Fetal Sacrococcygeal Teratoma

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Citation

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Abstract

Sacrococcygeal teratomas (SCT) are neoplasms, composed of a wide diversity of tissues foreign to the anatomic site in which they arise. We report a case of a multi gravida at 38 weeks with a Type 1 sacrococcygeal teratoma. The diagnosis was made prenatally and the patient underwent a cesarean section due to pre labor rupture of membranes. Surgical resection of the tumor was performed on the 5th postnatal day and histological examination confirmed the diagnosis of a benign SCT. The baby is being followed up regularly and at six months of age has no evidence of recurrence of the disease. Management of fetal sacrococcygeal teratoma requires a multidisciplinary team approach to maximize infant survival.

INTRODUCTION

Teratomas are defined as tumors that are derived from more than one embryonic germ layer. Sacrococcygeal teratomas (SCT) are neoplasms, composed of a wide diversity of tissues foreign to the anatomic site in which they arise. It is now generally accepted that teratomas are not a variant of fetus in fetu or twins, but originate from totipotent cells from Hansen's node or primitive germ cells during their migration from the yolk sac to the genital ridge[1]. They are mostly gonadal in origin and rarely present at extra-gonadal locations. They are of two types' i.e. benign (mature cystic teratoma) and malignant (immature teratoma). Fetuses with this malformation may have associated morbidity and mortality. However, improvement in ultrasonography and the early detection of associated complications have improved the perinatal mortality rate for this condition. Standard treatment for SCT is complete excision after birth. We report this case study in order to demonstrate how a multidisciplinary team approach can maximize infant survival when the diagnosis of SCT is made prenatally.

CASE REPORT

A 30 year old multigravida, at 38 weeks of gestation was received as referral to our hospital because of a disproportionately increased uterine size and breech presentation. She was non-diabetic, non-hypertensive, and there was no history of serious infections or exposure to teratogens. Fetal anomaly scan at 5 months of amenorrhea was said to be normal. Her personal and family history did not contribute significantly. On examination, the uterus was over distended, with polyhydramnios and breech presentation. Ultrasound showed a single live fetus corresponding to period of gestation with polyhydramnios and breech presentation with a large sacrococcygeal teratoma measuring 15 x 17.5 cms showing cystic and solid components (Fig 1). The patient was scheduled for planned cesarean delivery but due to pre labor rupture of membranes had an emergency cesarean section instead. It was a live female baby of 3.3 kg, with a tumour measuring 26 x 17 cms, extending from sacrum to upper part of the thigh (Fig 2). There was no other associated anomaly. On the 5th neonatal day, complete surgical excision of the tumour was performed. Baby withstood surgery well but for a transient anal sphincter incontinence on the second and third post-operative days. Baby was discharged on the 15th post operative day with no neurological deficit at discharge. The histopathology report confirmed an immature teratoma. The baby is now 6 months old, active, with good milestones and being monitored regularly for recurrence of the disease.

Figure 1

Figure 1: Ultrasound showing a fetus in breech presentation with a large sacrococcygeal teratoma with cystic and solid components.

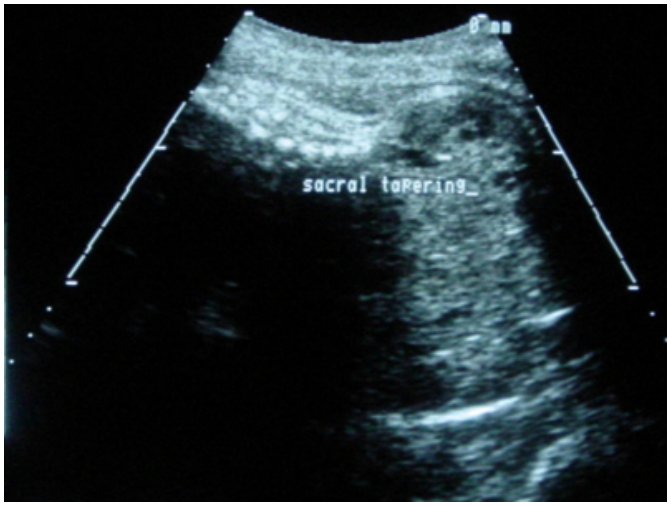
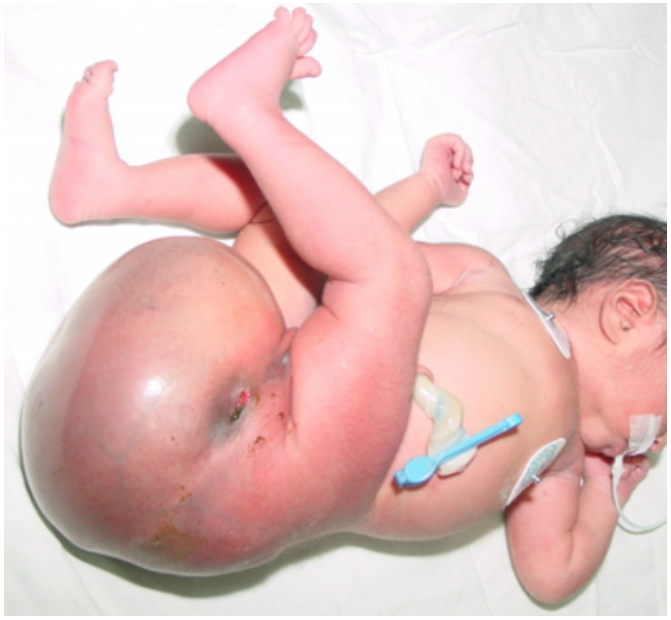


Figure 2

Figure 2: Baby with a tumour (Sacrococcygeal teratoma) extending from sacrum to upper part of the thigh



DISCUSSION

Sacrococcygeal teratoma (SCT) is the most common congenital neonatal tumor and has an incidence of 1-2 per 40,000 deliveries [2]. Teratomas are derived from all three germinal layers and contain neural elements, squamous and intestinal epithelium, skin appendages, teeth, and at times, calcium. The sacrococcygeal region is the most frequent site. The exact etiology of most SCTs is unknown; majority are thought to be sporadic. A few families with autosomal dominantly inherited presacral teratomas have been described in the literature [1, 3].

Commonly seen in girls than in boys (4:1), but it is more often malignant in boys. 15% of patients have associated congenital anomalies like imperforate anus, sacral bone defects, and duplication of uterus or vagina, spina bifida, meningomyelocele. Screening methods such as amniotic fluid -fetoprotein level or maternal serum screening have not been proven to be effective, but can be used postnatally to differentiate between benign and malignant SCT [1, 6].

The most common indication for antenatal ultrasound in SCT is a uterus that is large for gestational age either due to polyhydramnios or rapid growth of the tumour. On antenatal ultrasound, a SCT appears as a mass of cystic, solid or mixed consistency which arises from the sacral area[1]. Completely cystic or partially cystic lesions are usually benign, whereas solid lesions are frequently malignant.

Fetal complications include spontaneous haemorrhage into the SCT, nonimmune hydrops fetalis, malignancy, renal calyceal dilatation and bladder outlet obstruction. The presence of hypervascularity within SCTs is a risk factor for arteriovenous shunting, which may cause high-output cardiac failure which in turn may lead to the development of nonimmune hydrops fetalis. The development of hydrops fetalis is an ominous sign. It may be fatal or be associated with polyhydramnios and ultimately preterm labour[7, 8]. Obstetric complications associated with this condition are polyhydramnios, preterm labour, pre-eclampsia and dystocia. Large SCTs cause dystocia during delivery 6% to13% of the times.

Sacrococcygeal teratoma being a potentially malignant tumor, an early diagnosis helps in management. Intrauterine surgery has been suggested when the fetus develops hydrops prior to viability in an attempt to interrupt the feeding vessel[9]. The role of open intrauterine surgery however, is limited since it carries high morbidity to the mother and the fetus. Delivery in a tertiary center is recommended where an experienced neonatologist and pediatric surgeon will be immediately available. In a viable fetus, caesarean section is the mode of delivery of choice when SCT measures more than 5 cm in order to prevent dystocia or rupture of SCT during delivery. Sacrococcygeal teratomas should be excised as soon as possible, because small, undifferentiated foci may proliferate and become aggressive. Most infants can safely undergo a period of stabilization prior to surgery. Since the tumors are attached to the coccyx, excision of the tumour in

continuity with the coccyx should be achieved. Failure to remove the coccyx results in a 30 to 40 per cent risk of local recurrence. Malignant tumours require surgical excision, chemotherapy and radiation (metastases to lung, bone, liver). Prognosis depends on size and histology of the tumor, degree of prematurity, associated malformations, route of delivery and prompt and complete surgical removal. Risk of malignancy depends on the time of diagnosis, at less than 2 months of age, only 7-10% are malignant, at 1 year 37%, and at 2 years 50% are malignant. In benign tumors, disease free survival is greater than 90%, whereas a malignant tumour has significant mortality. Hence frequent follow-up is necessary with measurements of serum fetoprotein level, and surveillance by imaging is recommended for at least 3 years.

CORRESPONDENCE TO

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