A Case Report: Sacrococcygeal Teratoma

Professor Dr Abdul Baser Mangal 1

Assistant professor Dr Samsoor Walizada 2

1 Nangarhar University, Nangarhar Medical Faculty, Neurosurgery Department

2 Nangarhar University, Nangarhar Medical Faculty, Neurosurgery Department

Abstract:

Sacrococcygeal teratomas (SCT) are tumors that arise in the sacrococcygeal area containing tissue from all three germ layers. SCT has an incidence of approximately one in 35,000 – 40,000 live births with malignant transformation with increasing age. We report a case in a 20 day old which was excised completely via the post anal route with no operative or postoperative morbidity.

Sacrococcygeal teratoma (SCT) is the most common tumor presenting at birth. Commonly seen in female fetuses; this case presented, more or less with the whole feature of typical sacrococcygeal teratoma.

Key words: Childhood tumors, Sacrum, Sacrococcygeal teratoma

Introduction:

Sacrococcygeal teratomas (SCT) are tumors that arise in the sacrococcygeal area containing tissue from all three germ layers. SCT has incidence of approximately one in 35,000 – 40,000 live births with malignant transformation with increasing age1, 2.

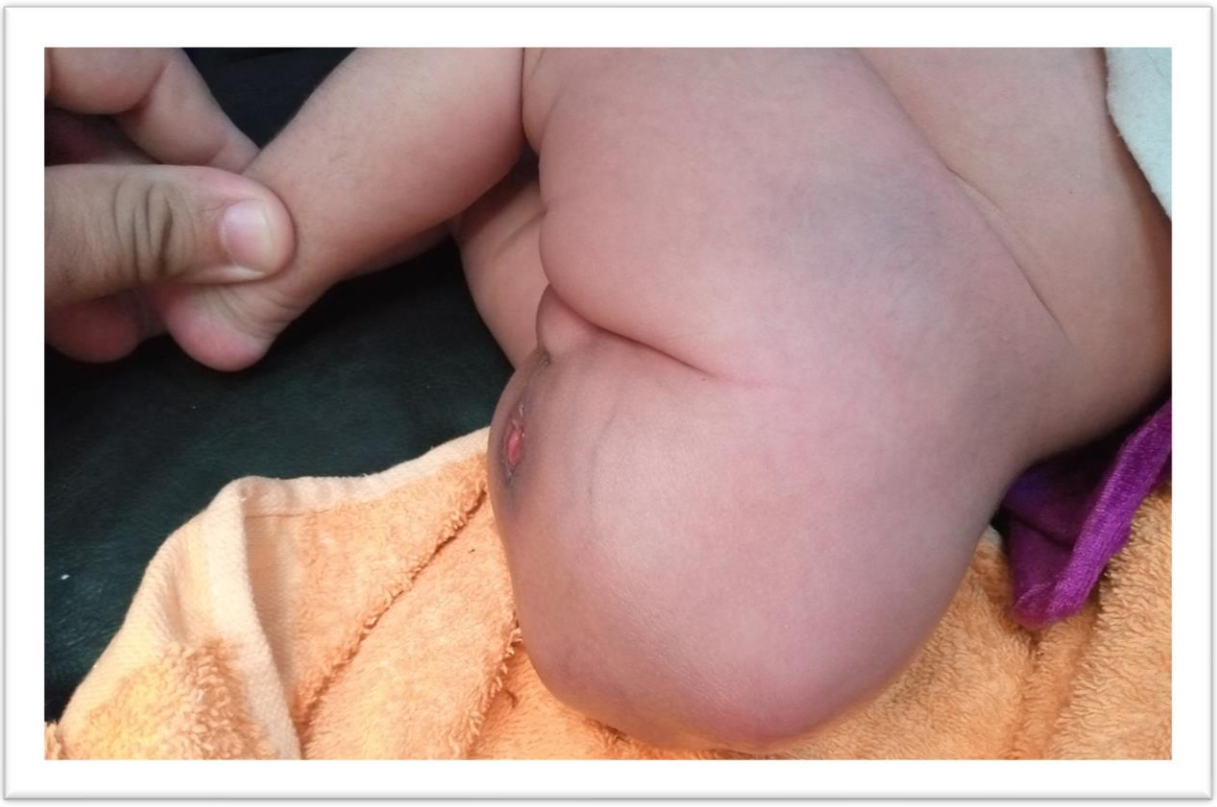
SCT is more common in females with a male, female ratio of 1:3-4, 3, 5.

SCT may present in three categories- fetal, neonatal and children. Prenatal ultrasound can detect fetal tumors with or without maternal symptoms. Those presenting at birth are usually mature or immature teratomas. Large sacral mass that has been increasing progressively is the commonest type of presentation in children. Those presenting late have a poorer outcome than the early detected ones.

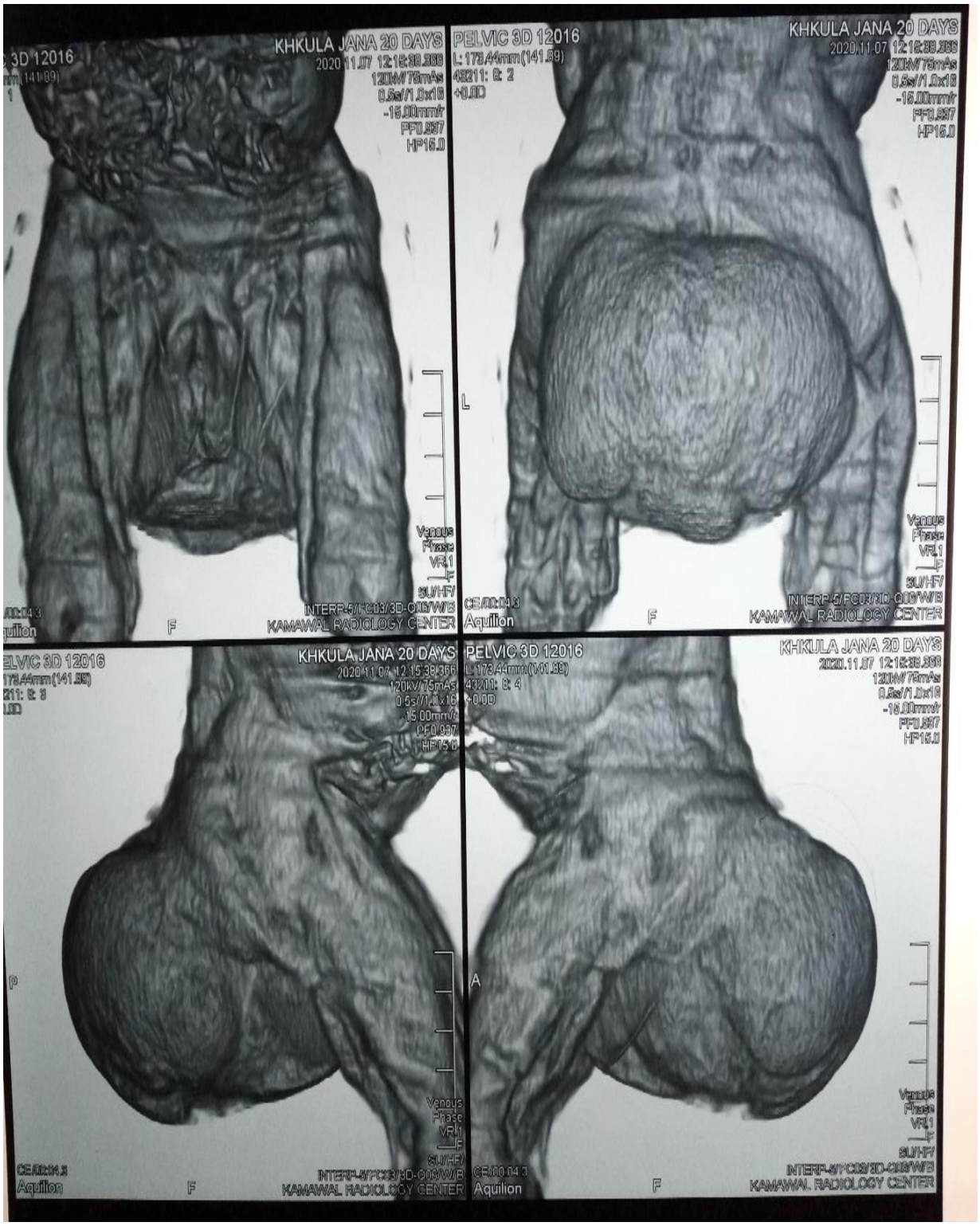
Case Report:

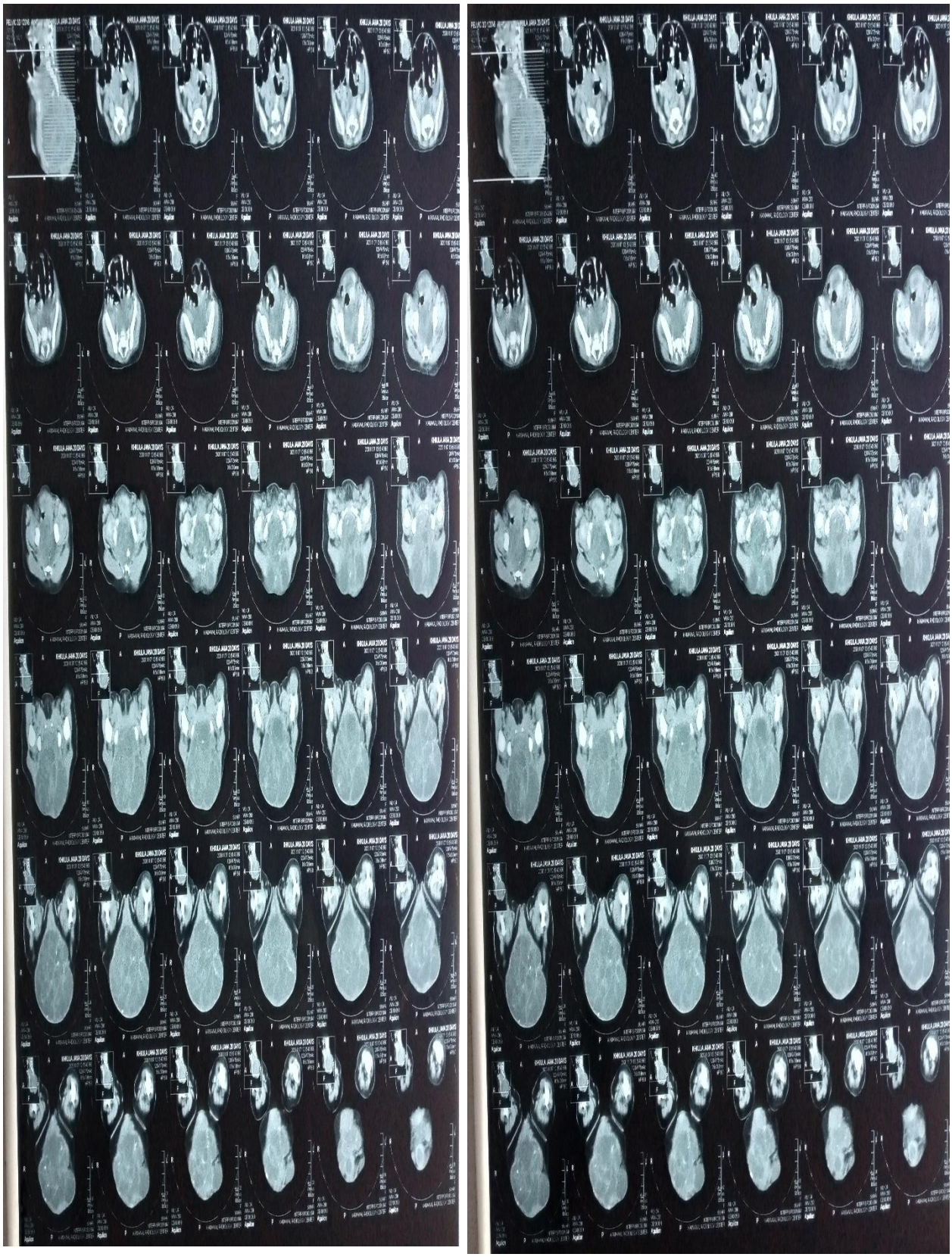
A 20 day old girl presented with a sacral mass in the perianal area since birth. The mass had been slowly and progressive growing to reach the present size. It had also grown to the extent that she could not sleep in supine position. There was no history of altered bowel or bladder habits or weakness of the legs.

On examination the child was alert, adequately built with a large solid-cystic mass of 30x25 cm in the sacrum with the anus grossly pushed anteriorly (Fig. 1).



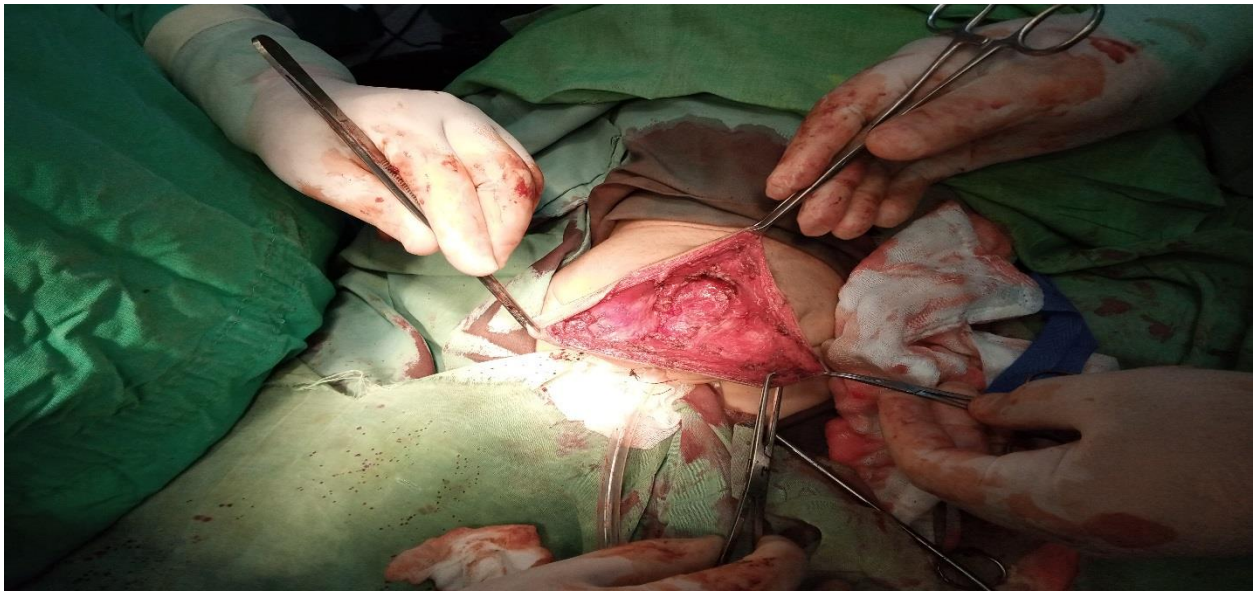
No weakness of the lower limbs or intrabdominal mass was palpable. The ultrasound of the abdomen did not reveal any genitourinary abnormality. Computed tomography (CT) revealed a large heterogeneous mass arising from the sacrococcygeal area with the rectum pushed anteriorly (Fig. 2).



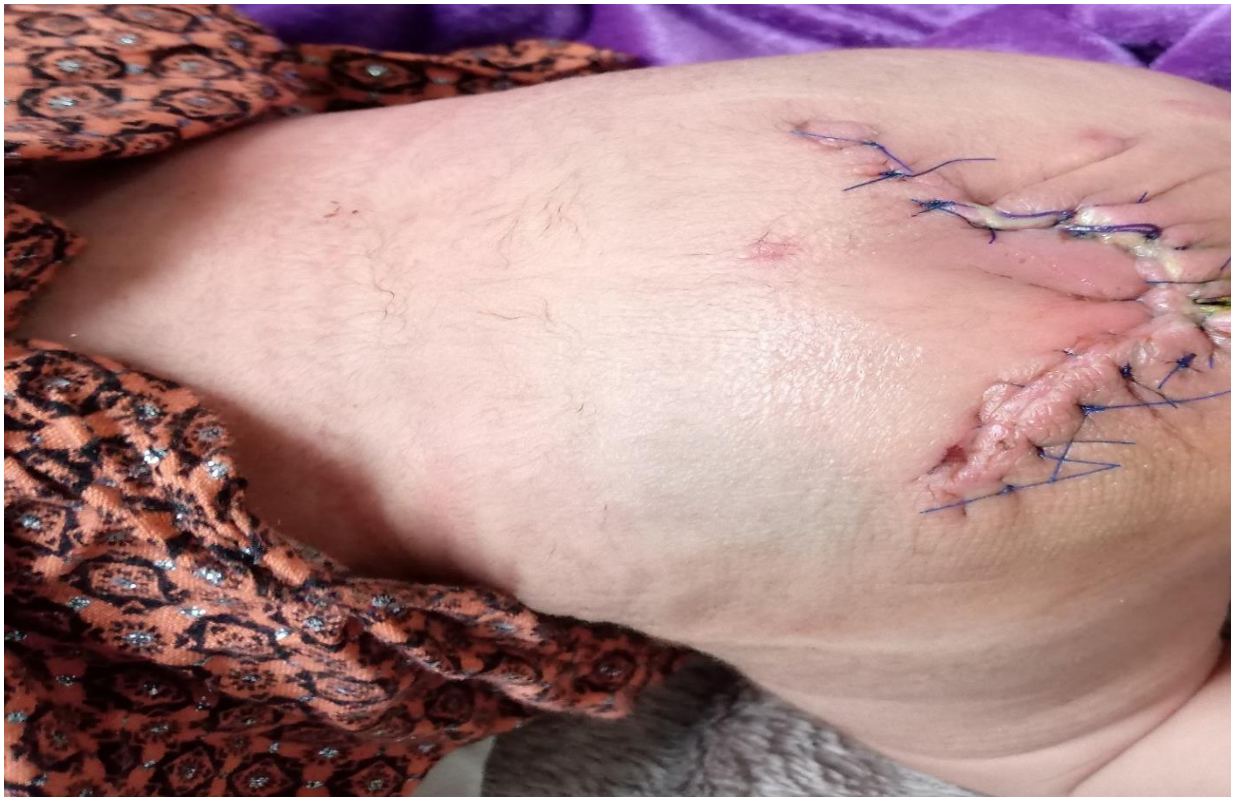


There was no spinal dysraphism. Preoperatively she was planned with full bowel preparation and consent for possible colostomy. She underwent complete excision of the mass by the post

anal route along with the lower sacrum with no intraoperative or postoperative problems (Fig. 3).



Intra operatively there was no infiltration into the pelvic structures. The sutures were removed on the 10th day and she was discharged (Fig. 4).



There was no postoperatively bowel or bladder incontinence.

Discussion:

The newborn with SCT has an excellent prognosis depending on the time of diagnosis, malignant potential of the tumor and the ease of surgical excision.

Early prenatal diagnosis is possible. SCT could be diagnosed from second trimester of pregnancy when there is polyhydraminos and/ or uterus larger than the gestational age. Prenatal diagnosis is of significance, since early prenatal presentation is associated with high fetal morbidity and mortality while presentation after 30 weeks is a relatively good prognostic indicator for fetal survival.

Monitoring for fetal distress during pregnancy is very important. Some large tumors have a very high blood flow that causes a shift in blood flow away from the baby towards the tumor. As it grows it can cause the baby to become sick and hydropic. This means the heart begins to fail and the baby becomes swollen. Other possible complications include bleeding inside the tumor, development of excess amniotic fluid and preterm labour. Progressive hydrops can be associated with a swollen and sick placenta. There is a rare condition called 'Mirror syndrome', where the mother mirrors the baby's sickness.

This is due to fluid retention in fetal compartments, water retention in mother also occurs and she suffers the same symptoms as the sick fetus. Mother will become ill and have signs of preeclampsia, water retention, and high blood pressure, protein in the urine, placetomegally and failing heart. If this occurs baby should be delivered immediately. Early diagnosis may necessitate delivery by cesarean section in centers with good neonatal facilities where early surgical treatment can be offered to the baby.14

That's what happened with our case due to prenatal diagnosis baby delivered safely with cesarean section and also surgical excision was done at earliest. Similarly fetal surgical procedures15 could be undertaken when the diagnosis is made early in the pregnancy. Well planned surgical excision including coccyx excision was done in order to forestall possible recurrence. Anesthetic challenges were mainly prematurity and blood loss. Atracurium was used for muscle relaxation considering prematurity of organs. Blood loss was replaced with blood (packed cell volume) immediately. Adequate pain relief was provided. Complete excision including coccygectomy is the primary therapy for all SCT and it is adequate if the tumor is benign. Altman type 1 SCT, meticulous anesthetic and surgical management, good postoperative monitoring and care in neonatal intensive care unit and benign nature of the tumor all these things contributed to the miraculous outcome of the baby. Extensive surgery in the pelvis and perineal region may involve disruption of nerves and muscles which supply urinary/anorectal sphincters and provide maximum support in normal working respectively. Alpha fetoprotein well known marker of teratomas, is valuable in differentiating between mature and malignant teratomas at presentation and during follow up of patients.

It may be utilized to detect early occurrence of malignancy. This is not only appropriate during first three postoperative years when recurrence is likely, but on long term basis as significant number of them suffer from deficient anorectal function and diminished quality of life.

SCT are tumors that arise in the sacrococcygeal area containing tissue from all three germ layers. SCT has a incidence of approximately one in 35,000 – 40,000 live births with malignant transformation with increasing age1,2.

SCT is more common in females with a male, female ratio of 1:3-4,3, 5.

These tumors are thought to arise in the Henson's node which contains pleuri potent cells and may present prenatally, at birth or later in life6, 7.

Antenatal detection of SCT by use of three-dimensional (3D) sonography has a worse outcome than those diagnosed after birth with survival rates ranging from 54-77%7, 8, 9.

Large tumors detected antenatally or those with excess amniotic fluid then early caesarean section must be planned. These tumors may present with varying symptoms like bowel or bladder incontinence, backache, weakness of limbs or fistula of the urogenital or gastrointestinal tracts. MRI shows the tumors relation with the adjacent organs with infiltration suggesting malignant transformation. Other congenital anomalies like defects in the cloacal and hindgut are associated in 10-24% of cases10.

Complete excision including the coccyx and sparing the sacral nerves leads to low recurrence. The recurrence is high in the first three years after surgery and therefore needs regular followup11.

Those tumors with malignant changes need further chemo radiotherapy.

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