

Research Article

Sacrococcygeal Teratomas: Role of Intermittent Cyst Drainage during Operation

Beger Burhan^{1*}, Sönmez Bülent², Tuncer Oguz², Mehmet Edip Akyol³, Simsek Metin⁴ and Soysal Lokman¹

¹Department of Pediatric Surgery, VanYuzunciyil University, Van, Turkey

²Department of Neonatology, VanYuzunciyil University, Van, Turkey

³Department of Neurosurgery, Van Yuzunciyil University, Van, Turkey

⁴Department of Pediatric Surgery, Van Training and Research Hospital, Van, Turkey

Abstract

Background: Sacrococcygeal Teratoma (SCT) is a quite rare tumor yet is the most frequent congenital tumor type. Its incidence rate is reported to be 1 in every 35000-40000 live births. In this study, we assessed the techniques used in diagnosis and treatment of SCT patients in a pediatric surgery department.

Materials and methods: This retrospective study included 14 SCT cases presented to Van Yuzunciyil University Faculty of Medicine. Patients demographic properties, perinatal situations, surgical details, tumor histology, recurrence, morbidity, and mortality rates were reviewed.

Results: 14 (4 Males and 10 Females) cases with a mean age of 8 (Range: 2-34) days were included in the study. 11 patients were treated using Chevron incision and posterior sagittal approach incision was performed on the rest 3, 2 of the patients treated with Chevron incision required a scar revision. In 9 cases, the tumor was removed as a complete mass. Mass excision was done using intra-operative intermittent cyst aspiration method in the rest 5 cases.

Conclusion: Even though posterior sagittal approach yields to more cosmetically appealing results, it is not very reliable in bigger masses due to its surgical field limitations. In huge masses, Chevron incision and intermittent cyst drainage can be beneficial in reducing surgical vessel injuries and rectal injuries associated with the surgery.

Keywords: Sacrococcygeal teratoma; Chevron incision; Intermittent cyst aspiration; Tumor

Introduction

Sacrococcygeal Teratoma (SCT), albeit rare, makes up about 25% of neonatal tumors and 1% to 3% of all childhood period tumors [1-5]. Tumor incidence rate is reported as 1 in every 35,000-40,000 live births. Most of them are benign and can be resected and it is rarely seen with other anomalies. Since this tumor is a quite rare entity; only short patient series could be researched during the years in the literature. The treatment is the total excision of the mass including coccyx. Although cases without intra-abdominal components could be operated without any issues; huge cysts with intra-abdominal placement have severe morbidity and mortality-related technical complications such as cyst rupture, rectal injury, peripheral vessel injury and may be spinal canal injury [6]. Moreover, to prevent such complications, patients receive a prophylactic colostomy, or a residual tumor tissue is left on the excised area.

Using intermittent cyst drainage during surgery can be beneficial in creating a sufficient cleavage, especially during intra-abdominal

huge cyst excisions; resulting in a safer surgical approach. In this study, we would like to share our 7 years of experience on SCT methods in a 3rd step medical center on the eastern part of Turkey and assess it with the literature.

Materials and Methods

This retrospective study included 14 SCT cases presented to Van Yuzuncu Yil University Faculty of Medicine. Patients' demographic properties, perinatal situations, surgical details, tumor histology, recurrence, morbidity, and mortality rates were reviewed.

Surgical technique

Following bladder and rectal catheterization, chevron incision is used to lift dermal and other sub-dermal tissues. The cyst wall is separated from skin structures using sharp and blunt dissections. Following dissections up to pelvis, if posterior of cyst is not clear despite manipulations, intermittent 5 cc aspirations using Veress Needle can decrease the tension on cyst walls (like a slightly deflated balloon). After defining muscular structures and rectum using manipulations, the cystic lesion is completely aspirated (almost a completely deflated balloon) and tumor mass is completely excised including coccyx (Figure 1).

Results

The study group consisted of 10 females and 4 males. Median age of patients was calculated as 8 (Range: 2-34) days. According to Altman classification; 3 patients were Type 1, 10 were Type 2-3 and 1 was Type 4. Two cases had small (2 cm to 5 cm), 9 had moderate (5 cm to 10 cm) and 3 cases had large (>10 cm) tumors (Table 1). Only 4 cases could be diagnosed using prenatal ultrasound. Preoperative MRI results and tumor markers were assessed in every patient to

Citation: Burhan B, Bülent S, Oguz T, Akyol ME, Metin S, Lokman S. Sacrococcygeal Teratomas: Role of Intermittent Cyst Drainage during Operation. *Int J Pediatr Surg.* 2020; 1(1): 1002.

Copyright: © 2020 Beger Burhan

Publisher Name: Medtext Publications LLC

Manuscript compiled: Mar 04th, 2020

***Corresponding author:** Beger Burhan, Department of Pediatric Surgery, VanYuzunciyil University, K2-17/4, Van, Turkey, Tel: 0905056418310; E-mail: burhanbeger@hotmail.com

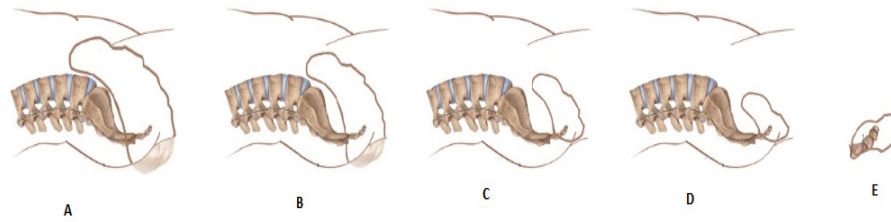


Figure 1: A: Initial Cyst, B: Partially Aspirated Cyst, C: Partially Aspirated, D:Maximal Aspirated Cyst, E: Excision of cyst and coccyx..

confirm the diagnosis (Figure 2). AFP level test was used routinely for postoperative follow-up. In 11 cases chevron incision performed whereas 3 had posterior sagittal approach incision (Figure 3). Two of the patients with chevron incision required scar revision. In 9 cases, tumor was removed as a complete mass (Figure 4). Mass excision was done using intra-operative intermittent cyst aspiration method in the rest 5 cases. No mortalities were seen in any of the patients during post-op follow-up period. Pathology results showed mature cystic teratomas (histological group 0) in all patients. Three of the follow-up patients developed constipation which responded to medical therapy, one patient showed recurrence. Patient follow-up was done using serum AFP levels and sacral-abdomen ultrasonography in 3-months period. One case with Type 4 according to Altman classification developed recurrence on 18th month and re-operated using abdominal approach. No additional anomalies were seen in any of the patients except 1 case with hydronephrosis which went into remission during follow up period.



Figure 2: IMR scan shows Cystic SCT.



Figure 3: Mini Chevron for SCT.

Discussion

Although SCT has no clear etiology, autosomal dominant familial cases were reported previously. It is about 4 times more frequent in

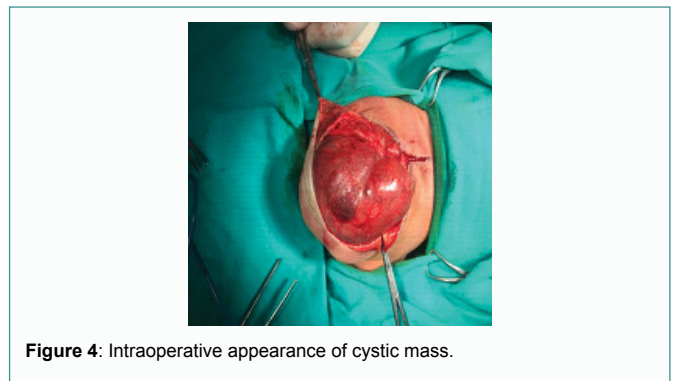


Figure 4: Intraoperative appearance of cystic mass.

Table 1: Gender of patients, classification according to Altman, mass size, surgical technique and results.

Patients	Gender	Altman Type	Mass Size	Surgical Technic	Result
1	F	Type 1	17 cm	CME	Cure
2	F	Type 2-3	8	CME	Cure
3	M	Type 1	4 cm	CME	Cure
4	F	Type 2-3	19 cm	ICA + CME	Cure
5	F	Type 2-3	9.5	ICA + CME	Cure
6	M	Type 2-3	10	ICA + CME	Cure
7	M	Type 1	16 cm	CME	Cure
8	F	Type 2-3	9	ICA +CME	Cure
9	F	Type 2-3	7.4	CME	Cure
10	F	Type 2-3	8	CME	Cure
11	F	Type 2-3	4.8 cm	CME	Cure
12	F	Type 4	9.6	CME	Reccurrence
13	M	Type 2-3	6	CME	Cure
14	F	Type 2-3	8.4	ICA + CME	Cure

females compared to males [7]. In our study, this ratio was calculated as 5:2 (F:M). SCT are germ cell tumors and roots from embryologic primitive knot and Hensen’s node [1]. Sacrococcygeal Teratomas are divided into 4 histologic groups. Group 0 is only mature elements, Group 1 consists of tumors with a small amount of immature embryonic neuro-ectodermal elements. Group 2 tumors have a moderate number of immature elements. Group 3 consist of tumors with a large number of immature elements and malignant endodermal sinus or yolk-sac tumor elements [8]. Altman et al. [9] classify SCT in 4 groups according to their radiological and operative appearance. Type 1 is tumors that can be viewed externally. In Types 2 and 3; there are both internal and external components. In Type 4, the tumor has no external components. Altman also classified SCT according to their size as small (2 cm to 5 cm), moderate (5 cm to 10 cm) and large (>10 cm) types. Types 1-3 has a low malignant transformation whereas Type 4 has a bigger risk of malignant transformation.

About 90% of SCT cases are diagnosed during early second trimester with a mixed echogenic mass view in fetal ultrasonography. As SCT is a fast progressing tumor, it could be presented with hydrops and can show early term radiological results such as polyhydramnios and placentomegaly. Prenatal diagnosis is crucial as to prevent mass obstruction-related complications in the baby with SCT. The reason for this low rate of prenatal diagnosis is related to most of the patients living in rural areas. Early diagnosis and surgery can prevent malignant transformation [10-13].

Intrauterine intervention time and indications are still debated. In cases with antenatal diagnosis; tumor-amniotic shunt or decompression with cystic aspiration can be beneficial in preventing complication development in some cystic tumors. Percutaneous drainage of cystic SCT can decrease risks for both the mother and the fetus [14]. However; although fetal therapy can prevent hydrops development by shrinking perinatal mass volume, it is frequently associated with intrauterine exitus or preterm deliveries [15-17].

The surgical procedure is total removal of the mass with coccyx. Coccyx removal decreases recurrence risks [13]. Chevron Incision (reverse V) is the incision done in classical SCT surgery. Jan et al. [18] reportedly obtained more aesthetically pleasing results by defining posterior sagittal intervention. In huge teratomas, modifications are possible with skin flaps. Two of the patients treated with chevron incision required scar revision in the long term. The posterior sagittal approach seems like an effective method in decreasing cosmetic morbidity in appropriate cases. In our Type 4 and Type 2-3 huge teratoma cases we used chevron incision to decrease iatrogenic injury risk by providing us a larger surgical viewing field. Some authors suggest protective colostomy in Type 4 and Type 2-3 huge teratoma cases which require abdominosacral excision [19,20]. Especially a colostomy performed at the beginning of the surgery in larger tumors stuck to rectum both facilitates tumor dissection and decrease morbidity in possible rectum injuries [20]. We used controlled-intermittent percutaneous drainage method during surgery in SCT cases with huge cystic components. As this method facilitates access to the posterior side of the mass, we think that this could reduce intraoperative vessel and rectum injury risks by easier mass excision without colostomy. In addition, it could also decrease the number of Type 4 cases which might require abdominosacral interventions. None of our patients received a protective colostomy. Serum AFP levels dropped in all infants following surgery. It is not only useful for diagnosis but also to detect any recurrences in those cases [10]. In all cases, preoperative AFP was >2000 yet decreased to normal levels in a month following surgery.

The perinatal mortality rate is 25% to 37% in prenatally diagnosed SCT cases. Mortality is associated with solid and fast-growing teratomas with a good blood supply with failure in higher cardiac output. Polyhydramnios, hydropsfetalis and preterm delivery risks are associated with this failure due to high cardiac output [15]. Fecal incontinence and constipation are frequently seen following SCT excision [1,2,10]. This is mainly associated with muscle and nerve damage during mass excision. About 18% to 20% of SCT cases also present additional anomalies. These anomalies can be anorectal, Currarino's Triad (anorectal fistula, anterior sacral defect, pre-sacral masses), renal anomalies (hydronephrosis, renal dysplasia), pulmonary hypoplasia, musculoskeletal anomalies (club feet, hip dysplasia), ventricular septal defect and neural tube defects (myelomeningocele, spina bifida) [5].

Recurrence rate is reported to be about 12.5% (Range: 2-35) and can be seen on 4-34th month after surgery [4]. Incomplete primary resection and malignant histology were proven to increase recurrence risks [21]. Only one of our cases showed recurrence. A low number of patients who received intraoperative cyst drainage can be the main limitation of our study. Therefore; this method needs to be prospectively used in larger series.

Conclusion

Operated patients should be followed up for at least 3 years with serum AFP levels and sacral-abdominal USG. Although more aesthetically pleasing results can be obtained with posterior sagittal approach; it is not very reliable in preventing rectal injuries in huge masses. Type 3-4 SCT cases should be followed up for urinary incontinence, fecal incontinence, and constipation. Chevron incision and intraoperative cyst drainage in Types 3-4 and huge cysts can provide a control over hemorrhages related to surgical vessel injuries. Moreover, this method can lift protective colostomy requirement by decreasing rectal injuries.

References

1. Draper H, Chitayat D, Ein SH, Langer JC. Long-term functional results following resection of neonatal sacrococcygeal teratoma. *Pediatr Surg Int*. 2009;25(3):243-6.
2. Isserman RS, Nelson O, Tran KM, Cai L, Polansky M, Rosenbloom JM, et al. Risk factors for perioperative mortality and transfusion in sacrococcygeal teratoma resections. *Paediatr Anaesth*. 2017;27(7):726-32.
3. Yoon HM, Byeon SJ, Hwang JY, Kim JR, Jung AY, Lee JS, et al. Sacrococcygeal teratomas in newborns: a comprehensive review for the radiologists. *Acta Radiol*. 2018;59(2):236-46.
4. Ho KO, Soundappan SV, Walker K, Badawi N. Sacrococcygeal teratoma: the 13-year experience of a tertiary paediatric centre. *J Paediatr Child Health*. 2011;47(5):287-91.
5. Kremer MEB, Althof JF, Derikx JPM, van Baren R, Heij HA, Wijnen MHWA, et al. The incidence of associated abnormalities in patients with sacrococcygeal teratoma. *J Pediatr Surg*. 2018;53(10):1918-22.
6. Ishibashi S, Tajima Y, Nakada S, Ota Y, Manako J, Ago M. Sacrococcygeal Teratoma with Spinal Canal Invasion. *J Pediatr Surg Case Report*. 2018;32:1-4.
7. Ayed A, Tonks AM, Lander A, Kilby MD. A review of pregnancies complicated by congenital sacrococcygeal teratoma in the West Midlands region over an 18-year period: population-based, cohort study. *Prenat Diagn*. 2015;35(11):1037-47.
8. Valdiserri RO, Yunis EJ. Sacrococcygeal teratomas: a review of 68 cases. *Cancer*. 1981;48(1):217-21.
9. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma. The American Academy Of Pediatrics Surgical Section Survey-1973. *J Pediatr Surg*. 1974;9:389-98.
10. Lee MY, Won HS, Hyun MK, Lee HY, Shim JY, Lee PR, et al. Perinatal Outcome of Sacrococcygeal Teratoma. *Prenat Diagn*. 2011;31(13):1217-21.
11. Baumgarten HD, Gebb JS, Khalek N, Moldenhauer JS, Johnson MP, Peranteau WH, et al. Preemptive Delivery and Immediate Resection for Fetuses with High-Risk Sacrococcygeal Teratomas. *Fetal Diagn Ther*. 2019;45(3):137-44.
12. Roybal JL, Moldenhauer JS, Khalek N, Bebbington MW, Johnson MP, Hedrick HL, et al. Early Delivery as an alternative management strategy for selected High-Risk fetal sacrococcygeal teratomas. *J Pediatr Surg*. 2011;46(7):1325-32.
13. Osman Ma, Ibrahim AI. Sacrococcygeal teratoma: 10-year experience in upper Egypt. *Ann Pediatr Surg*. 2015;8(2):45-8.
14. Abubakar AM, Nggada HA, Chinda JY. Sacrococcygeal teratoma in Northeastern Nigeria: 18-years experience. *Pediatr Surg Int*. 2005;21(8):645-8.
15. Van Mieghem T, Al-Ibrahim A, Deprest J, Lewi L, Langer JC, Baud D, et al. Minimally invasive therapy for fetal sacrococcygeal teratoma: case series and systematic review of the literature. *Ultrasound Obstet Gynecol*. 2014;43(6):611-9.

16. Gebb JS, Khalek N, Qamar H, Johnson MP, Oliver ER, Coleman BG, et al. High Tumor Volume to Fetal Weight Ratio Is Associated with Worse Fetal Outcomes and Increased Maternal Risk in Fetuses with Sacrococcygeal Teratoma. *Fetal Diagn Ther.* 2019;45(2):94-101.
17. Akinkuotu AC, Coleman A, Shue E, Sheikh F, Hirose S, Lim FY, et al. Predictors of poor prognosis in prenatally diagnosed sacrococcygeal teratoma: a multi institutional review. *J Pediatr Surg.* 2015;50(5):771-4.
18. Jan IA, Khan EA, Yasmeen N, Orakzai H, Saeed J. Posterior sagittal approach for resection of sacrococcygeal teratomas. *Pediatr Surg Int.* 2011;27(5):545-8.
19. Wakhlu A, Misra S, Tandon RK, Wakhlu AK. Sacrococcygeal teratoma. *Pediatr Surg Int.* 2002;18(5-6):384-7.
20. Cigdem MK, Önen A, Otcu S, Okur H, Akay H. Sakrokoksigeal Teratomlara Yaklasim. *Çocuk Cerrahisi Dergisi.* 2007;21(2):108-12.
21. Wang Y, Wu Y, Wang L, Yuan X, Jiang M, Li Y. Analysis of recurrent sacrococcygeal teratoma in children: clinical features, relapse risks, and anorectal functional sequelae. *Med Sci Monit.* 2017;23:17-23.