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Case Study

A STUDY OF SACROCOCCYGEAL TERATOMAS IN FETUSES, NEONATES & ADULTS IN CORRELATION WITH EMBRYOLOGICAL CONCEPT

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ABSTRACT

Introduction: The Sacrococcygeal Teratomas (SCTs) are rare and most common congenital neoplasms in neonates, but rare in adults. Usual presentation is, a mass in the sacrococcygeal region at the time of birth and arise from the caudal end of the spine, displacing the anal canal anteriorly. The SCT results in multiplication of totipotent cells of Henson's node (primitive node) which fails to involutes at the end of the embryonic period.

Materials and Methods: We report three cases, with clinical manifestations & imaging aspects. The first case was an abortuses of 12 weeks old with SCT diagnosed by Ultrasonography, second was a female neonate 1 day old with huge SCT and third case was 24 years old female diagnosed as sacral tumor by MRI report.

Conclusion: The antenatal and proper management is carried out after baby is born. It can be diagnosed by prenatal sonography, if necessary MRI during pregnancy to avoid unnecessary complications. In adult, SCTs are diagnosed with abdomino-pelvic ultrasound scan. In this article a brief review of literature and embryological correlation has been presented.

KEY WORDS: Sacrococcygeal Teratomas (SCTs), Ultrasonography, MRI.

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INTRODUCTION

The SCTs are neoplasms present most frequently in infancy and childhood. Willis defined 'Teratomas' as true tumors or neoplasms composed of multiple tissues of kinds foreign to the part in which it arises. The SCTs are composed of several types of tissues usually derived from two or three germ layers [1].

The Ultrasound determines accurate diagnosis of SCT location particularly in infancy and childhood. The SCTs may involve perineum, retroperitoneal, Mediastinum, testis or ovary and they protrude towards perineum or gluteal region. They may be cystic, solid or mixed masses in sacral region. The SCTs have incidence of approximately 1 in 35,000 to 1 in 40,000 live births. Female to male ratio is 4:1 [2,3]. It is rare in adults, less than a hundred cases of Teratomas have been documented in literature. Most of SCTs are cystic and benign and 2% are malignant [4].

The neonatal SCT are benign and has excellent prognosis after surgical treatment. The recurrence rate after surgical resection varies

from 2 to 35% which may be due to incomplete surgical excision [5].

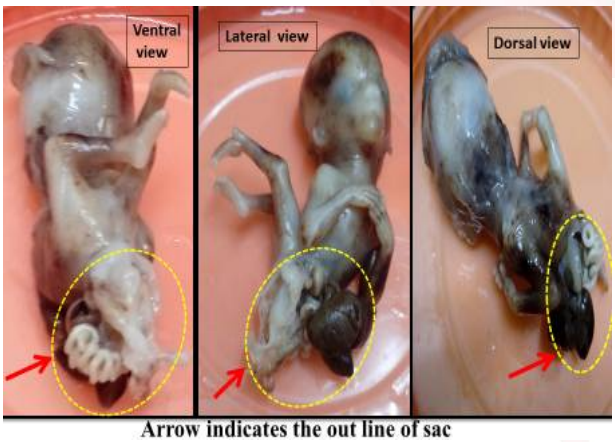
The purpose of the study was designed to determine the SCTs in foetus, neonate and adults with different types, clinical presentations, surgical approaches and correlating embryological implications.

CASE REPORT AND RESULTS

Case report 1: (Fig 1)

A 26 year old G1P1 with Rh negative pregnancy was consulted during 10-12weeks of gestation. An ultrasound examination was performed showing singleton pregnancy associated with anomalous fetus. The diagnosis was sacrococcygeal teratomas of extra pelvic development. Pregnancy was terminated with the consent of the patient. The foetus was delivered with cystic sacrococcygeal teratomas and the sac was ruptured. On examination tumor had fat, muscle, intestines, liver, stomach, pancreas, cartilaginous and bony structures. The spine appeared intact & lower limbs were hyper flexed dorsally. Based on above findings diagnosis of benign SCT of type I was made.

Fig. 1: Case Report 1, Foetus 10 Weeks showing Sacrococcygeal Teratoma with ruptured sac.



Arrow indicates the out line of sac

Case report 2: (Fig 2)

A woman 30 years of age G2P1, non-consanguineous marriage delivered a 2.5 kg girl baby by Caesarian section at peripheral hospital. The mother belonged to poor socio-economic family and did not have any kind of antenatal checkup or ultrasound.

On presentation the girl baby had very large mass of 20 cms x15cms in the sacrococcygeal region. The surface of the mass was highly edematous and there were numerous dilated veins.

On Ultrasound examination, the mass had cystic areas with good blood supply and no intrapelvic extension. The tumor was classified as SCT type I of benign nature.

The general condition of the baby was unstable clinically. The conservative treatment with oxygen, intravenous fluids, and broad spectrum antibiotics were started immediately. The baby died within 3 hours of admission despite of aggressive resuscitation.

Fig. 2: Case Report 2, Neonate 1 Day old, showing large cystic Sacrococcygeal Teratoma.

Case report 3: (Fig 3)

A female patient 24 years old came with low back ache since 1 year, difficulty and pain while walking since 3 months. She also had urinary incontinence since 15 days.

MRI Report:

1. Sacro- coccygeal tumor was moderate in size with extension of mixed hypo/ hyper intense lesions, arising from the sacrum and coccyx involving S2 to S5. This lesion is seen extending posteriorly and laterally abutting gluteus maximus and posterior para-spinal muscles. The mass measures 12x9x5cms displacing the rectum anteriorly.

2. Multiple intramural and sub-serosal uterine fibroids.

3. Follicular cyst in the left ovary.

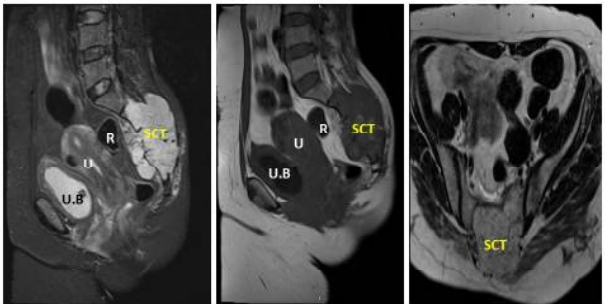
4. Multiple urinary bladder diverticula.

5. The rest of the pelvic musculature and other soft tissues are unremarkable.

These findings are suggestive of? Sacrococcygeal teratoma (SCT) of type IV or Sacral Chordoma.

The patient was operated successfully under General Anesthesia. The tumor was removed & close follow up was done regularly for any recurrence. Histopathology was in favor of SCT.

Fig. 3: Case Report 3, Female 24 years old, showing Sacrococcygeal Tumor (MRI).



Sacrococcygeal tumor-SCT; Urinary Bladder-UB; Uterus-U; Rectum-R

DISCUSSION

The SCT is the most common tumor found in new born and very rare in adults. "Sacro" refers to Sacrum, "Coccy" refers to coccyx and "Teratomas" refers to the type of tissues that forms the growths. Teratomas are derived from 3 germ layers i.e. ectoderm, endoderm and mesoderm. The type of tissues may be fat, bone, nerves, muscles, glands, lung, GIT and hair that are found in an area that they are not normally found. The cause of SCT is not known. One theory is failed twinning attempt, another theory may be an abnormal migration of set of germ cells or pluripotent cells.

The majority of SCTs are benign teratomas and these tumors have tendency for malignant degeneration. The benign are mature and malignant are immature. The mature teratomas are more common in neonates & children [6].

The pluripotent cells from Hensen's node of Primitive streak containing all types of cells from 3 germ layers and their midline distribution can be explained by arrest or aberrant migration of Primordial germ cells. [7] These Pluripotent cells can differentiate into mature or immature teratomas or extraembryonic choriocarcinomas or yolk sac Teratomas. The metastatic spread is rare and it is found in regional lymph nodes, liver, lungs and rarely in bone and vertebrae [8,9].

The Sacrococcygeal tumors are classified according to Altman classification based on American Academy of Pediatrics surgical section [10] (AAPSS) and graded as 4 types:

Type I (47%): Predominantly external tumor with minimal pre-sacral component.

Type II (35%): Present externally but with significant intrapelvic extension.

Type III (8%): Apparent externally but predominantly a pelvic mass extending into the abdomen.

Type IV: (10%): Pre-sacral with no external presentation.

Type IV Sacrococcygeal teratomas may occur as familial form, inherited as an autosomal dominant trait. In this case the pre-sacral mass may be associated with anal stenosis and defect in sacrum. All 4 types may have an intra spinal component and may be associated with neurologic deficit [11,12].

The teratomas may be cystic, solid or mixed. The cystic lesions are predominantly benign. Calcifications occur in 1/3rd of SCTs and frequently seen in benign than malignant tumors [13].

According to Gonzalez-Crussi system SCTs graded microscopically from 0 to 3 [14]

Grade 0: tumors contain only mature tissue.

Grade 1-3: SCTs have immature tissues, with Grade 1containing rare foci of immature tissue, Grade 2 with moderate immature tissue and Grade 3 containing large quantities of immature cells.

Antenatal ultrasound diagnosis of SCT is very crucial to prevent fetal and neonatal deaths. Majority of the fetuses diagnosed as SCT are likely to die before delivery. These tumors associated with complications like malignant invasion, hemorrhage into tumor, high cardiac output failure, obstruction of umbilical flow may lead to perinatal or neonatal mortality. Other complications of SCT is development of hydrops and risk of preterm labor. Hydrops is an abnormal accumulation of fluid in 2 or more areas of fetal body cavities like ascites, pleural effusion, and pericardial effusion or under the skin (anasarca).

MRI followed by three dimensional Ultrasonography provides an excellent visualization of sacral tumors.

In adults SCT may be present with varying symptoms like bowel and bladder incontinence,

backache, numbness of lower limb, fistula of urogenital or gastrointestinal tract and congenital anomalies like cloacal defects [15]

In contrast to presentation in neonates in which 90% of SCTs are externally visible, most of the adult SCTs are internal, asymptomatic and diagnosed accidentally during PR digital examination or retro-rectal radiological investigation presenting with a large mass. The CT followed by the MRI gives better tomography evaluation of the tumor [16,17].

The SCTs have malignant tendency which parallels with increase in age. The incidence of malignancy at neonatal period is 10% followed by 100% at the age of 3 years. Complete resection of tumor after birth provides an excellent prognosis.

Each child with sacrococcygeal germ cell tumor should be investigated for alpha fetoprotein (AFP), which forms an important diagnosis to monitor the therapy and detects metastases or recurrence after therapy. Increase in AFP is commonly seen in both serum and in tissues [18] Excision of the coccyx may be necessary because the bone may contain a nidus of pluripotent cells which increases the risk of recurrence [19]. In adult, renal function tests, liver function tests and tumor markers like alpha feto protein and beta human chorionic gonadotrophin (HCG) should be evaluated before and after the operation [20].

The present pathological study of 3 cases revealed sacrococcygeal teratomas according to Altman classification.

Our first case was female aborted fetus of 10 weeks old is benign SCT of type I. The second case was female baby with large benign SCT of type I which lived only for 3 hours because of poor vital condition. The third case was an adult female 24 years old also presented with**?** malignant SCT of type IV.

The study also agrees with most of the authors presented in the literature with predominance that all cases affected were females.

The purpose of this study is to evaluate our experience with SCTs present in different age group, different types, clinical presentation, surgical outcome of management and treatment strategies and coordinating with embryological correlation.

CONCLUSION

The SCTs are common congenital tumors that develop during early fetal life and very rare in adults. All pregnancies should have regular antenatal check up and prenatal diagnosis by ultrasound and MRI. These investigations play an important role in SCT babies from obstructed labor.

The recommended treatment is surgical resection of the tumor en bloc with coccyx to decrease the risk of recurrence and continue follow up for at least 5 years.

It is my hope that these cases will stimulate lively discussion and give all of us an opportunity to learn something new about these rare anomalies.

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Conflicts of Interests: None

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