CASE REPORT

Unusual Sacrococcygeal Teratoma (SCT) With Accessory Penis, Anorectal Malformation, Bifid Scrotum, Bilateral Inguinal Hernias and Hypospadias

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ABSTRACT

SCT are congenital tumors originating from all three germinal layers: ectoderm, mesoderm and endoderm. The reported incidence is one in 35000-40000 live births. We are reporting an unusual case with sacrococcygeal teratoma (SCT) with accessory penis, anorectal malformation, bifid scrotum, hypospadias and B/L inguinal hernias. A 13*13 cm mass was excised with os coccyx and histopathology confirmed it teratomatous growth.

Keywords: SCT, bilateral inguinal hernia, hypospadias

INTRODUCTION

SCT are rare congenital tumors, generally discovered at birth[1]. These originate from one or more of the three germinal layers. These are found in neonates, infants and children upto 4 years[2]. Teratomas are seen in ovaries, testes, sacrococcygeal region and retroperitoneal space[3]. These can be benign or malignant. Histopathology is usually needed for confirmation of diagnosis. A grading system based on histology divides teratomas into 2 types: mature (grade 0) and immature teratoma (grade 1,2,3). Mature teratoma is characterized by well-differentiated tissues of all the three germinal layers. On the other hand immature teratoma has elements of immature neuroectodermal tissue along with other germinal layers[4]. They usually arise in midline and para-axial region[5]. Any organ can develop from the three germinal layers of teratoma i.e. hair, bones, teeth, limbs, penis and so on. Only one case has been reported with an unusual spinal teratoma with an accessory penis on back[6].

We are reporting an unusual sacrococcygeal teratoma (SCT) with accessory penis, anorectal malformation, bifid scrotum, distal penile hypospadias and bilateral inguinal hernias. To our knowledge, this will be the first such case reported in English literature.

CASE REPORT

A one day male neonate weighing 4 kg was brought to the emergency with a huge swelling over the sacrococcygeal area with an unusual accessory penis and scrotum over it. Patient also had anorectal malformation, bifid scrotum, bilateral inguinal hernia and distal penile hypospedias.

Patient was healthy-looking infant with stable vital signs. There were no signs of cardio-respiratory distress. His abdomen was moderately distended with absent anal opening, so Digital Rectal Examination was not possible to assess the intrapelvic extent of the mass. The full-term neonate was born at home by uneventful spontaneous vaginal delivery with assistance of traditional birth attendant. Child cried well immediately after birth.

Initial abdominal examination revealed moderate abdominal distension with some signs of dehydration. Base line laboratory studies were normal. Cross-table X-ray abdomen showed a high variety anorectal malformation and a soft tissue shadow in sacrococcygeal area with diffuse calcifications. Ultrasound of mass revealed a mixed echogenicity mass of 10*12cm over sacrococcygeal area with no evidence of intrapelvic extension (type I). CT scan of abdomen and pelvis showed a soft-tissue mass at sacrococcygeal region having predominantly fat component and calcific foci suggestive of teratomatous growth. A hyperdense band-like structure is seen within the mass. Penis having corpora is seen at the top of mass along with rudimentary scrotum but no testicles identified. Serum AFP and beta HCG were not done because of financial constrains.

Our pre-operative suspicion was sacrococcygeal teratoma with accessory penis, bilateral inguinal hernias, anorectal malformation, bifid scrotum and distal penile hypospadias or sacrococcygeal teratoma with asymmetrical twins.

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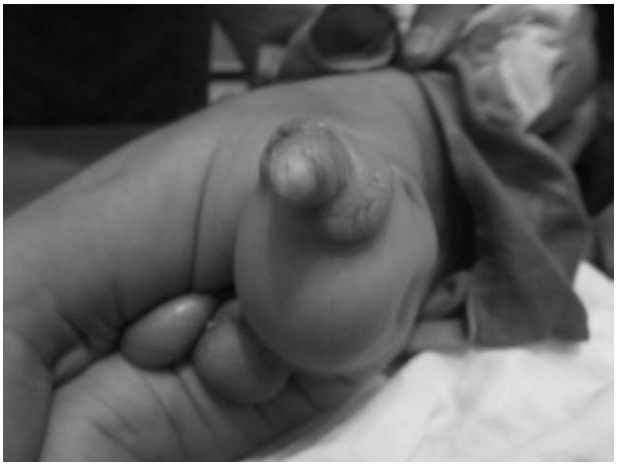
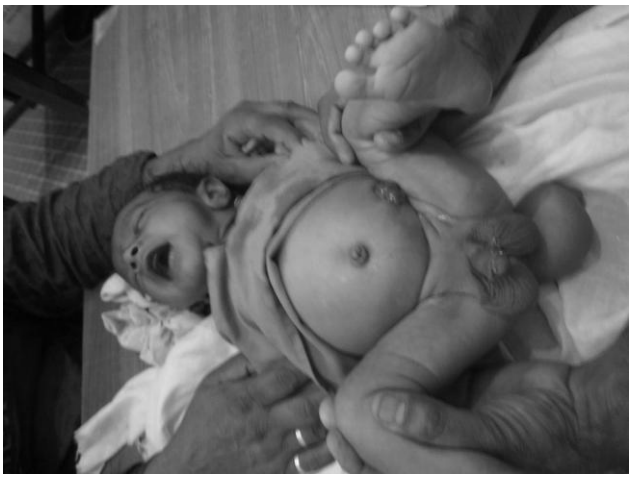
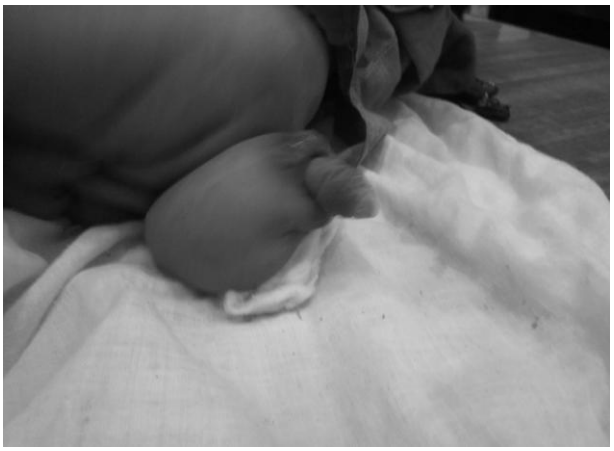
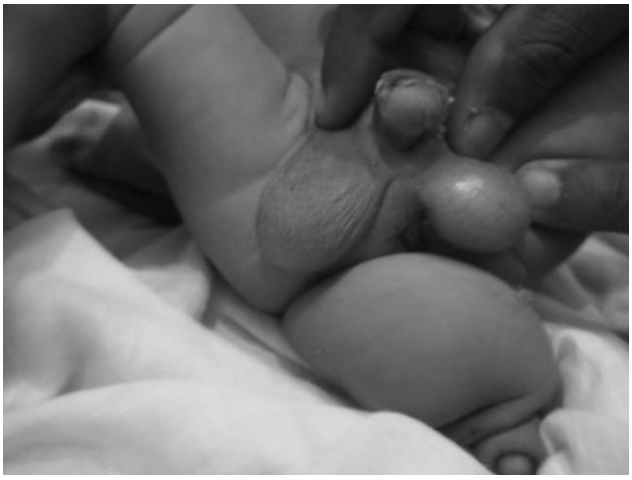
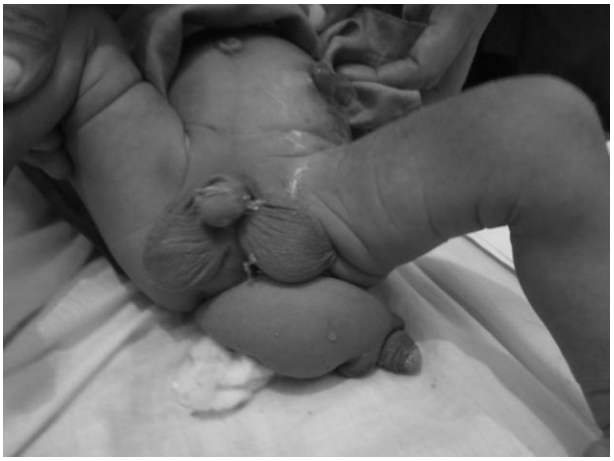
The child was taken up for surgery and a diverting sigmoid colostomy was made in first stage. After uneventful recovery from first surgery his second surgery was done after 15 days of first surgery and excision of sacrococcygeal teratoma along with accessory penis was done. The specimen was sent for histopathology which confirmed a mature variety of teratoma with clear resection margins. Patient made uneventful recovery after second surgery also and sent home on 10th post-operative day. The attendants were explained about follow up and future surgeries (PSARP, colostomy reversal, bilateral herniotomy and hypospadias repair).

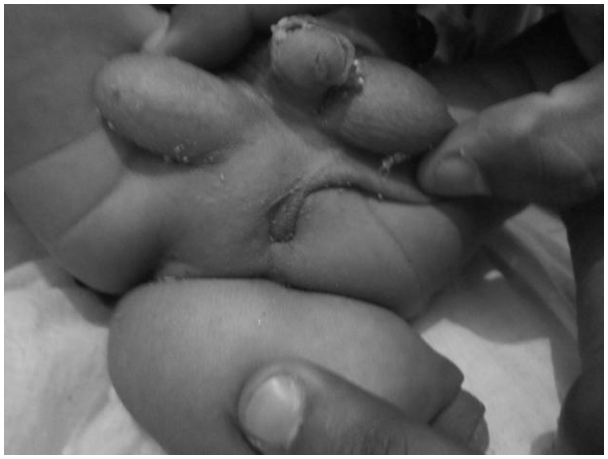


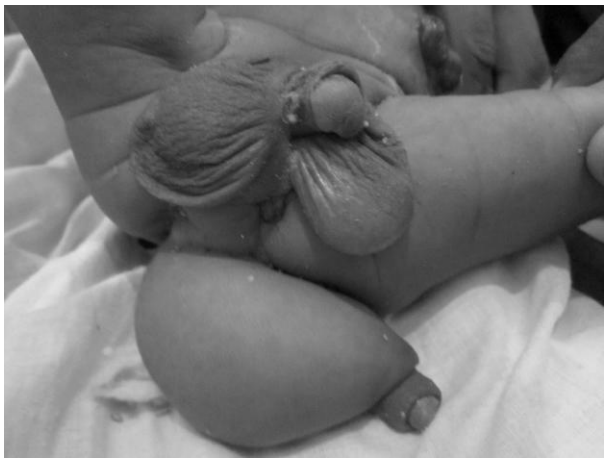












DISCUSSION

SCT are rare congenital tumors, generally discovered at birth[1]. These originate from one or more of the three germinal layers. These are found in neonates, infants and children upto 4 years[2]. Sacrococcygeal teratomas are seen in ovaries, testes, sacrococcygeal region and retroperitoneal space[3]. These can be benign or malignant. Histopathology is usually needed for confirmation of diagnosis. A grading system based on histology divides teratomas into 2 types: mature (grade 0) and immature teratoma (grade 1,2,3). Mature teratoma is characterized by well-differentiated tissues of all the three germinal layers. On the other hand immature teratoma has elements of immature neuroectodermal tissue along with other germinal layers[4]. They usually arise in midline and para-axial region[5]. Any organ can develop from the three germinal layers of teratoma i.e. hair, bones, teeth, limbs, penis and so on. Only one case has been reported with an unusual spinal teratoma with an accessory penis on back[6].

Teratomas not only present over body surface but can also arise from any intra-abdominal organ. More than 100 cases of gastric teratoma have been reported[7,8]. SCT are usually diagnosed in 2nd trimester via ultrasonography. SCT is associated with polyhydroamnios and/or uterus larger than gestational age. Early prenatal diagnosis before 30th gestational week is associated with greater morbidity and mortality for both neonate and mother. It can lead to maternal and child mortality due to obstetric complications namely obstructed labour, prolonged labour, distocia and damage to maternal birth canal[9].

As in most congenital abnormalities, parents of these patients are psychologically disturbed and a good counseling about disease and results of complete surgical excision can decrease their anxiety to some extent. A well planned surgical excision including pre-operative skin markings to enable flap reconstruction of defect has excellent prognosis. Important prognostic factors are age at diagnosis and treatment, extent of resection, histologic type and stage at time of resection. Complete surgical excision is mainstay of treatment, however chemo- and radiotherapy are necessary in malignant cases[10].

After complete excision prognosis is excellent however a long term follow up for surveillance of recurrence is necessary.

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