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Case Report

A Case of Immature Teratoma in a Pregnant Female

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Abstract

Neuroectodermal tumors are rare lesions that arise from cells of the primitive neuroectoderm located in the neural crest. Their occurrence in pregnancy is extremely rare and only a couple of case reports have been reported.

This case report looks at a 20-year-old pregnant female who presented at estimated gestational age of 22 weeks and 4 days with rapidly increasing abdominal girth and was found to have ascites and an ovarian mass. Pathology confirmed the diagnosis and patient underwent surgical resection with adjuvant chemotherapy.

Keywords: Ovary; Immature teratoma; Pregnancy; Chemotherapy

Abbreviations

WHO: World Health Organization; EGA: Estimated Gestational Age; CBC: Complete Blood Count; MRI: Magnetic Resonance Imaging; FIGO: International Federation of Gynecology and Obstetrics; POD: Post-Operative Day; CT: Computerized Tomography; BEP: Bleomycin Etoposide Platinum; NSE: Neuron Specific Enolase; GFAP: Glial Fibrillary Acidic Pattern; EMA: Epithelial Membrane Antigen; GP: Gliomatosis Peritonei; PNETs: Primitive Neuroectodermal Tumors; CAV/IE: Cyclophosphamide Adriamycin Vincristine Ifosphamide Etoposide

Introduction

Immature teratomas of the ovary represent 3% of all teratomas, 1% of all ovarian cancers and 20% of malignant ovarian germ cell tumors. It mostly affects women in the first 2 decades of life. According to the World Health Organization (WHO), immature teratoma is defined as a teratoma containing a variable amount of immature embryonal type (generally) neuroectodermal tissue [1]. Immature teratomas that contain large amounts of immature neural tissue and those that have ruptured or disseminated through the abdominal cavity are treated as malignant [2]. Kleinmann *et al.* (Am. J. Surg. Pathol. 1993; 17: 764-778), demonstrated that tumors composed exclusively of immature neuroectodermal tissue should be separated from other teratomas and treated as a distinct group of neoplasms [3].

Herein, we report a case of neuroectodermal ovarian teratoma that occurred in a pregnant subject. To our knowledge, there have only been a few isolated case reports on this topic, including that of a primitive neuroectodermal ovarian tumor and a neuroectodermal renal tumor; both of which were diagnosed in subjects who were pregnant at the time.

Case Summary

20-year-old female gravida 1 presented at 22 weeks and 4 days Estimated Gestational Age (EGA) with complaints of a 2-week history of abdominal pain and swelling. She reported that her abdominal size had greatly increased over the preceding weeks in a way which she felt was disproportionate to the progression of her pregnancy. She reported appropriate fetal movement throughout this period, and was seen for routine prenatal care at another facility where she was told there was no need for concern. The subject stated that an ultrasound exam of her abdomen and pelvis performed at that facility revealed no unusual findings.

On examination at our institution, vital signs were normal except for mildly elevated blood pressure of 138/83. She had a distended abdomen that appeared tense and nearly rigid, consistent with massive ascites. Fundal height was difficult to palpate due to the tense nature of the abdominal wall. Fetal heart tracing was normal with heart rate in the 150's. No contractions were noted on external tocometer.

A Complete Blood Count (CBC) and complete metabolic panel were all normal. Lactate dehydrogenase was 225mmol/L, alpha feto-protein 132.7ng/mL and beta human chorionic gonadotropin 117.0mIU/mL. A urine protein to creatinine ratio was 0.039. Abdominal ultrasound showed massive ascites with an intrauterine pregnancy of 21 weeks and 5 days gestation. She was initially thought be having nephrotic syndrome in the setting of decreased urine output, proteinuria, hypoalbuminemia and edema. A repeat ultrasound exam showed a left complex left sided pelvic mass, 9cm by 13cm by 7cm, consistent with an ovarian tumor. A Magnetic Resonance Imaging (MRI) of the abdomen/pelvis further defined the mass as measuring 12cm by 10cm by 9cm. Mass was centered within the cul-de-sac which appeared to be closely associated with the left adnexa. It demonstrated a cauliflower appearance and was isodense on T1 and T2 imaging. It also demonstrated restriction on diffusion weighted imaging as well as enhancement with gadolinium.

After extensive counseling, the subject agreed to performance of an exploratory laparotomy, with possible left salphingo-oophrectomy and surgical staging. Approximately 12 liters of ascitic fluid was aspirated intraoperatively. The left ovary appeared grossly abnormal, and was removed surgically, along with the left fallopian tube. Pathologic exam *via* rapid frozen section was performed which showed a malignant tumor, favoring ovarian teratoma. On immunostains, tumor was reactive for synaptophysin, neuron specific enolase NSE, S-100 protein, neurofilament and Glial Fibrillary Acidic Protein (GFAP). The final pathologic diagnosis confirmed the presence of a malignant teratoma (neuroectodermal monodermal teratoma) that

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measured 12.9 by 10.2 by 9.8cm. The tumor showed regional necrosis, neurophil formation and extensive immature neuroectodermal tubular structures resulting in primitive neural tubes. The stroma adjacent to the tumor showed stromal thecal luteinization. The ovary showed focal cystic follicles with the tumor occupying the interior of the ovary. The ovarian surface was ruptured but the fallopian tube was uninvolved. An omental biopsy showed focal gliomatosis peritonei fibrovascular adhesion but with no immature neuroectodermal elements were noted. A cul-de-sac nodular implant was positive for tumor involvement. Ascitic fluid cytology was negative for malignant involvement but did have mesothelial cells and macrophages in a background of proteinaceous fluid-pT2bpNx International Federation of Gynecology and Obstetrics (FIGO) stage IIB.

The subject tolerated the surgery well. She began to experience abdominal swelling again on Post-Operative Day (POD) 3, as well as decreased fetal movement. Subsequently, she went into spontaneous labor on POD 5. Within 1 hour of first complaining of abdominal cramping on that day, she was delivered vaginally of a male infant at 24 weeks and 2 days EGA. The baby was incubated and taken to the neonatal intensive care unit, where he died of extreme prematurity and respiratory failure on day 2 of life. The mother experienced an uncomplicated postpartum course, other than continued abdominal distension. Computerized Tomography (CT) scanning revealed a large amount of ascites (but no other abnormalities) was present in the abdomen.

The subject underwent extensive counseling regarding fetal demise, as well as prognosis and subsequent therapy for her cancer. She agreed to receive chemotherapy, including a combined regimen of Bleomycin, Etoposide and Cisplatin (BEP), given intravenously once every 4 weeks for 3 total cycles starting on POD 11. After the first administration of BEP, the ascites resolved, and the subject reported that she was feeling much better. She tolerated the treatment well with no reported toxicities, and a CT scan performed after the third (final) cycle showed no evidence of disease.

Discussion

The incidence of ovarian cysts during pregnancy varies from 1 in 100 to 1 in 2000 pregnancies and malignant ovarian tumors during pregnancy are uncommon [4]. Further, ovarian germ cell tumors, while rare overall, usually consist of mature teratomas and most often occur in postmenopausal women, Hinshaw *et al.* [5]. Primitive neuroectodermal tumors are presumed to arise from the cells of the primitive neuroectoderm located in the neural crest [6].

Pathologists usually restrict the term teratoma to tumors differentiating tissues from all 3 germ layers [7], and many ovarian teratomas do in fact contain tissue from all 3 layers. However, monodermal ovarian teratomas with 1-sided tissue differentiation (monophyletic teratomas), such as Struma Ovaarii are frequently reported.

Interestingly in this case, immunostaining revealed the neuropil/neuroglial tissue was reactive for synaptophysin, NSE, S-100 protein, neurofilament and GFAP. The primitive neural tube/medulloepithelioma patterns were not reactive but did stain for pankeratin, as well as EMA (weakly) and glypican-3. Medulloepithelioma has been reported to be reactive for glypican-3. The medulloepithelioma component had a very high nuclear proliferation index with the background neuroglial tissue having only moderate reactivity.

In general, ovarian malignant neuroectodermal tumors are highly aggressive and the prognosis is poor especially in the presence of extra-ovarian spread. The Gliomatosis Peritonei (GP) in this patient is not necessarily an indicator of poor prognosis as its behavior is benign, since mature glial cells are not aggressive and remain stable for long periods of time. However, on rare occasions, GP can induce florid vascular proliferation that may result in peritoneal hemorrhage and shock or transform into a secondary malignant glial tumor [8]. The patient's presentation with massive ascites makes her prognosis somewhat more concerning.

The rarity of this tumor precludes a randomized clinical study and the management in the setting of pregnancy further complicates this case. Fertility-preserving surgery followed by chemotherapy for early stage primitive neuroectodermal tumors PNET of the ovary with successful pregnancies has been reported (by Demirtas *et al.*, 2004) but the outcome in more advanced cases is uncertain. The optimum chemotherapy regimen for the treatment of PNET transformed from germ cell tumors of the ovary remains unknown [9]. Management of these cases is based on case reports and largely extrapolated from our knowledge of the transformed male germ cell testicular tumors. For PNETs that are transformed from germ cell tumors, some authors advocate for the use of platinum based chemotherapy. Many other authors however have felt that despite arising as germ cell tumors, PNETs are usually resistant to cisplatin-based treatment [10] and therefore advocate chemotherapy regimens comprising of doxorubicin, ifosphamide and cyclophosphamide, directed at the transformed PNET component [3,11]. In one of the largest series by Ehrlich and colleagues CAV/IE (cyclophosphamide/doxorubicin/vincristine alternating with Ifosphamide and etoposide), was the treatment of choice for PNETs transformed from testicular teratoma [11]. Yet other authors had used integrated chemotherapy regimen that targets both PNET and germ cell tumor [12,13]. Although published evidence is limited, a few cases of PNET have been reported in pregnancy, suggesting a possible effect of maternal hormones.

Conclusion

PNET of the ovary during pregnancy is rare. The management involves a combination of surgery and chemotherapy. Disease stage appears to be the most important prognostic factor for the mother, and EGA at diagnosis is the most important consideration for the infant. Survival at 24 weeks EGA (as in this case) is uncommon and frequently complicated by multiple developmental defects. The mother/patient should be carefully counseled regarding all these factors, preferably in a multidisciplinary setting which includes an obstetrician, oncologist and neonatologist.

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