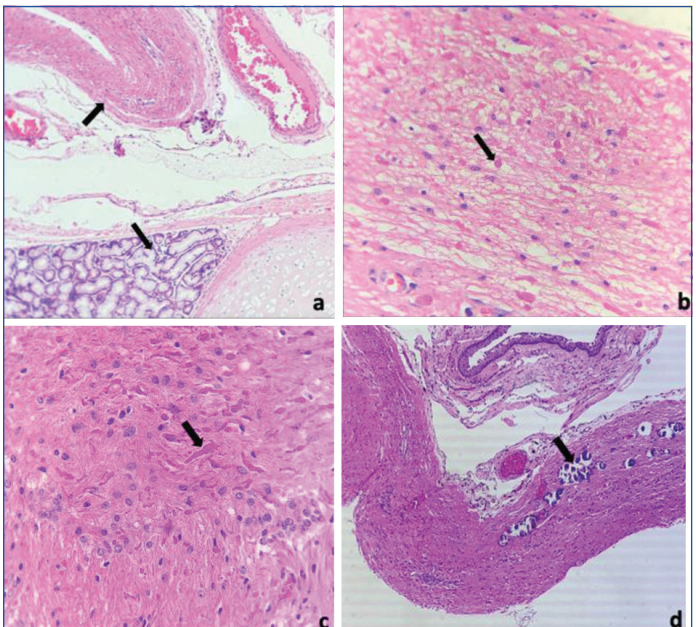
Patient underwent exploratory laparotomy and right ovarian cystectomy demonstrating 8×8 cm large cyst replacing the right ovary along with bilateral tubectomy. Intraoperatively, right and left fallopian tubes, uterus and left ovary were unremarkable. Right ovarian cyst and bilateral tubectomy specimens were submitted for histopathology. Provisional diagnosis of dermoid cyst was made based on MRI impression and intraoperative findings. On gross examination, already cut opened cyst measured 9×6×2 cm completely replacing ovary, along with seperately lying bilateral tubes. On extending the cut, cyst drained clear fluid. Cut surface showed multiloculated cyst with visible hair follicles admixed with fat and pultaceous material, maximum cyst wall thickness was 1 cm



[Table/Fig-2]: MRI abdomen and pelvis image revealed heterogenous fluid and fat signaling complex large ovarian cyst of size 7.9×7.9×6.2 cm.

with few solid areas [Table/Fig-1c]. Few cystic areas were filled with thick mucoid material. Specimen was sampled adequately.

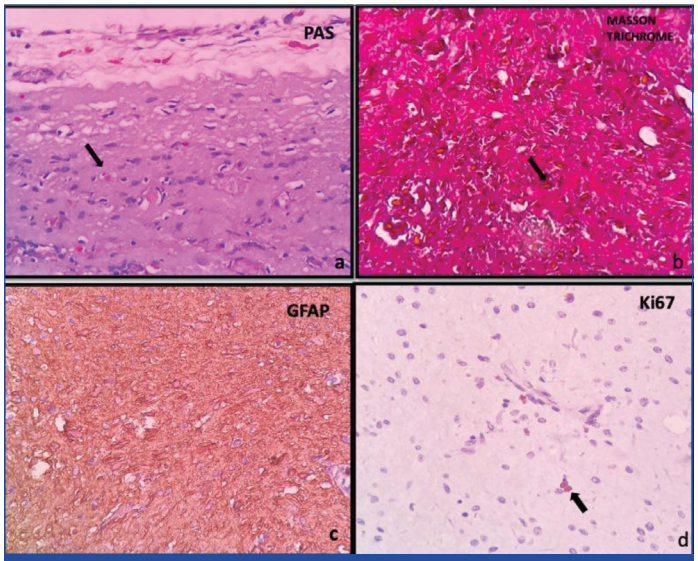
Histologically cyst wall with mature derivatives of all three germ cell layers namely ectoderm, endoderm and mesoderm were identified. Stratified squamous epithelium with underlying good number of sebaceous glands, mature cartilage, foci of respiratory and transitional epithelium were identified. Areas of sharply demarcated glial tissue with biphasic morphology noted, predominantly composed of loose microcystic area with eosinophilic granular bodies and compact areas with plenty of bright-red sausage or corkscrew-shaped Rosenthal fibres and astrocytes with elongated nuclei were noted. Degenerative change in the form of psammomatous as well as patchy type calcification appreciated at places [Table/Fig-3a-d]. There was no evidence of necrosis, endothelial proliferation or increase in mitotic activity. Final impression was released as mature cystic teratoma with pilocytic astrocytoma component- World Health Organisation (WHO) grade 1 [1].



[Table/Fig-3]: Mature teratoma with Pilocytic astrocytoma histology: a) Glial tissue (top arrow) mature sebaceous glands (arrow below) and cartilage (H&E stain, 10X); b) Loose microcystic areas with eosinophilic granular bodies (H&E stain, 40X); c) Compact areas with Rosenthal fibres (arrow) (H&E stain, 40X); d) Degenerative changes as patchy calcification and psammoma bodies (H&E stain, 10X).

Periodic acid schiff histochemical stain highlighted eosinophilic granular bodies and Masson trichrome stain demonstrated bright Rosenthal fibers [Table/Fig-4a,b]. Immunohistochemical stain Glial Fibrillary Acidic Protein (GFAP) also performed to confirm glial origin and to highlight the Rosenthal fibres, came out to be strong and diffuse positivity. Ki-67 stain showed low proliferation index of 1 to 2% compatible with low grade (grade 1) tumour [Table/Fig-4c,d].

The postoperative period was uneventful. On follow-up visits till 11 months of postoperative period, patient was asymptomatic without any complaints. Written consent for publication was obtained from the patient.



[Table/Fig-4]: a) Periodic Acid Schiff (PAS) stain showing darkly stained eosinophilic granular bodies (arrow) (PAS stain10X); b) Masson trichrome stain highlights Rosenthal fibers, (MT stain, 40X); c) GFAP IHC stain is strong and diffusely positive in the tumour cells (40X); d) Ki67 IHC stain showing occasional cell positivity (arrow), 1 to 2% score (40X).

DISCUSSION

Benign (mature) cystic teratomas are one of the most common ovarian neoplasms occurring in women of reproductive age group, accounts for 20% of all ovarian neoplasm and 95% of all ovarian germ cell tumours [2]. Peak incidence is 20-29 years. Mature teratomas/dermoid cyst, a germ cell tumour contains well differentiated tissue of all three layers of germ cells, including ectoderm, endoderm, and mesoderm [3,4]. Mature cystic teratomas are usually benign, but malignant transformation identified in 0.17-2% of cases [5]. Malignant cells can arise from cell of any type, but most common malignant transformation is squamous cell carcinoma (80% cases) followed by adenocarcinoma [6].

While neural tissue is identified in nearly 80% cases of mature cystic teratomas [7], neuroectodermal tissue with malignant transformation is very rare, however neuroectodermal tumours including glioblastomas, oligodendrogliomas, ependymomas, and astrocytomas were reported previously [8]. In the detailed literature search, only 44 cases of gliomas including the present case have been described, out of these cases 37 cases were found within a mature cystic teratoma and seven developed from immature type. Four cases diagnosed as fibrillary astrocytomas WHO grade ranging from I–II, 16 cases were ependymomas and 13 cases found to be grade IV glioma [9]. Oigodendroglial, ependymal tissues, ganglia of sympathetic type, and Schwann cells may be identified in mature cystic teratomas. Pilocytic astrocytoma of central nervous system is the most common childhood tumour occurring in cerebellum, classified as grade-I tumour by World Health Organisation. This should be differentiated from other types such as infiltrating or diffuse type of astrocytoma since these types tend to infiltrate adjacent tissues. To the best of our knowledge this is the third case showing Pilocytic astrocytoma component, first case reported as pure pilocytic astrocytoma [10] and the second case reported as collision glial neoplasms composed of pilocytic and ependymal components [9].

Due to routine antenatal USG examination, incidental diagnosis of adnexal masses are on rise. Though there is spontaneous resolution of few masses, some cases end up in complications like torsion, interference in normal labour or rupture resulting in surgical emergency, which adversely affects maternofetal outcome [11].

Late detection may increase the risk of malignant transformation, however the risk of occurrence of malignancy in pregnancy ranges from 1 in 10000 to 1 in 50000 [12]. The most common ovarian tumour occurring in pregnancy was mature teratoma followed by serous cyst, bilateral borderline serous carcinoma was identified in one patient [13]. The present case is the first unique case of pilocytic astrocytoma originating from a mature cystic teratoma occurring in a pregnant female. The treatment modes and prognosis are notwell prevalent for these tumours due to rarity.

While the literature knowledge is limited about the behaviour of low-grade astrocytoma arising in a mature cystic teratoma, the patients described in the previous studies were cured with surgical resection itself [8]. As far as gynaecological tract's astrocytomas are considered, authors believe that patients with low grade glial tumours undergoing complete surgical resection will have good prognosis. Szczepa Ska M et al., studied about ovarian lesions occurring in pregnant females 35% of cases were surgicly intervened to excise the tumour and 65% of the cases were managed by non surgical conservative modality [13]. Gliomas of all types arising within mature or immature teratomas of the ovary tend to present as ectopic site lesions such as in peritoneal cavity and sometimes in the lymph nodes [14]. This theory appears not suitable for this case.

The identified risk factors involving malignant transformation of mature cystic teratoma are larger size tumour, older age, postmenopausal status and elevated CA-125 levels [15]. The exact mechanism is not understood, however it was suggested that presence of mature cystic teratomas without excision for longer period may be related to malignant transformation. Teratomas presenting in postmenopausal and older women may present for several years before excision. This long-term presence can be related with overexposure to carcinogens or causing increased mutations over a period of years [16]. Interestingly, in the present case report the patient presented with larger size tumour which is the only known risk factor of malignant transformation.

CONCLUSION(S)

This case report presented extremely rare entity of pilocytic astrocytoma originating from ovarian mature cystic teratoma occurring in a pregnant female. Where radiology gave preliminary diagnosis however, extensive histopathological examination and immunohistochemistry helped in definite diagnosis, emphasising the need of appropriate sampling to identify rare components with proper grading and subtyping. This case report will be contributing to the minimal information obtainable for primary pilocytic astrocytoma of female genial tract showing malignant ectodermal transformation in the background of mature cystic teratoma. Many more case studies and follow-up details are surely needed for helping physicians to plan for better treatment and to understand the prognosis of these rare entities.

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