Management of Vaginal Delivery in a Parturient with Ewing's Sarcoma of the Adrenal Gland.

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Citation

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Abstract

Ewing's sarcoma is a tumor affecting bone and soft tissue which usually occurs in childhood. Extraosseous primary disease is uncommon. Patients initially diagnosed with local disease have a good cure rate with resection, multiagent chemotherapy and radiation treatment. Older patients with disseminated disease have worse outcomes.[1] We present the rare case of a 31 year old parturient diagnosed with an adrenal tumor during pregnancy, later found to be an Ewing's sarcoma. As malignancies in pregnancy are rare, few reports of this tumor during pregnancy can be found and those discuss delivery via cesarean.[1,3,4] This is a rare case of a patient who delivered vaginally with this tumor. The location and size of this tumor present several considerations for the anesthesiologist if a vaginal delivery is planned.

INTRODUCTION

Ewing's sarcoma is a tumor affecting bone and soft tissue which usually occurs in childhood. Extraosseous primary disease is uncommon. Patients initially diagnosed with local disease have a good cure rate with resection, multiagent chemotherapy and radiation treatment. Older patients with disseminated disease have worse outcomes.[1] We present the rare case of a 31 year old parturient diagnosed with an adrenal tumor during pregnancy, later found to be an Ewing's sarcoma. As malignancies in pregnancy are rare, few reports of this tumor during pregnancy can be found and those discuss delivery via cesarean.[1,3,4] This is a rare case of a patient who delivered vaginally with this tumor. The location and size of this tumor present several considerations for the anesthesiologist if a vaginal delivery is planned.

CASE REPORT

A 31 year old G4P2 parturient was diagnosed with a complex left adrenal mass (14x13.4x10 cm) at 24 weeks gestation during an ultrasound. This mass was believed to be an adrenal hematoma after urine metanephrines at that time ruled out the likelihood of pheochromocytoma. Repeat ultrasound at that time also revealed a slight decrease in the size of the mass, therefore, she was sent home with instructions to follow up in two weeks. Her past surgical history included salpingectomy resulting from an ectopic pregnancy five years ago. The mass was followed by ultrasound and the obstetric plan was for cesarean delivery and possible tumor resection under CSE anesthesia at 35 weeks. At 34 weeks the tumor was noted to have significant enlargement (14.1x23.2x26 cm), increased vascularity concerning for adrenal cortical carcinoma. Two subcentimeter hepatic lesions were also noted. A dexamethasone suppression test was positive, indicating functional adrenal insufficiency as a result of cortisone secretion from the mass, and she was started on stress dose hydrocortisone, 100 mg every eight hours to continue until 24 hours after delivery, in addition to hydromorphone for severe left flank pain.

It was at this time that she presented to anesthesia for consultation. Airway exam revealed a Grade I Mallampati view, adequate cervical motion, good jaw mobility and adequate hyomental distance, implying a good airway for endotracheal intubation. Urine metanephrines were again checked by the obstetric team and found to be elevated. A third sample was normal, and endocrine consultation revealed no evidence of pheochromocytoma. Discussion between the anesthesia, obstetric and surgical teams gave rise to a differential diagnosis including cortical carcinoma, lymphoma, and ovarian cancer; although we continued to consider pheochromocytoma as a result of the one elevated set of labs. As a result of the tumor's continued enlargement, significant vascularity, and possible metastases, feasible excision during the cesarean was impossible and, therefore, a vaginal delivery was decided upon. Cesarean delivery would only be performed for obstetric or fetal indications if

necessary. Believing that the mass was likely benign, our oncologic surgeon planned to excise the mass a several weeks after delivery to allow involution of the uterus.

The anesthetic plan was for epidural anesthesia with controlled titration of the anesthetic level, as well as confirmation of the availability of the operating room for possible emergent cesarean section. Epidural anesthesia would provide analgesia for labor, as well as allow for extension of the level for cesarean section, if necessary. Also, the sympathectomy provided by an epidural anesthetic may provide moderation of hypertension if the tumor did manifest properties of a pheochromacytoma intraoperatively, although this has mixed results in patients treated with alpha blockers for known pheochromocytoma.[9,10] Labor was induced with a foley bulb, followed by oxytocin. Two large bore intravenous lines were also placed at this time, and equipment was available for an arterial line if the patient became unstable. The immediate availability of blood products was also confirmed. In consideration of the possibility of a pheochromocytoma, availability of nitroprusside, labetolol, esmolol, phentolamine, and other antihypertensives were confirmed. If the patient began having any hypertensive episodes or labile blood pressures, and arterial line would be placed and appropriate therapies would be given. When the patient began making adequate cervical change, as determined by the obstetric team, an epidural was placed without difficulty at the L4 level, dosed with 10ml 0.125% Bupivicaine to a T10 level, and maintained with 0.1% bupivicaine plus 1.5 mcg/mL fentanyl. The patient labored for approximately five and a half more hours and a 5.5 lb baby girl was delivered vaginally (APGARS $9^{1}/9^{5}$) without complication. No hypertensive episodes occurred, consistent with the tumor having an origin other than pheochromocytoma.

Post partum CT and MRI imaging revealed a 20-cm adrenal mass with renal invasion, but no hepatic metastases. An exploratory laparotomy and biopsy was performed on postpartum day 32 after involution of the uterus. Unfortunately, the large tumor size and encasement of the celiac axis and aorta precluded resection. Initial pathology showed a small cell tumor, while further examination and staining of the tumor revealed an extraskeletal Ewing's Sarcoma (primitive neuroectodermal tumor – PNET), which had not been considered in her differential diagnosis. The patient subsequently underwent four cycles of chemotherapy with alternating etoposide/ifosfamide and cyclophosphamide, doxorubicin, and vincristine (CAV); followed by chemoembolization. She had significant decrease in her tumor size for six months, at which point she had a relapse treated with gemcitabine and taxotere. She expired from complications of her disease approximately ten months after diagnosis.

DISCUSSION

There are four reported cases of adrenal tumors in pregnancy in the English language literature over 22 years – one pheochromocytoma, one neoplasm associated with Von Hippel Lindau disease, and two benign adrenal adenomas.[3] Ewing's sarcoma is rare; PNET is extremely rare and occurrence during pregnancy has only been reported a few times. Ewing's sarcoma family of tumors describes a group of tumors with similar histological, immunochemical and chromosomal features.

Malignancies in pregnancy are uncommon, (0.02% to 0.10% in all pregnancies); adrenal tumors are even more rare.[2] The most common of all adrenal tumors are pheochromocytomas. In this case, as pheochromocytoma was eventually excluded, we assumed a vascular tumor based on early ultrasound findings. Thus, our anesthetic plan included preparation for massive bleeding while providing analgesia for the expected mode of delivery.

In our case, the multidisciplinary team (Maternal-Fetal-Medicine obstetrician, oncologic surgeon, endocrinologist, obstetric anesthesiologist), agreed to deliver the baby first and then approach definitive treatment for this tumor as the chances of malignancy appeared small.

Of seven case reports of peripartum Ewing's sarcoma, only one case was extraosseous, two cases had symptoms which began after conception, and all cases involved cesarean delivery.[4,5,6] Although no formal relationship between Ewing's sarcoma and pregnancy has been established, these cases lead one to believe that this tumor grows rapidly during pregnancy. When PNET occurs in a solid organ, it has been found in the kidney (most often), ovaries, uterus, abdominal cavity/retroperitoneum, small bowel, and lung.[7]

The prognosis for the mother with PNET is poor, but fetal outcomes have been good. In several reported cases, chemotherapy was initiated prior to delivery without apparent injury to the fetus.[6] Our patient's initial diagnosis of adrenal hematoma would not require immediate treatment and was further delayed until definitive diagnosis at laparotomy was established. It is unclear whether the delay in definitive diagnosis impacted her overall prognosis, as the

tumor was large at initial presentation and may have already metastasized. Disease free survival for a large soft tissue based Ewing's tumors with metastases is less than 30% at five years.[8]

Anesthetic considerations in a parturient with a large adrenal mass are many. First, regardless of the route of delivery one must anticipate an increased blood loss in this case than from a routine delivery. Therefore, blood and platelet counts should be evaluated and blood products made readily available. Second, adrenal function, electrolyte abnormalities, and blood pressure fluctuations should be evaluated and treated. In our case, the only confirmed abnormality, adrenal insufficiency, was appropriately treated with steroids. Although unlikely to be needed, nitroglycerin was also immediately available for blood pressure control in the rare instance that this tumor was indeed a pheochromocytoma. An epidural was placed and slowly titrated to provide labor analgesia and minimize hypotension. Lastly, airway evaluation and plans for emergent cesarean delivery are important.

In conclusion, this is the first reported case of a vaginal delivery in a patient with PNET arising from the adrenal gland in pregnancy. These tumors are extremely rare, difficult to treat, and require multidisciplinary cooperation. Anesthetic management for these cases requires understanding of not only the airway and physiology of obstetric patients, but also of the potential complications from this rare tumor.

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