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PRENATAL DIAGNOSIS OF SACROCOCCYGEAL TERATOMA ON ULTRASONOGRAPHY : A CASE REPORT

Radiology

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ABSTRACT

Teratomas are germ cell tumors which arise from ectopic pleuripotent stem cells which fail to migrate from yolk sac endoderm to the urogenital ridge. It contains elements from all three embryological layers – endoderm, mesoderm and ectoderm. "Teratoma" is a Greek word for "monstrous tumor", so named because it contains hair, teeth, bone, neurons and large variety of tissue types. Depending upon tissue of differentiation, they are divided into mature, immature and malignant. Sacrococcygeal teratomas (SCT) are the most common extragonadal germ cell tumors. The incidence of SCT in infants and children is 1 in 35000 to 40000 live births. Here we report a case of SCT arising from the sacrococcygeal region in a 17 weeks fetus. The patient was a 20 year old female who was gravida 1 para 0 and abortion 0 (G1P0A0) and was diagnosed during her routine prenatal ultrasound examination at 17 weeks of gestation. The patient was counselled for termination of pregnancy and type I SCT was diagnosed clinically. We present this case to highlight the importance of prenatal B-mode and color doppler two dimensional ultrasonography (2D US) in evaluation of SCTs.

KEYWORDS

Sacro-coccygeal Teratoma, Extra-gonadal, Germ cell tumor, Ultrasonography, Prenatal diagnosis

INTRODUCTION:

Germ cell tumors arise primarily within the gonads, but a significant minority can also be found in extragonadal midline locations, including the sacrum, presacral space, coccyx, mediastinum, retroperitoneum, and pineal/intracranial space.[1] SCTs are believed to originate from pleuripotent cells in Hensen's nodule which is located on anterior surface of the sacrum and coccyx. Multiple works of literature have been published to date about the prenatal diagnosis of sacrococcygeal teratoma in second trimester of pregnancy.

The incidence of SCT is 1:40000 and female to male ratio of 4:1.[2] At prenatal ultrasound, majority of these tumors are seen as solid or mixed cystic and solid, external caudal mass. Fetuses with large teratomas present a high perinatal mortality rate, caused by high-output heart failure resulting from arteriovenous communication, with subsequent polyhydramnios and hydrops.[3]

Although majority of these tumors are histologically benign but they can cause multiple fetal and maternal complications due to mass effect like dystocia, traumatic delivery and intratumoral hemorrhage.

Case Report :

A 20 year old primigravida (G1P0A1) came to obstetrics OPD at our institution. There was no history of consanguineous marriage and no family history of anomalous babies.

On general examination her BP was 120/80 mmHg, pulse rate was 82 bpm and there was no pallor or icterus. On clinical examination, respiratory and cardiovascular system was normal. Per abdomen examination showed a relaxed uterus of 15 to 20 weeks. Blood investigations revealed complete blood count, blood sugar level, liver function tests, kidney function tests within normal limits.

The patient was advised second trimester prenatal ultrasonography as a part of routine investigation in 2nd trimester. A two dimensional (2D) ultrasound (US) examination was performed in our institution on "MINDRAY DC-60" ultrasonography equipment, which revealed a single live pregnancy of 17 weeks 2 days with fetus presenting a bulky loculated solid mass in sacrococcygeal region with cystic components and few calcific foci within measuring 27 x 27 x 34 mm in size. On color doppler there was internal flow within the solid component. The mass was completely extra-pelvic in location.

To avoid the complications the patient was counselled for termination of pregnancy. After the termination of pregnancy diagnosis of type I SCT was confirmed clinically.

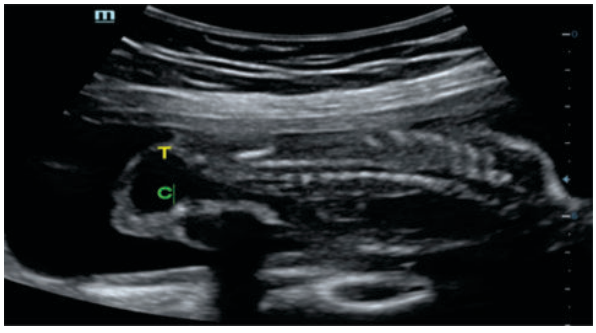

Fig.1: Sagittal section on ultrasound showing a hypoechoic solid cystic tumor mass arising from sacrococcygeal region of fetus suggestive of SCT. T: tumor mass, C: cystic area

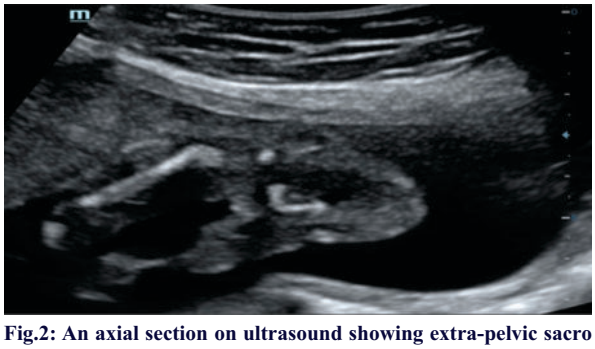

Fig.2: An axial section on ultrasound showing extra-pelvic sacro coccygeal mass (Arrow).

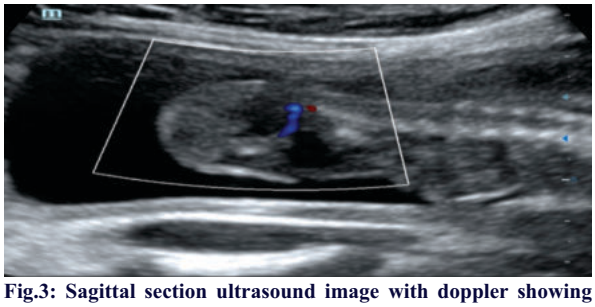

Fig.3: Sagittal section ultrasound image with doppler showing internal vascularity of the tumor

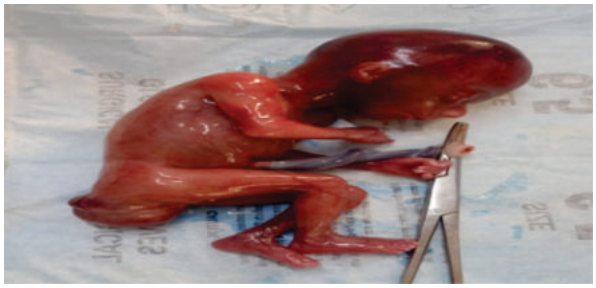

Fig.4: Gross specimen of fetus showing SCT confirming the prenatal ultrasonography findings

DISCUSSION:

Sacrococcygeal teratomas (SCT) are the most common extragonadal germ cell tumors (GCT) in infants. The prenatal diagnosis of SCT is of utmost importance as undiagnosed cases may undergo grave outcome to the mother and fetus at the time of delivery because large tumors can lead to congestive heart failure, hydrops, and high rates of perinatal mortality and maternal complications like postpartum hemorrhage at the time of delivery due to dystocia. Differentiation between histological types and their potential for malignancy is an important factor in decision making regarding termination of pregnancy and counselling of parents.

Sacrococcygeal teratoma (SCT) is an extragonadal germ cell tumor (GCT) that develops in the fetal and neonatal periods. SCT is a type I GCT in which only teratoma and yolk sac tumors arise from extragonadal sites. SCT is the most common type I GCT and is believed to originate through epigenetic reprogramming of early primordial germ cells migrating from the yolk sac to the gonadal ridges.[4] These pluripotent cells fail to migrate from the control of embryonic inductors and differentiate into various tissues which are aberrant to sacrococcygeal region and thus give rise to SCT. The association of SCT with the common location of sacrococcygeal region is thought to be due to the large number of pluripotential cells which are usually found in the caudal region of the embryo, closely associated with the distal sacrum and coccyx. [5,6]

SCTs are either benign (mature) or malignant (immature, composed of embryonic elements). Mature teratomas are more common in both neonates (68%) and older children (73%).[7]

Altman *et al*. [8] have classified SCTs into four types. Type I Altman tumors are totally extra-pelvic in location with no or minimal pelvic component; type I being the most common type of SCT and almost all cases are benign.

Type II Altman tumors are predominantly extra-pelvic but have a significant pelvic component (hour-glass pattern).

Type III Altman tumors have a larger proportion of intra-abdominal and intrapelvic component than the external component; and Type IV Altman tumors are exclusively presacral with no external component.

SCT are generally described as large, well encapsulated, multi lobulated masses with both solid and cystic component within them.[9] There is an internal vascular stalk in SCT which can be used as point of differentiation of this tumor from meningomyelocele which is a close differential to sacral region mass on ultrasound.

Nowadays the practise of routine serial obstetrical ultrasound has increased the number of SCT cases that are being diagnosed in utero. The SCT that was reported here was described as a completely extra-pelvic, bulky, multiloculated solid mass in sacrococcygeal region with cystic components and few calcific foci within. On color doppler there was internal flow within the solid component accounting to its vascularity. The sonographic appearance of the SCT can vary as it depends on multiple components of the tumor like calcification, cystic components, necrosis and intra-tumoral hemorrhage. The increased echogenicity with post acoustic shadowing can signify the dystrophic calcification in the areas of hemorrhage or rudimentary fragments of maldeveloped bones.

There are many significant prognostic factors for SCT which can be determined on prenatal ultrasound. Large proportion of fetal SCTs are benign tumors but still they are associated with significant morbidity and mortality. Normal development of other internal vital organs of fetus like kidneys and lung can be restricted by compression effect of the tumor. Even lower grades of UTD (urinary tract dilatation) found on prenatal ultrasound can be taken as unfavourable prognostic factor as this hydronephrosis or dysplasia may be caused secondary to urethral obstruction by perineal component of the tumor or direct invasion of ureter or urethra by large retroperitoneal component of SCT.[10]

Other prenatal poor prognostic signs of SCT include appearance of fetal hydrops, increased placental thickness (more than 4 cm), skin thickening on ultrasound.[11,12] Cause of fetal hydrops may be direct compression of major fetal vessels by tumor. If the hydrops develops in a fetus after the gestational age of lung maturity, then emergency delivery of the fetus followed by resection of tumor is appropriate approach for the management. But if it develops before the lung maturity then fetal surgical intervention by an expert surgeon involving debulking and devascularization of tumor may be the only possible way to save fetus. Close observation of mother and fetus by serial ultrasound and corticosteroid administration may aid in the prognosis by hastening fetal lung maturity.[13]

Congenital anomalies may be present in association with SCT, including genitourinary, anorectal, and lower vertebral malformations, and need to be ruled out during prenatal sonography.

CONCLUSION:

This case report will add on the importance of prenatal ultrasonography in diagnosis, prognosis of SCTs and appropriate management and counselling of parents in cases diagnosed with SCT. This case report will help in reducing future complications and to prevent the risk of maternal and fetal morbidity and mortality due to SCTs by early diagnosis with the help of ultrasonography and appropriate management approach.

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