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ORIGINAL ARTICLE

OUTCOME OF SURGICAL TREATMENT FOR NEONATAL SACROCOCCYGEAL TERATOMA: EXPERIENCE WITH 13 CASES

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Abstract

Background and Objective: Sacrococcygeal teratomas (SCT) are the most common congenital tumors, occurring in 1/35000 live births. It presents a major neonatal surgical problem. The objective was to revise and describe our experience as regard diagnosis, operative management and finding, postoperative care as well as surgical outcome in neonates with SCT in our locality.

Patients and Methods: Thirteen patients with SCT were treated surgically by one surgeon during the period from October 2005 to December 2011. All cases subjected to surgical treatment with the same protocol of preoperative evaluation and follow-up protocol. Perioperative data, clinical presentation, intraoperative findings, tumor characteristics, postoperative complications and outcome of surgical treatment all were reported and analyzed.

Results: The mean birth weight was 3093.75±822.2 gm; mean gestational age was 37.54±2.18 weeks and mean age at time of operation was 10.92±14.87 days. There were 3 (23.1%) males and 10 (76.9%) females. Prenatal diagnosis made in one (7.7%) case and diagnosis was mainly clinical. Two cases had associated with congenital anomalies. The approach was sacrococcygeal in 12 (92.3%) cases and it was combined with abdominal approach in one (7.7%) case. Mean weight of the excised mass was 573.85±343.15 gm. Postoperative complications occurred in the form of wound infection in 3 (23.1%) cases, Poor cosmetic outcomes in 4 (30.8%) cases and clinically weak anal tone with manifest soiling in 2 (15.4 %) cases.

Conclusion: Sacrococcygeal teratoma presents a major neonatal surgical problem. A good prognosis is related to several factors. Adequate surgical treatment which includes complete excision at neonatal period with coccygectomy carries good prognosis and remains the mainstay of treatment.

Keywords: Sacrococcygeal teratoma.

INTRODUCTION

Although generally a rare condition, sacrococcygeal teratomas (SCT) are the most common tumors diagnosed among newborns.[1] It has a reported incidence of approximately one in 35,000 – 40,000 live births.[2,3] It is more common in females, with a male: female ratio of about 1: 3–4.[4,5] There are many conflicting theories related to the origin of SCT. They are composed of two or three germ cell layers; have multiple tissue types, and can present in various sizes and shapes.[6,7] Believed to have arisen from the

totipotent somatic cells that originate from the primitive knot (Hensen's node), they are usually attached to the coccyx and escape normal inductive influences.[8]

In contrast to the newborn with SCT, the fetus with SCT remains at high risk of perinatal complications and death.[9] Fetuses with SCT detected antenatally have three times mortality rate compared with postnatally diagnosed neonates.[10] Neonatal death may result from maternal obstetric complications of tumor rupture, preterm labor, or dystocia.[11,12] The fetus is also at risk of high output cardiac failure, placentomegally and hydrops.[13]

The postnatal outcomes are related to surgical treatment and the histological features of the tumor.[1]

SCT are commonly diagnosed in the prenatal period using three-dimensional sonography.[14-17] Prenatal MRI offers superior anatomical evaluation regardless of fetal orientation.[18-21] which is critical for counseling the parents and planning surgical options including fetal surgery.[22-25] Hydrops and prematurity are the two main factors that contribute to mortality.

Postpartum morbidity associated with SCT is attributable to associated congenital anomalies, mass effects of the tumor, recurrence, intraoperative and postoperative complications. Ten to twenty-four percent of SCT are associated with other congenital anomalies.[26,27]

Tumor classified as benign (mature) which is more common and malignant or immature (composed of embryonic elements).[28] Surgical Section of the American Academy of Pediatrics, Altman and associates report morphological classification system: [5] Type I (45.8%): predominantly external; Type II (34%): have both an external mass and significant presacral pelvic extension. Type III (8.6%): visible externally, but the predominant mass is pelvic and intraabdominal. Type IV (9.6%): not visible externally but entirely presacral.

Surgical resection remains the mainstay of therapy, with an excellent prognosis provided it is early and complete. Recurrence is rare following complete excision.[29] The overall survival varies according to several factors.[30] Long-term follow-up study found that functional symptoms tended to improve with time.[31]

PATIENTS AND METHODS

A prospective study of thirteen patients with SCT was carried out at Pediatric Surgical Unit Sohag University Hospitals, local health insurance hospital and private sector during the period from October 2005 to December 2011.

Detailed history regarding antenatal period, type and place of delivery were reported. All patients subjected to physical examination included rectal examination to evaluate evidence of intrapelvic extension. Laboratory studies included routine one and serum alpha fetoprotein (AFP) in all cases. Imaging studies incorporated ultrasound, plain lumbosacral radiograph, CT and/or MRI abdomen and pelvis to differentiate SCT from neural tube defects delineate the tumor and evaluate anatomic features. Malignant cases which presented with distant metastases referred and treated by an oncologist and excluded from our series.

As routine, preoperative antibiotics were administered, any laboratory abnormalities were corrected and cross-matched blood unite for transfusion was ready. After an informed consent, surgical excision was carried out electively as early as possible.

Under general anesthesia with relaxant technique intravenous line was secured and patient hydrated well. Patient positioned prone and complete excision of the mass with enbloc coccygectomy was carried out through a chevron-shaped buttock incision alone or with additional approach through the abdomen in cases with pelvic extension. Muscles of the rectal sphincter were preserved and reattachment of the small muscles and ligaments formerly attached to the coccyx were performed. I would like to underline technical details which play an important role in improving the outcome and achieving low morbidity rates namely enbloc coccygectomy, sphincter muscle preservation and reattachment of the small muscles.

Demographic data, clinical presentations, investigatory workup, operative details, tumor characteristics and surgical outcomes all were documented. Follow up was achieved via clinical evaluation and serum AFP every 3 months. Regarding clinical evaluation of anal tone status a through history is obtained with special attention to frequency of bowel movements. Complete physical examination focused on inspection of the perineum and external anus for radial skin creases and patulous anus. Perianal sensation is assessed and presence of the anocutaneous reflex suggests an intact reflex arc. Rectal examination occasionally performed to evaluate sphincter resting tone and fecal impaction. Numerical data are expressed in the form of mean ± standard deviation using the appropriate statistical method.

RESULTS

Thirteen patients with SCT were treated surgically in this series. They were 3 (23.1%) males and 10 (76.9%) females. Birth weight was reported in eight cases and it ranged between 2150-4600 gm (mean 3093.75±822.2 gm).The mean gestational ages was 37.54±2.18 weeks (range 34-41 weeks). Mean age at time of operation was 10.92±14.87 days (4-60 days). Delivery was vaginally in 7 (53.8%) cases and by Caesarian section in 6 (46.2%) cases. Two cases (15.4%) had associated congenital anomalies (case with congenital heart disease CHD and one case with palpable undescended testis PUDT).

Prenatal diagnosis was made in one (7.7%) case via antenatal ultrasonography in which the mass and

polyhydramnios were detected. The antenatal period however was uneventful in all cases. In neonates and infants the diagnosis was mainly clinical all patients were presented by their parents with a mass at the lower back in the sacrococcygeal region since birth (Fig. 1).

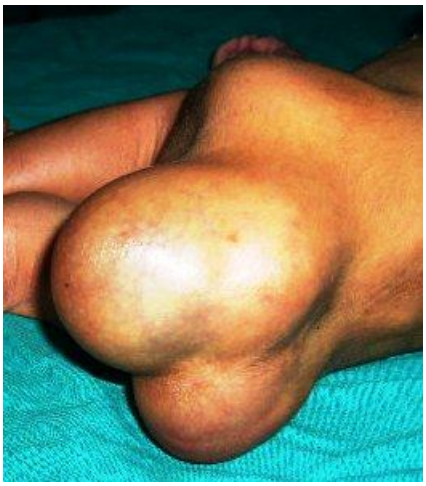

Fig 1. SCT.

Masses had lobulated surface and variable consistency it was solid in 4 cases, cystic in 2 cases and heterogeneous in 7 cases. Systemic examination was normal in 11 (84.6%) case, the remaining two cases showed clinical manifestation of CHD and UDT. Anus was displaced anteriorly in 10 (76.9%) cases (Fig. 2). Soft tissue US of the swelling was applied to all cases and revealed mixed echogenicity. Also, abdominal US was performed in all cases and showed intrapelvic extension in 2 (15.4%) cases. Serum AFP level was elevated in all cases compared to age reference range, then it decreased to normal levels within 6-10 weeks postoperatively.

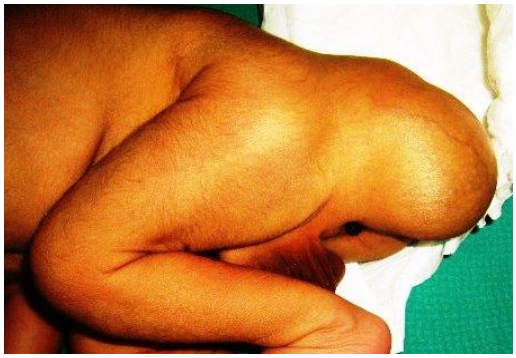

Fig 2. Anteriorly displaced anus.

According to Altman's classification there were 9 (69.2%) cases type I, three (23%) cases type II and one (7.8%) type III. By both clinical and sonographic finding the tumour was mixed in 7 (53.8%) cases, solid in 4 (30.8%) cases and cystic in 2 (15.4%) cases (Fig. 3).

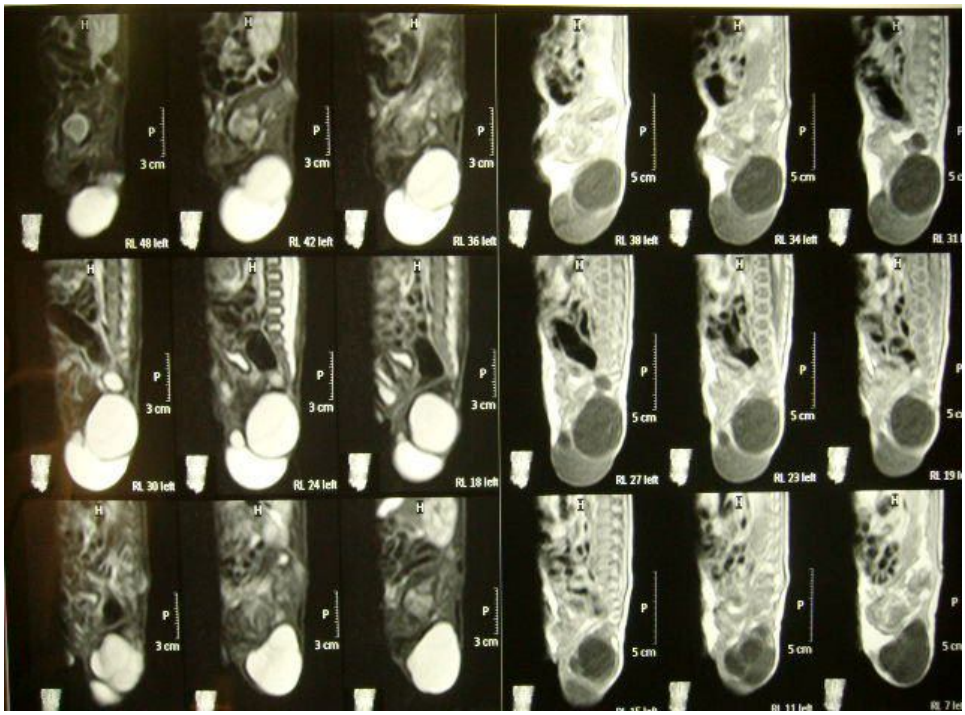

Fig 3. MRI.

Surgical treatment in the form of complete excision of the tumour with coccygectomy, in order to anticipate possible recurrence, had been carried out in all patients. The approach was sacrococcygeal with Chevron incision in 12 (92.3%) cases and it was combined with abdominal approach in one (7.7%) case. Intra-operative blood transfusion was required in 3 (23%) cases (Figs. 4,5). The mean operative time was 90±16.71 minutes (range 70-115 min). By calculating the difference between preoperative and postoperative weight of the neonate, the mean weight of the excised mass was 573.85±343.15 gm (range 85-1065 gm). Histopathological examination and analysis of the excised mass revealed mature SCT (differentiated or benign) in 11 (84.6 %) cases and immature teratoma in 2 (15.4 %) cases which showed immature tissue in some parts but with no evidence of malignant components (Fig. 6).

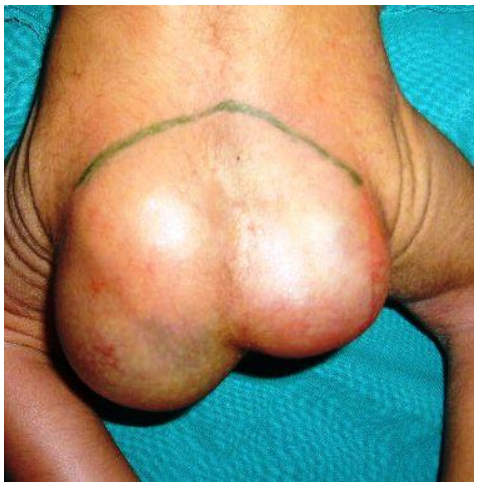

Fig 4. Chevron incision.

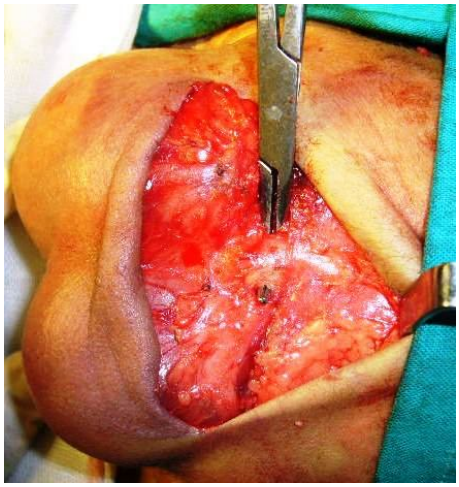

Fig 5. Coccygectomy.

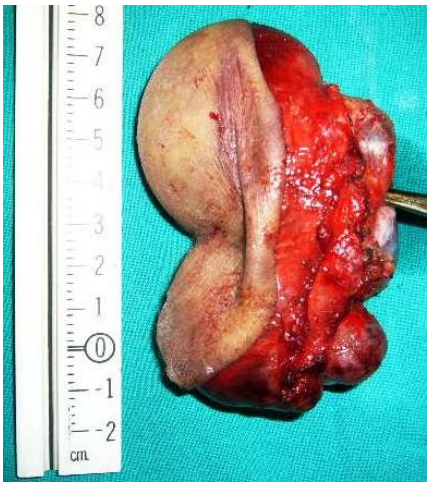

Fig 6. Excised mass.

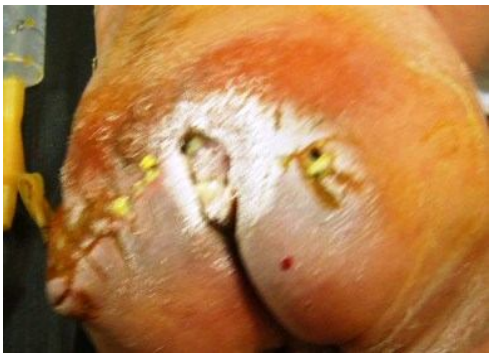

Fig 7. Wound infection with skin disruption.

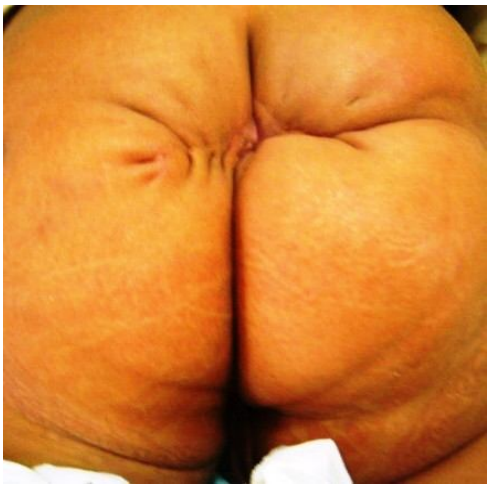

Fig 8. Cosmetically unsatisfactory scar.

Regarding postoperative complications wound infection was reported in 3 (23.1%) cases and skin disruption occurred in two of them. These cases were successfully managed by controlling the infection and left to heal with secondary intention (Fig. 7).

From a cosmetic point of view, the surgical results were satisfactory in 9 (69.2%) cases. Poor cosmetic outcomes were reported in 4 (30.8%) cases in the form of dimples or disfiguring scars (Fig. 8).

Normal anal tone was preserved in 11 (84.6%) cases while clinically weak anal tone with manifest soiling was noted in 2 (15.4%) cases. There was no clinical or biochemical evidence of recurrence during the follow up period which ranged from 45 days to12 months.

DISCUSSION

Sacrococcygeal teratoma is the commonest variety of tumors encountered in neonates and most commonly found in females.[32] Perrelli et al in 2002, in his case series reported that female to male ratio was 2.4:1 with mean birth weight was 2916 gm, mean gestational age was 35 weeks and mean age at time of surgery was 16.3 days.[1] Kamal et al 2006, documented female sex predominance with ratio 4:1, mean birth weight of the studied cases was 3137.27±781.74 gm, gestational age ranged between 34-42 week and the mean age at time of surgery was 27.6±17 days. Vaginal delivery reported in 26.7% of his cases and associated anomalies presented in 20% of cases. All data were comparable with our results.[33]

Prenatal diagnosis ranged between 12% and 29% in many series.1,[34] Our results showed antenatal diagnosis only in 7.7% of cases which may be due to lack of sufficient antenatal care services and knowledge in our locality.

Gabra et al and Makin et al reported that the most common presentation in neonates is a large, predominantly benign tumor protruding from the sacral area that is noted at the time of delivery.[27,35] The tumor may be solid, cystic or both in nature.[12,33] Serum AFP is a useful tumor marker with elevated level in most cases and decreased after complete resection.[12,34]

According to Altman's classification the percentage of distribution widely varied among many reports. Osman et al in 2012, documented in his case series type I was presented in (22%) of cases, type II (49%), type III (27%) and type IV in (2%) of cases. Other reports described type I in (80%) of cases, type II (13%), type III (7%) and no cases belong to type IV.[33] Our results showed that (69.2%) of cases was related to type I, type II (23%), type III (7.8%) and no cases with type IV SCT.

Complete excision including coccygectomy, through a chevron-shaped buttock incision with preservation of rectal sphincteric muscles, is the primary therapy for all SCT.[36] The coccyx always should be resected enbloc with the tumor, as failure to do so results in a 35-40% recurrence rate.[37] The preferred approach is through the perineum; a large SCT may require an additional approach through the abdomen. Surgery should perform within the first few days of life and Include reattachment of the small muscles formerly attached to the coccyx.[38-41]

In this study combined sacrococcygeal with abdominal approach used only in one (7.7%) case.

Perrelli et al in 2002, had mean weight of the excised mass 548 gm (range 30±1380 gm). [1]

Histologically, in our case series immature SCT was presented only in 15.4% of cases and no malignant teratoma was documented which may be attributed to early excision. Our results slightly varies from other studies in which immature teratoma represented in about 23% of cases and malignant type ranged between 12% and 29% of cases.[34,42] Before the age of one month, the risk of SCT being malignant in only 5%, by one year 60% and in children older than one year it is about 75%.[43]

Unsatisfactory cosmetic appearance was the commonest complication and occurred in 30.8% of cases, followed by wound infection which occurred in 23.1% (two cases out of 3 cases with wound infection experienced wound disruption). Clinically weak anal tone with fecal soiling occurred in 15.4% of our cases. In one review of 25 patients the most frequent complication was an unsatisfactory appearance of the surgical scar.[44] Wound infection reported in (9.7%).[34] However, later complications of surgery may include neurogenic bladder, other forms of urinary incontinence or fecal incontinence.[45]

In our series there was no recurrence during follow up period which may be attributed to complete excision with coccygectomy in all cases at early age and benign type of these tumors.

Local recurrence is rare unless the coccyx has been spared.[46] Other stated that a recurrence as high as 37% has been reported if the coccyx is not removed in the primary surgery.[47,48] Other reports documented recurrence in 12% of cases.[34]

In conclusion Sacrococcygeal teratoma is a well-known tumor of the newborn, mostly benign and common in females. Early diagnosis and management minimize the risks.

Adequate surgical treatment which includes complete excision at neonatal period with coccygectomy carries good prognosis and remains the mainstay of treatment. Complete and early excision guard against malignant transformation. On the other hand, coccygectomy markedly reduce recurrence rate. However, longer period of follow-up is required, in order to further evaluation of functional outcomes.

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