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ORIGINAL ARTICLE

Sacrococcygeal Teratoma: A 12-Year Experience Of A Single Center

Fatma Tuğba GÜVENÇ[1], [MD]
Burak ARDIÇLI*, [MD]
İbrahim KAHRAMAN[1], [MD]
Derya ERDOĞAN[1], [MD]
Engin YILMAZ[1], [MD]
Haşim Ata MADEN[1], [MD]
Ayşe KARAMAN[1], [MD]
İsmet Faruk ÖZGÜNER[1], [MD]

1 Dr. Sami Ulus Maternity and Children's Hospital, Department of Pediatric Surgery, Ankara, Turkey

* Corresponding Author: Burak Ardıçlı, Dr. Sami Ulus Maternity and Children's Hospital, Department of Pediatric Surgery, Ankara, TURKEY

e-mail: burakardicli@gmail.com

Phone: +90 312 305 6217

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ABSTRACT

Objective: Sacrococcygeal tumor is a rare benign or malign germ cell tumor of infancy and early childhood. We report a 12-year single-center experience.
Materials and Method: A total of 16 children operated with sacrococcygeal teratoma from 2004 to 2016 were reviewed retrospectively for their clinical features, treatment and follow-up.
Results: There were five boys and eleven girls between 1 day to 4 years of age. The most common symptom was sacrococcygeal mass. Complete resection of tumor involving coccyx was performed in all patients. Recurrence was seen in one case. Metastasis was also seen in one case.
Conclusion: Sacrococcygeal tumor is the most common tumor in newborn. Total excision is the treatment. The prognosis is excellent despite large tumor size and local invasion.

Key Words: Sacrococcygeal teratoma, child, newborn

INTRODUCTION

Sacrococcygeal teratoma (SCT) is a rare tumor. However, it is the most common congenital and neonatal neoplasm [1-2]. Studies have reported a SCT incidence of 1:14.000–1:40.000 [3]. Complete resection involving coccyx usually provides an excellent prognosis. Prenatal ultrasonography or postnatal examination is usually noted as a mass in the sacrococcygeal region. However, some presacral teratomas are noticed lately.

Altman et al. [1] suggested a classification of SCTs based on anatomical site and pelvic extension of the tumor: Type I is predominantly external, type II is external with significant intra-pelvic extension, type III is noticeable externally but with predominantly pelvic and abdominal extension, and type IV is completely presacral.

The predominant benign histology is mature teratoma and the predominant malignant histology is endodermal sinus tumor (yolk sac tumor; YST) [4]. Studies have reported a SCT recurrence rate of 2% to 35% [2-5-6].

We aimed to remind this rare tumor when diagnosing prenatal, infantile and childhood tumoral masses and to emphasize the importance of complete resection.

MATERIALS and METHOD

We analyzed the hospital records of children treated for sacrococcygeal teratoma between the period from January 2004 to June 2016, retrospectively. Age at diagnosis, gender, anatomic site, presenting symptoms, physical findings, radiological examination results, tumor markers (alpha-fetoprotein and beta human chorionic globulin), Altman classification, surgical management, postoperative complications, tumor histology, metastases, recurrence, and outcome were noted.

Statistical analysis was performed using SPSS 13.0 software. Descriptive data are expressed as the

mean ± SD, minimum and maximum values

RESULTS

We evaluated 16 children (see Table 1), with M/F ratio of 5/11 and a mean age of 236 days (1-1460). Seven cases of SCT (43%) were diagnosed prenatally. Six patients presented after the neonatal period. The most frequent presenting symptoms were sacrococcygeal mass (62%) and abdominal pain (18%). The diagnosis was based on physical examination findings in most cases and confirmed by ultrasonography (n = 9) and computerized tomography (n = 3) when required. Physical examination findings were palpable tumor in the sacrococcygeal (n=11) and abdominal areas (n=2).

All of the antenatal diagnoses of sacrococcygeal tumors were delivered with cesarean section. A presacral mass was detected in a patient who was operated for anorectal malformation. This case was diagnosed as a Currarino triad.

All patients had surgery and the tumor was removed with the coccyx as soon as the diagnosis was made. Surgery comprised sacral approach with total excision (n = 10), abdomino-sacral approach with total excision (n =4) and abdominal approach with total excision (n = 2). Reoperation was required for recurrence of SCT in one of the patients (5%).

Histological examination results were found as mature cystic teratoma (n=7), immature cystic teratoma (n=3), yolk sac tumor (n=2), mature solid teratoma, teratocarcinoma (n=1) and pure cystic teratoma(n=1).

Postoperative course was uneventful in 14 patients. Two patients who were operated with sacral approach had wound infection. One of the patients with yolk sac tumor is still on chemotherapy because of lung metastases. A patient with immature teratoma was operated 3 times because of local recurrence.

DISCUSSION

Sacrococcygeal teratoma (SCT) is a rare tumor. Benign tumors include mature teratoma (cystic, mixed or solid) and immature teratoma, while malignant tumors are pure endodermal sinus tumor (yolk sac tumor; YST) or mixed germ cell tumors containing embryonal carcinoma (EC). In addition, the sacrococcygeal area has the highest incidence of malignancy considering the teratomas. The frequency of neonatal sacrococcygeal teratomas with yolk sac tumor was 2.5% to 25% [7]. We had two patients with yolk sac tumor who were over a year old.

Eight patients were operated on newborn period and 7 of them had prenatal diagnosis (47%). Other studies have reported 50% to 74% prenatal detection rates [8-10]. We think that the devices and experience are important in this regard.

SCT patients often have a good prognosis when the diagnosis is made postnatally. The rates of morbidity and mortality associated with prenatally diagnosed (particularly before 30 weeks) SCT remain high [7]. The majority of SCT cases are female [1]. In accordance with the literature, our patients were predominantly female (M/F: 5/11).

A patient with recurrence had immature teratoma and she was operated on the 8th day of life. She was later operated 3 times for local recurrences. A patient with lung metastases had YST and she is still on chemotherapy.

The study noted an 11% tumor recurrence rate with mature teratoma and a 4% recurrence with immature teratoma. Recurrence occurred as late as 34 months postoperatively emphasizing the need for long-term follow-up. It is well known that recurrence risk increases with age [2]. Benign recurrences are usually located within the pelvis and can be managed with surgery. Malign tumors are associated with metastatic disease and can be managed with chemotherapy [11].

Hemorrhagic control may be difficult especially giant SCT and premature neonate. Several surgical approaches have been described to minimize bleeding during surgery. For example; median sacral artery ligation (laparoscopic), clamping of the abdominal aorta above the bifurcation and tumor embolization. In our cases, tumor embolization and clamping of the abdominal aorta were never used and there was no need for massive transfusion during or after surgery.

Sacrococcygeal teratoma has excellent prognosis in children. However, since early and late recurrences are major problem long-term close follow up is mandatory.

Table 1: Sacrococcygeal teratoma clinical and follow-up data

Case no.	Age (day	Sex	Altman classification	Prenatal diagnoses	Preoperative AFP	Preoperative B-HCG	Approach surgery	Histological diagnoses	Recurrence	Metastases
1	3	F	Type 1	+	High	High	Sacral	Mature cystic teratoma		
2	1460	F	Type 4		Normal	Normal	Abdominal	Mature cystic teratoma		
3	1	F	Type 2	+	High	High	Sacral	Mature cystic teratoma		
4	1	F	Type 2	+	High	High	Sacral	Mature cystic teratoma		
5	12	F	Type 3	+	Normal	Normal	Abdominal Sacral	Pure cystic		
6	8	M	Type 2	+	Normal	Normal	Sacral	Mature cystic teratoma		
7	2	M	Type 1	+	Normal	Normal	Sacral	Mature cystic teratoma		
8	8	F	Type 2		Normal	Normal	Sacral	Immature teratoma		
9	400	M	Type 4		Normal	Normal	Abdominal	Teratocarcinoma		
10	30	M	Type 3		Normal	Normal	Abdominal Sacral	Mature solid teratoma		
11	90	F	Type 3		High	High	Abdominal Sacral	Mature cystic teratoma		
12	1	M	Type 1	+	Normal	Normal	Sacral	Immature teratoma		
13	30	F	Type 2		Normal	Normal	Sacral	Immature teratoma		
14	520	F	Type 3		Normal	Normal	Abdominal Sacral	Yolk sac tumor		
15	730	F	Type 4, Currarino triad		Normal	Normal	Sacral	Mature cystic teratoma		
16	490	F	Type 2		High	High	Sacral	Yolk sac tumor		Lung

CONFLICT of INTEREST

None

REFERENCES

[1] Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973. J Pediatr Surg 1974; 9: 389-398.

[2] Yao W, Li K, Zheng S, et al. Analysis of recurrence risks for sacrococcygeal teratoma in children. J Pediatr Surg 2014; 49: 1839-1842.

[3] Hambraeus M, Arnbjornsson E, Borjesson A, et al. Sacrococcygeal teratoma: A population-based study of incidence and prenatal prognostic factors. J Pediatr Surg 2016; 51: 481-485.

[4] Calaminus G, Schneider DT, Bokkerink JP, et al. Prognostic value of tumor size, metastases, extension into bone, and increased tumor marker in children with malignant sacrococcygeal germ cell tumors: a prospective evaluation of 71 patients treated in the German cooperative protocols Maligne Keimzelltumoren (MAKEI) 83/86 and MAKEI 89. J Clin Oncol 2003; 21: 781-786.

[5] De Backer A, Madern GC, Hakvoort-Cammel FG, et al. Study of the factors associated with recurrence in children with sacrococcygeal teratoma. J Pediatr Surg 2006; 41: 173-181; discussion 173-181.

[6] Derikx JP, De Backer A, van de Schoot L, et al. Factors associated with recurrence and metastasis in sacrococcygeal teratoma. Br J Surg 2006; 93: 1543-1548.

[7] Isaacs H, Jr. Perinatal (fetal and neonatal) germ cell tumors. J Pediatr Surg 2004; 39: 1003-1013.

[8] Swamy R, Embleton N, Hale J. Sacrococcygeal teratoma over two decades: birth prevalence, prenatal diagnosis and clinical outcomes. Prenat Diagn 2008; 28: 1048-1051.

[9] Pauniaho SL, Tatti O, Lahdenne P, et al. Tumor markers AFP, CA 125, and CA 19-9 in the long-term follow-up of sacrococcygeal teratomas in infancy and childhood. Tumour Biol 2010; 31: 261-265.

[10] Gabra HO, Jesudason EC, McDowell HP, et al. Losty PD. Sacrococcygeal teratoma--a 25-year experience in a UK regional center. J Pediatr Surg 2006; 41: 1513-1516.

[11] Bilik R, Shandling B, Pope M, et al. Malignant benign neonatal sacrococcygeal teratoma. J Pediatr Surg 1993; 28: 1158-1160.