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OUTCOME OF NEONATAL SACRO-COCCYGEAL TERATOMA AT A TERTIARY CARE CENTRE

Surgery

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ABSTRACT

Aim: To analyze the clinical characteristics and outcome of neonatal Sacrococcygeal teratoma (SCT) treated at our centre.

Materials and Methods: A retrospective study was conducted on 17 neonates, who were treated for SCT from January, 2013 to December, 2017. Hospital records of these patients including the clinical features, surgical details and follow-up details were retrieved and data thus collected were analyzed. Patients who did not come for follow-up were contacted telephonically and called for periodic checkups.

Results: Out of 17 patients, 12 girls and 5 boys presented with a mean age of 11 days (range 1- 28 days).One of the patients presented at 28 days after birth in profound sepsis with a fungating ruptured sacrococcygeal mass which led to her demise on the day of her presentation. Remaining 16 patients were operated in the neonatal period. Only 5 of these patients had antenatal ultrasound detection. Two patients presented with tumor rupture. 13 patients had a large tumor size with diameter exceeding 10cm. One patient with a small tumor was misdiagnosed as meningomyelocoele. While 15 patients were Altman Type I, 2 patients had Altman Type II tumor. All the 16 surviving patients underwent complete surgical excision with coccygectomy. Histopathological examination showed mature cystic teratoma in 14 patients and immature elements in 2 patients. Following surgery, wound infection was seen in 3 patients and one patient had neurogenic bladder. No mortality was seen in the 16 patients who were operated. Mean follow-up was 30 months (range – 6months to 36months) in twelve patients. 4 patients were lost to follow-up after surgery.

Conclusions: Complete surgical excision of Neonatal Sacro-coccygeal teraroma is usually possible, considering the early presentation of Altman Type I and II. Histopathology mostly shows mature elements showing good prognosis. Considering the long term risks of recurrence, periodic follow-up with clinical and tumor marker assessment is a must to detect recurrences early and intervene promptly.

KEYWORDS

Neonates; Prognosis; Recurrence; Sacrococcygeal teratoma

INTRODUCTION

With an incidence of 1/40,000 live births, Sacro-coccygeal teratoma (SCT) is the commonest congenital tumor in neonates [1]. Depending upon the relative sizes of the external and intra-pelvic/intra-abdominal components of the tumor, 4 types of SCT have been described by Altman [1]. Most neonatal SCT are detected soon after birth due to large external tumor (Altman Types I and II) leading to early surgery with good prognosis. Types III and IV usually present late and have poorer prognosis due to increased likelihood of finding immature elements due to late presentation [1]. Being known for recurrences, follow-up of SCT patients is very important. We present our experience of 17 neonatal SCT patients.

MATERIALS AND METHODS

A retrospective study was conducted on 17 neonates who were treated for SCT from January, 2013 to December, 2017 in a tertiary care hospital. Hospital records of these patients including the clinical features, surgical details and follow-up details were retrieved and data thus collected were analysed. Patients who did not come for follow-up were contacted telephonically and called for periodic checkups.

RESULTS

(1) Demographic details: **Table 1** summarizes the demographic details of the study.

Table 1: Demographic Details

Total No. of patients in the study	17
Male: Female ratio	5:12
Mean age at the time of presentation	11 days(range 1 -28 days)
Antenatal detection on USG	5 patients
LSCS: Normal vaginal delivery	5:12
Preterm : Term delivery	2: 15
No. of patients operated	16
No. of Deaths in the study	1
Mean Follow-up	30 months (range – 6 months to 36 months)
No. of Patients lost to Follow-up	4

USG: Ultrasound

A total of 17 SCT patients in the neonatal age group were seen in the study of which 12 were girls and 5 boys. Mean age at presentation was 11 days with a wide range of age at presentation from 1 day to 28 days. Of these 17 patients, only 5 had antenatal detection on third trimester Ultrasound (USG) although 14 of these children had hospital delivery. Five neonates were born by lower segment Caesarean section (LSCS) and the remaining patients had had normal vaginal delivery. Two patients had preterm delivery. The mean weight of the babies at presentation was 2790g (range 2030 – 3750g).

While all the 17 patients had a lower back mass at birth, one of them presented with rupture after delivery with bleeding and had to be operated in the emergency operation theatre. In another patient, a small ruptured mass with leakage of clear fluid led to an improper diagnosis of ruptured meningo-myelocoele in the preoperative period. One patient presented at 28 days after birth in profound sepsis with a fungating ruptured sacrococcygeal mass which led to her demise on the day of her presentation.

In the rest 16 patients, after initial evaluation and routine investigations, complete excision of the mass with coccygiectomy was done. 4 of these patients are lost to follow-up while the remaining patients have a mean follow-up of 30 months.

(2) Tumor details: Table 2 depicts the type of the tumor in the SCT patients included in this study. 13/17 patients had a large tumor exceeding 10cm size. Most of the tumors (15/17) were of Altman Type I variety while two patients had Altman Type II tumor. Of the 16 patients whose tumors were excised and sent for Histopathological examination (HPE), 14 had mature elements while the remaining two patients showed immature elements in their tumor.

Table 2: Type of SCT encountered in the study

Type of Tumor	Description
No. of patients with a large tumor (size>10cm)	13

Altman Type	Type I – 15; Type II - 2
Histopathology (in 16 operated patients)	Mature cystic teratoma – 14; Immature teratoma - 2

One of these patients with immature elements on HPE, had a recurrence at 13 months of age which was detected by a raised Serum Alpha fetoprotein level and a palpable mass on per-rectal examination posteriorly.

(3) Table 3 depicts the complications seen during the management of these neonates.

Table 3: Complications

Complications	Frequency
Wound infection	**3** (managed conservatively)
Poor cosmesis/ Bad scar	**5**
Neurogenic bladder	**1** (Crede's manoeuvre initially followed by vesicostomy at 2 months of age and vesicostomy closure at 3 years of age; now on Clean intermittent catheterization)
Increased Stool Frequency	**2** (conservatively managed)
Death	**1 ; No mortality in any operated patient**
Recurrence	**1** (At 13 months of age with raised Serum AFP level and a mass on per-rectal examination posteriorly; received 4 cycles of Chemotherapy (Bleomycin, Etoposide, Cisplatin) followed by resection and normalization of Serum AFP and two subsequent cycles of chemotherapy; Patient asymptomatic on follow-up at present.

AFP- Alpha feto protein

Of all the 16 patients who were operated for SCT, 3 had wound infection which was treated conservatively; 5 patients had a very bad scar and their parents seemed concerned about the appearance of these scars. One child had features of neurogenic bladder following surgery which was initially managed by Crede's manoeuvre of bladder evacuation but because it was not very effective, Blocksom's vesicostomy had to be done to ensure proper bladder evacuation and protection of the upper renal tract. At 3 years of age just before going to school, vesicostomy closure was done following which child was managed on clean intermittent catheterization (CIC) during daytime and continuous night-time drainage.

One patient, who had immature elements on HPE at the time of initial surgery, presented with raised Serum alpha-fetoprotein (AFP) level for her age indicating recurrence and on per-rectal examination, she was found to have a palpable mass posteriorly. After 4 weeks of chemotherapy using Bleomcin, Etoposide, Cisplatin(BEP), AFP level normalized and mass lessened in size. It was then excised and repeat biopsy showed features of necrosed tissue with no viable tumor tissue. He has been given additional 2 cycles of BEP and is doing well on follow-up visits.

DISCUSSION

Being the commonest tumor seen in the neonatal age group, SCT presents unique challenges particularly in developing countries like India where there are deficiencies in antenatal detection, late presentation, mismanagement at inappropriate, unskilled centers and poor follow-up. Although several studies on SCT can be found in the western literature, studies from resource-challenged countries are fewer [2, 3]. This study summarizes our experience in managing neonatal SCT.

Similar to the observation of other researchers [1, 4 and 5], there was a preponderance of females in the study group with a female to male ratio of 12:5. No definite reason is known for this predilection.

According to the size of the tumor, Altman et al classified SCTs into small (2 to 5 cm diameter); moderate (5 to 10 cm diameter); large (>10 cm diameter) [1]. In our series, 13 out of 17 patients had a large tumor. The significance of tumor size lies in the vulnerability of larger tumors to injury, rupture and bleeding, increased incidence of pressure symptoms, difficulties during labour and increased likelihood of finding immature elements on HPE [1, 6]. Both tumors with immature histology in our study were large tumors and the tumor which had ruptured and bled following delivery had a diameter of 18cm.Tumor size also increases the chances of hydrops foetalis and is associated with additional difficulties during surgery including increased blood loss [6].

Finding Altman types I (15/17) and II (2/17) tumors in the frequency as observed in this study is similar to the observation of other researchers [7]. Also, there were no patients of Altman Type III or Type IV as expected in the neonatal age group.

Sacrococcygeal masses can have several differential diagnoses [8]. However, as evident in one of our patients, sometimes a Type I or II SCT may be confused with meningo-myelocoele (MMC). Although magnetic resonance imaging (MRI) scan could have helped in identifying the correct diagnoses, we did not do it due to difficulty in performing this scan in a neonate.

Gonzalez-Crussi et al. have described the histological grading of SCTs (Grades 0-3) based on the presence /absence of immature neural elements and their relative quantities [9]. Grade 0 being the most common accounts for around 75% of the cases, while grades (1-3) reflect different degrees of immaturity with fewer incidence [10]. Malignant lesions in neonatal age group have been reported in around 13.2% cases [10]. Our study showed benign grade 0 lesions to be found in 14/16 patients and immature elements in 2/16 patients. Although some authors believe that finding immature elements in the tumor does not correlate with overall prognosis of the tumor [5], we had the only recurrence in our study in a patient who had had immature elements in initial HPE.

Although associated malformations have been reported to occur in as many as 18% of patients of SCTs (most common being anorectal/ genital, vertebral anomalies) [4, 11and 12], we did not have any anomaly in any of our patients.

Poor antenatal detection of SCT on sonogram in our study (5/17 patients) reflects the sorry state of antenatal care and knowhow in a resource-challenged country like ours. This is, in contrast to the developed countries, where most of the cases are diagnosed by prenatal sonogram in the 24^{th} to 34^{th} week of gestation [11]. SCTs usually appear as well circumscribed exophytic masses at the caudal end of the fetus. Presence of placentomegaly, cardiomegaly or hydrops fetalis indicate poor prognosis [13, 14]. Fetal MRI may further add on to the anatomical details of the lesion, if needed [15].

Although it is recommended that patients with large SCT (>10cm diameter) and vascular tumors should be delivered by LSCS after fetal lung maturity has been attained [4, 16], in our study most of the children had normal delivery due to poor antenatal detection and care. This may explain the increased incidences of tumor rupture and bleeding due to trauma during delivery in developing nations. Tumor rupture has been reported to increase the chances of mortality of the patient [14].

Antenatally diagnosed SCTs have a high risk of perinatal complications and death. Evidence of placentomegaly, cardiomegaly and hydrops in such patients indicate increased likelihood of high output cardiac failure due to arterio-venous shunting through the tumor leading to fetal death [17]. Such patients need shunt reversal to save the patient which can be done by urgent tumor debulking either in the fetus or immediately after Caesarean section. Definitive tumor resection in such cases is done after the child stabilizes in the neonatal period. Adzick et. al. reported the first successful fetal SCT resection in 1997 in a fetus with a large SCT, polyhydramnios and impending hydrops fetalis [18]. Graf et. al. reported their observation of dissapearance of malignant foci and presence of mature histology in definitive surgical resection specimens of patients who had immature or malignant histology at the time of fetal tumor decompression [19]. The exact reason for this observation is not known. Other antenatal interventions include fetal cyst aspiration, amnioreduction and amniotic catheter drainage to lessen distension, avoid preterm labour and optimize physiology [20, 21].

All these developments signify the contrast between the developing and developed countries. While the former are still struggling with antenatal detection of SCTs, the later have started doing fetal intervention.

Complete excision of the tumor with coccygectomy within the first week of life is the recommended treatment. Because intra-operative bleeding may be significant, it is important to arrange cross matched blood sample beforehand. The most important factor to prevent recurrence is to ensure complete surgical excision of the tumor along with the coccyx. In cases where coccyx has not been removed, recurrence rates as high as 37% have been reported [9].

Sacral approach to Altman Types I and II SCT is the most common approach [22] and it is possible to perform complete excision through this approach. All the 16 neonates in our study were operated by the Chevron incision through the sacral route. None of our patients needed any additional abdominal incision to achieve complete excision of the tumor.

Several complications of surgery done for SCTs have been reported in literature [4], wound infection being the most common considering the excessive dissection and its proximity to the anus. Poor cosmesis of the healed surgical incision has been often reported [2, 4, 23 and 24]. Neuropathic bladder [5], temporary diarrhea [4], bowel incontinence and constipation [25] have been variously reported by different researchers. Neurogenic bladder and continence issues are more common in cases of large pelvic component necessitating pelvic dissection which causes increased likelihood of damage to the pelvic nerves. Wang et al., in a recent study, found significant anorectal functional sequelae following surgery for SCT [26].

In our study, we had wound infection in 3/16 patients who improved with conservative treatment.5 of our patient (31.25% of operated patients) had bad scar over their lower back after wound healing. We have concluded that good cosmesis after surgery for SCT is a challenge in the neonatal period due to lack of local tissues, large size of tumours, stretched out muscles, inadequate tissue for skin flaps and high chances of postoperative wound infection. However, once the child has grown up and there is no tumor recurrence, cosmetic surgery can be taken up to address this issue.

One of our patients developed neurogenic bladder and could not evacuate her bladder. Initially, Crede's manoeuvre of bladder evacuation was taught to her parents but when it was not very effective, we had to perform Blocksom's vesicostomy. Child did well thereafter and vesicostomy closure was done at the age of 3 years when she was about to join school. Following that, she was managed on clean intermittent catheterization and continuous night-time drainage.

Risk factors for malignancy in SCTs include a tumor size >10cm [27], Altman Types III and IV due to a delay in diagnosis, presence of solid areas within the tumor [4, 14]and delayed presentation beyond 2 months of life [1]. We had two patients with immature elements in their tumor one of which later recurred and had to receive chemotherapy and second surgery for tumor re-excision.

SCTs are known to recur even after complete excision and recurrence rates can be as high as 11-22% [22, 27]. So, it is very important to keep these patients on regular follow-up for clinical examination and serum AFP monitoring. Sometimes, increase in Serum AFP may be the first indicator for recurrence without any clinically detectable mass [27].

Reasons for high recurrence rates in SCTs are not very obvious. Incomplete excision, intra-operative spillage and non-performance of coccygectomy have all been suggested as a few possible causes [4, 9, 28 and 29].Malignant degeneration in mature teratoma has been postulated as one of the theories [29].

All 16 operated patients in our study have survived and one child who presented late died without surgery in sepsis. High survival rates have been reported by several other investigators [18].

CONCLUSION

All Sacro-coccygeal masses in neonates should be properly evaluated and promptly treated. Complete surgical excision with coccygectomy should be done in all cases and patients must be kept on regular long term follow-ups to detect recurrences early.

REFERENCES

1. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973. J Pediatr Surg. 1974; 9:389-98.
2. Aly KAE, Shoier M, Badrawy T. Sacrococcygeal Teratoma: a neonatal surgical problem. Ann Pediatr Surg. 2006; 2:106-11.
3. Wakhlu A, Misra S, Tandon RK, Wakhlu AK. Sacrococcygeal teratoma. Pediatr Surg Int. 2002; 18:384-7.
4. Ein SH, Adeyemi SD, Mancer K. Benign sacrococcygeal teratomas in infants and children: a 25 year review. Ann Surg. 1980; 191:382-4.
5. Keslar PJ, Buck JL, Suarez ES. Germ cell tumors of the sacrococcygeal region: radiologic-pathologic correlation. Radiographics. 1994; 14:607-20.
6. Noseworthy J, Lack EE, Kozakewich HPW, Vawter GF, Welch KJ. Sacrococcygeal germ cell tumors in childhood. J Pediatr Surg. 1981; 16:358-64.
7. Sinha S, Sarin YK, Deshpande VP.Neonatal Sacrococcygeal Teratoma:Our Experience with 10 cases. J Neonatal Surg.2013;2:4
8. Lemire RJ, Beckwith JB. Pathogenesis of congenital tumors and malformations of the sacrococcygeal region. Teratology. 1982; 25:201-13.
9. Gonzalez-Crussi F, Winkler RF, Mirkin DL. Sacrococcygeal teratoma in infant and children. Arch Pathol Lab Med. 1978; 102:420–5.
10. Valdiserri RO, Yunis EJ. Sacrococcygeal teratomas. Cancer. 1981; 48:217-21.
11. Tuladhar R, Patole SK, Whitehall JS. Sacrococcygeal teratoma in the perinatal period. Postgrad Med J. 2000; 76:754-9.
12. Lahdenne P, Heikinheimo M, Jääskeläinen J, Merikanto J, Heikkilä J, Siimes MA. Vertebral abnormalities associated with congenital sacrococcygeal teratomas. J Pediatr Orthop. 1991; 11:603–7.
13. Winderl LM, Silverman RK. Prenatal identification of a completely cystic internal sacrococcygeal teratoma (type IV). Ultrasound Obstet Gynecol. 1997; 9:425-8.
14. Holterman AX, Filiatrault D, Lallier M, Youssef S. The natural history of sacrococcygeal teratomas diagnosed through routine obstetric sonogram: a single institution experience. J Pediatr Surg. 1998; 33:899– 903.
15. Danzer E, Hubbard AM, Hedrick HL, Johnson MP, Wilson RD, Howell LJ, et al. Diagnosis and characterization of fetal sacrococcygeal teratoma with prenatal MRI. AJR Am J Roentgenol. 2006; 187:W350-6.
16. Kum CK, Wong YC, Prabhakaran K. Management of fetal sacrococcygeal teratoma. Ann Acad Med. 1993; 22:377–80.
17. Bond SJ, Harrison MR, Schmidt KG. Death due to high-output cardiac failure in fetal sacrococcygeal teratoma. J Pediatr Surg. 1990; 25:1287–91.
18. Adzick NS, Crombleholme TM, Morgan MA, Quinn TM. A rapidly growing fetal teratoma. Lancet. 1997; 349:538.
19. Graf JL, Housely HT, Albanese CT, Adzick NS, Harrison MR. A surprising histological evolution of preterm sacrococcygeal teratoma. J Pediatr Surg. 1998; 33:177-9.
20. Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, Johnson MP, Wilson RD, et al. Sacrococcygeal teratoma: Prenatal assessment, fetal intervention and outcome. J Pediatr Surg. 2004; 39: 430 – 8.
21. Garcia AM, Morgan WM, Bruner JP. In utero decompression of a cystic grade IV sacrococcygeal teratoma. Fetal Diagn Ther. 1998; 13:305–8.
22. Rescorla FJ, Sawin RS, Coran AG, Dillon PW, Azizkhan RG. Long-term outcome for infants and children with sacrococcygeal teratoma: a report from the Children's Cancer Group. J Pediatr Surg. 1998; 33:171–6
23. Azizkhan RG, Caty MG. Teratomas in childhood. Curr Opin Pediatr. 1996; 8:287-92.
24. Bittmann S, Bittmann V. Surgical experience and cosmetic outcomes in children with sacrococcygeal teratoma. Curr Surg. 2006; 63:51-4.
25. Draper H, Chitayat D, Ein SH, Langer JC. Long-term functional results following resection of neonatal sacrococcygeal teratoma. Pediatr Surg Int. 2009; 25:243-6.
26. Wang Y, Wu Y, Wang L, Yuan X, Jiang M, Li Y. Experimental and Clinical Resarch. Medical Science Monitor: International Medical Journal of Med Sci Monit.2017;23:17-23.
27. Bilik R, Shandling B, Pope M, Thorner P, Weitzman S, Ein SH. Malignant benign neonatal sacrococcygeal teratoma. J Pediatr Surg. 1993; 28:1158–60.
28. Lahdenne P, Heikinheimo M, Nikkanen V, Klemi P, Siimes MA, Rapola J. Neonatal benign sacrococcygeal teratoma may recur in adulthood and give rise to malignancy. Cancer. 1993; 72: 3727-31
29. De Backer A, Madern GC, Hakvoort-Cammel FG, Haentjens P, Oosterhuis JW, Hazebroek FW. Study of the factors associated with recurrence in children with sacrococcygeal teratoma. J Pediatr Surg. 2006; 41:173-81.