Prenatal and Neonatal MRI of Sacrococcygeal Teratoma With Surgical Correlation

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging

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Abstract

We report a case in which a sacrococcygeal teratoma was identified and characterized on prenatal and postnatal MRI, and correlated with subsequent surgical resection.

Case Report

A 19-year-old gravid woman at 29 weeks gestation underwent routine prenatal sonography. A large fluid filled, intrauterine structure was found and she subsequently was referred to a tertiary care facility for further evaluation and management. She denied nausea, vomiting, acute abdominal pain, or recent trauma, and her pregnancy had been progressing well. An MRI was obtained to further delineate the lesion found on sonography.

MRI showed large cystic mass arising from the sacrococcygeal region of the fetus was depicted suggestive of a sacrococcygeal teratoma (Fig. 1). There was no evidence of polyhydramnios. The patient was counseled and monitored until she presented in labor eight weeks later, with contractions every ten minutes, nausea, vomiting, and apparently decreased fetal activity. She was taken urgently to the operating room where a cesarean section was performed and the fetus was delivered without complication. At the time of delivery, the female neonate was noted to have a large sacrococcygeal mass (Fig. 2). The mass measured 19.1 cm by 9.7 cm by 13.2 cm. The neonate underwent a MRI scan to characterize the mass and delineate its anatomic extent and relationships with other structures. MRI showed that the mass was cystic with large extrapelvic and small intrapelvic components (Fig. 3). The mass had no solid components, and there was no involvement of the neural tube.

The neonate was subsequently taken to surgery where the lesion was resected and coccygectomy was performed. Pathological examination of the resected specimen revealed multiple cystic components with the presence of three germ cell layers. The ectodermal component consisted of stratified squamous epithelium, the mesodermal component included both fat and smooth muscle, and the endodermal layer showed with cuboidal and columnar cells. The final pathologic diagnosis was benign, well-differentiated sacrococcygeal teratoma. Post-operatively, the mother and child both did well and were discharged home in stable condition.

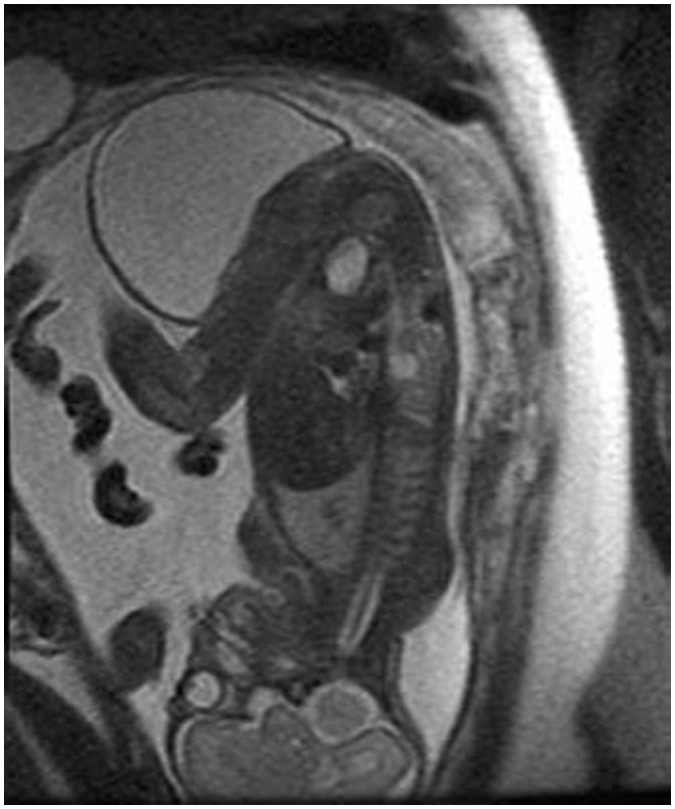

Figure 1. MRI (T2 weighted images) of the maternal pelvis depicting a large cystic mass arising from the sacrococcygeal region of the fetus suggestive of a sacrococcygeal teratoma

Figure 2. Gross image of the sacrococcygeal teratoma following cesarean section delivery of the neonate.

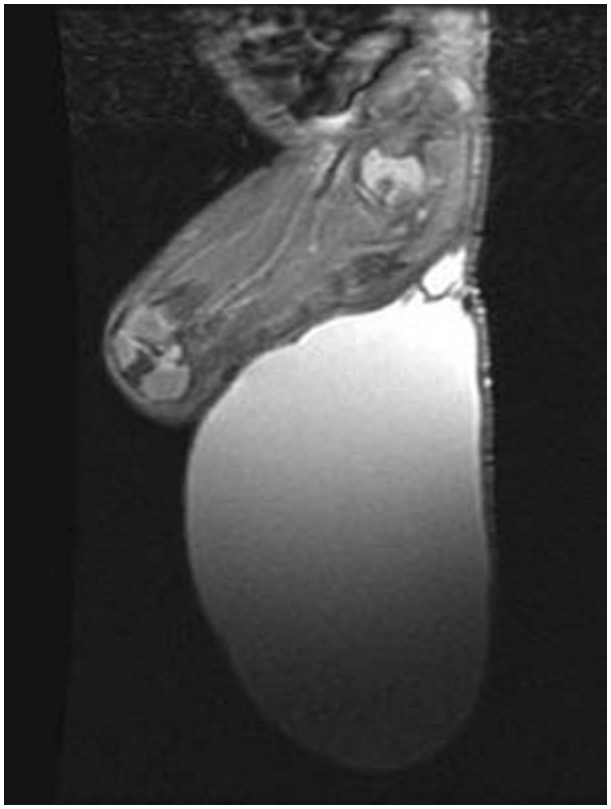

Figure 3. RI (T2 weighted images) of a cystic mass with a large extrapelvic component and demonstrating a small intrapelvic component. The mass depicts a predominantly fluid signal intensity without any evidence of soft tissue, calcification or fat.

Discussion

Sacrococcygeal teratomas are the most common tumors in newborns with an incidence of 1 per 20,000 - 40,000 births. They range from benign well differentiated cystic lesions to malignant solid masses. Female prevalence is particularly higher with a female-to-male ration of 3-4:1. The most common location is the sacrococcygeal region, accounting for 57% of cases, followed by the ovaries and testes respectively [1]. Infrequent locations include the retroperitoneum, mediastinum, and intracranium. Teratomas consist of a consortium of parenchymal cells representing more than a single germ layer, usually derived from totipotential cells that are midline or paraxial. The major complication is malignant degeneration which is manifested in 11-35% of teratomas, more commonly exhibited when in the presence of squamous components. Although the potential recurrence of these tumors can be monitored with the tumor marker alpha-fetoprotein, the absolute serum level of this tumor marker does not have prognostic significance [2,3].

The diagnosis of sacrococcygeal teratoma is often made in the prenatal period. Although it is commonly diagnosed in utero, it may also present with presacral tumors manifesting as masses that create an asymmetry of the gluteal fold in babies up to four months of age. It is imperative that sacorcoccygeal tertatomas be differentiated from other neural tube defects, particularly meningoceles or meningomyeloceles. Our patient underwent an MRI which depicted the large mass extending from the fetus without involvement of the neural tube. Sonography does have the capability to delineate the lesion, however does not offer the sensitivity of MRI, especially when imaging the lower spine. One study revealed the sensitivity and specificity of transvaginal ultrsonography to be 84.6% and 98.2%, respectively, in differentiating ovarian teratoma from other pathology [4]. CT scanning is also a useful modality for evaluating teratoma size and consistency, however, it does expose the fetus and mother to radiation. MRI has the ability to differentiate tissue density and thus serves as an excellent tool to evaluate teratomas, particularly vascular anatomy and peripheral margins. Evaluation of the pre-sacral space with ultrasound, CT or MRI is also important and contrast may be administered to demonstrate anterior displacement of the rectum and bladder.

There does exist a staging system for sacrococcygeal teratomas (Table 1) [1,5]. Most tumors are type I and amenable to surgical management, with several studies documenting cure rates in excess of 95% [5]. The lesion in our case was Altman type I. Complete resection offers cure to the patient, and usually is completed through a chevron shaped buttock incision with particular attention paid to the preservation of the rectal sphincter. The coccyx is part of the surgical specimen primarily due to the high recurrence rate (35-40%) should it be left behind. Hemorrhage is the most common complication attributed to the middle sacral and hypogastric vessels. Fetuses with sacrococcygeal teratomas measuring larger than 5 cm should undergo cesarean section delivery to prevent tumor rupture. Larger lesions have the potential go undergo tumor hemorrhage, polyhydramnios or nonimmune hydrops fetalis. Significant vascular shunting can predispose the mother to high-output cardiac failure and warrants emergent delivery. Development of hydrops prior to 30 weeks gestation carries an ominous 93% infant mortality rate stemming primarily from lung immaturity. Emergent delivery addressing hydrops after 30 weeks gestation is still is associated with a 25% mortality rate. Tumors which present with significant extension and/or metastases have been treated with neo-adjuvant cisplatinum-based chemotherapy with success. One study reported a median of eight cycles of neo-adjuvant chemotherapy followed by surgical resection rendered an 84% patient survival for locally advanced disease [6,7].

Recurrent malignant sacrococcygeal teratomas present a challenge for both the patient and clinician. Salvage surgery remains the mainstay of therapy, however sometimes does not eradicate the tumor completely. Preoperative platinum based chemotherapy in combination with regional hyperthermia have conferred some success [8]. There also is some evidence for the role of radiotherapy should microscopic margins remain positive for cancer [8]. The complete eradication of malignancy via complete surgical resection remains the best management and the ability to confer hope for the patient, particularly a young child.

Table 1: Altman Classification of Sacrococcygeal teratomas [5]

Type	Description
I	Predominantly external, attached to the coccyx. No evidence of metastases.
II	External mass and significant presacral pelvic extension (34%) minimal metastases rate
III	Tumors visible externally, mass is pelvic and intrabdominal. 20% rate of metastases.
IV	Tumors not visible externally, entirely presacral.

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