Sacrococcygealteratoma in Newborns: Management and Outcome

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ABSTRACT:

BACKGROUND:

Sacrococcygeal teratoma is the most common tumor in the newborn.. Approximately 90% of sacrococcygeal teratomas excised during the neonatal period are benign. The risk of malignancy increases in lesions removed after 6 months of age.

OBJECTIVE::

To discuss the management of newborns with sacrococcygeal teratomas and the importance of early surgical intervention in avoiding the risk of malignant transformation.

PATIENTS AND METHODS:

11 neonates with sacrococcygeal teratomas were managed during the period from April 2008 through April 2013. In all cases, age, sex, mode of delivery, details of surgery, extent of resection, macroscopic tumor appearance, tumor histology, Altman classification and postoperative morbidity and mortality were all documented and analyzed. A two years follow up plan was designed to assess any complication and tumor recurrence.

RESULTS:

Classical sacrococcygeal approach was performed in all of the patients operated upon and complete excision of the tumor along with the coccyx was possible in all cases. According to the Altman classification 8 (72.7%) of the tumors were of type I. Macroscopically, 7(63.6%) tumors were mixed,. Histopathological examination of the excised tumors revealed the diagnosis of benign mature teratoma in all cases. All of the newborns survived with the exception of one patient with a giant sacrococcygeal mass who died because of heart failure . No tumor recurrence was documented in all of the patients operated on.

CONCLUSION:

Early detection including antenatal diagnosis is essential to define fetuses with poor prognostic signs and deciding the appropriate mode of delivery, and in some centers even fetal intervention. Early excision of these tumors during the neonatal period is essential to avoid the risk of malignant transformation.

KEYWORDS: sacrococcygeal, newborn, teratoma.

INTRODUCTION:

Sacrococcygea lteratoma is the most common tumor in the newborn. The estimated incidence is 1 per 35,000 to 40,000 live births, with an unexplained female preponderance of 3:1[(1)].

Sacrococcygeal teratomas are complex lesions arising from totipotent cells of the yolk sac endoderm and contain tissues derived from the three embryonic layers: ectoderm, mesoderm and endoderm [(2)].

The diagnosis is generally made immediately after birth as most of these lesions are seen as visible masses. The main differential diagnosis includes meningocele, hemangioma, lymphangioma, lipoma, rare tumors like chordoma and several other rare conditions [(1, 2)].

Prenatal diagnosis is possible with the increased application of antenatal ultrasonography. The fetus should be delivered by cesarean section if the tumor is larger than 5 cm to avoid the problem of dystocia during vaginal delivery. The ultrasonographic findings of placentomegally or hydrops fetalis with large vascular tumors are indicators of fetal high-output cardiac failure and impending fetal death. These findings may lead to urgent cesarean delivery. In utero fetal surgical intervention has been performed with success in some centers [(1,2)].

Approximately 90% of sacrococcygeal teratomas excised during the neonatal period are benign. In lesions removed after 6 months of age the risk of malignancy is 50% or more. The risk of malignant

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degeneration approximates 100% with lesions removed at 3 years of age [1,2,3]. Therefore early surgical intervention is the standard approach of treatment to avoid the risk of apparent malignant transformation, as well as the risks of spontaneous ulceration and hemorrhage that may occur because of the tumor's rich vascular supply.

Excision of the coccyx is mandatory in all cases. Failure to remove the coccyx results in 30-40% recurrence rate, with a higher probability of malignancy [3].

Prognosis depends also on the tumor size, the histological type as well as the degree of prematurity [3].

PATIENTS AND METHODS:

Eleven neonates with sacrococcygeal teratomas were managed in the pediatric surgery departments in Basrah Children's Specialty Hospital and Medical City Hospital of Baghdad during the period from April 2008 through April 2013. Demographic data including age, sex and mode of delivery were documented. After confirmation of the diagnosis, patients were admitted to the intensive care unit. Preoperative preparation included routine laboratory tests with abdominal and pelvic ultrasound to asses any intrapelvic or intraabdominal extension of the mass.

In all cases, the details of surgery, extent of resection, macroscopic tumor appearance, tumor histology, the Altman classification and postoperative morbidity and mortality were all documented.

A two years follow up plan was designed at 3 months interval for the first year and 6 months interval for the second year. The plan elements included clinical examination, abdominal and pelvic ultrasound, serum alphafetoprotein evaluation after the age of 9 months with the aim to assess any complication and tumor recurrence.

In all of the patients operated on, the standard sacrococcygeal approach was used. After intubation and induction of anesthesia, the patient is placed in the prone position. The rectum is cleaned with povidone-iodine enema to prepare it for digital manipulation during the course of dissection. A chevron incision is made with its apex over the sacrum, continuing around the dorsolateral surface of the tumor. The tumor capsule is usually well defined and separate from other tissues. The tumor is then dissected out enbloc from the rectum , the pelvic diaphragm, and the gluteal muscles. The tumor including the coccyx is then removed. The procedure is finished with reconstruction of the pelvic floor. The posterior and superior portions of the levator muscles are sewn to the presacral fascia. This maneuver allows the anus to assume a near normal configuration and gives the best cosmetic outcome. Pelvic reconstruction is then followed by approximation of the gluteus maximus muscles and skin closure.

RESULTS:

Of the patients studied 7(63.6%) were females and 4 (36.4%) were males (Figure. 1). The age at presentation ranged from 4 hours to 30 hours according to the time of referral with a mean of 12.36 ± 7.17 hours (Table.1). Visible sacrococcygeal mass was the principal presentation in all cases and the diagnosis was obvious in all. Eight (72.7%) patients were delivered by cesarean section and 3 (27.3%) were delivered vaginally. Antenatal diagnosis was achieved by prenatal ultrasonography in only 4 patients.

Classical sacrococcygeal approach was performed in all of the patients operated upon and complete excision of the tumor along with the coccyx was possible in all cases. According to the Altman classification of sacrococcygeal teratoma, 8 (72.7%) of the tumors were of type I (predominantly external with minimal presacral component), and 3 tumors were of type II (present externally but with significant intrapelvic extension). Macroscopically, 7(63.6%) tumors were mixed (partly solid and partly cystic), 3 tumors were solid and only 1 tumor was purely cystic. Histopathological examination of the excised tumors revealed the diagnosis of benign mature teratoma in all cases.

All of the newborns operated upon survived with the exception of one patient who had a giant sacrococcygeal mass (22X16 cm) .Death had occurred on the second postoperative day because of heart failure that usually complicates newborns with giant lesions.

Postoperatively, 1 patient had mild wound infection that was treated successfully with antibiotics and frequent dressings and 1 patient had wound dehiscence that required secondary suturing two weeks later after resolution of the infection. Wound healing was good in all other cases.

Blood transfusion was indicated in one patient to replace blood loss during surgery.

Of the 10 patients who survived, 6 attended regular follow up plan at the scheduled dates for 2 years. 2 patients completed the first year plan, and 2 patients completed only 6 months of follow up. No recurrence was documented in all of the patients.

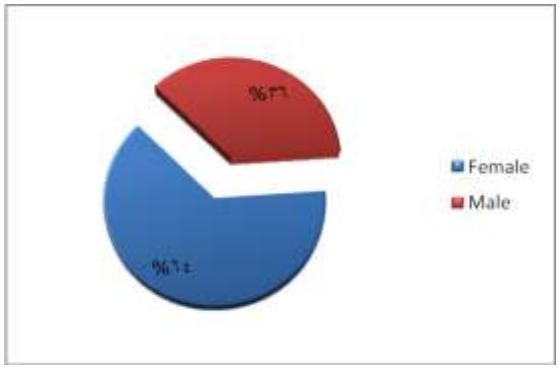

Figure 1: Male to female ratio

Table 1: Age at presentation and timing of surgery.

Patient	Age at presentation	Age at surgery
No.1	4 hours	28 hours
No.2	6 hours	30 hours
No.3	6 hours	30 hours
No.4	8 hours	36 hours
No.5	12 hours	40 hours
No.6	16 hours	44 hours
No.7	22 hours	50 hours
No.8	24 hours	48 hours
No.9	24 hours	52 hours
No.10	24 hours	60 hours
No.11	30 hours	66 hours

Table 2: Size of excised tumors.

Patient	Mass Size
No.1	12X10 cm
No.2	15X12 cm
No.3	22X16 cm
No.4	10X8 cm
No.5	15X10 cm
No.6	16X12 cm
No.7	7X6 cm
No.8	6X5 cm
No.9	12X8 cm
No.10	8X6 cm
No.11	10X5 cm

Table 3: Association between sex and tumor type according to Altman classification in our study.

Type of tumor	Sex	
	Male no. (%)	Female no. (%)
Altman type I	3 (75 %)	5 (71.4 %)
Altman type II	1 (25 %)	2 (28.6 %)
Total	4 (100 %)	7 (100 %)

X2 = 0.02 p.value> 0.05

Table 4: Demographic variables in comparison with other study .

Study	No. of Patients	Sex			Mode of delivery		
Ours	11	Male	4	36.40%	Vaginal	3	27.30%
		Female	7	63.60%	C/S	8	72.70%
Aly KA et al.	15	Male	3	20.00%	Vaginal	4	26.70%
		Female	12	80.00%	C/S	11	73.30%

Table 5: Tumor characteristics according to Altman classification in comparison with other study.

Study	Altman type I	Altman type II	Altman type III
Ours	72.70%	27.30%	0.00%
Perrelli et al.	75.00%	18.75%	6.25%

Figure 2: Complete excision of giant sacrococcygeal teratoma in 2 day old newborn.

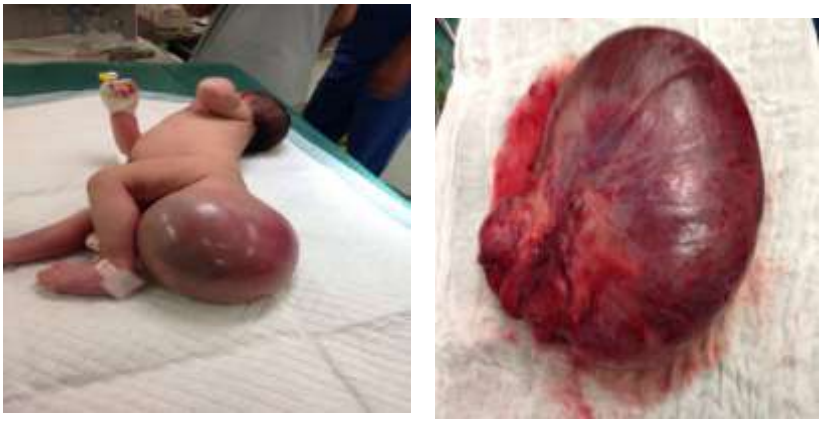

Figure 3: Complete excision of huge sacrococcygealteratoma in 1 day old newborn

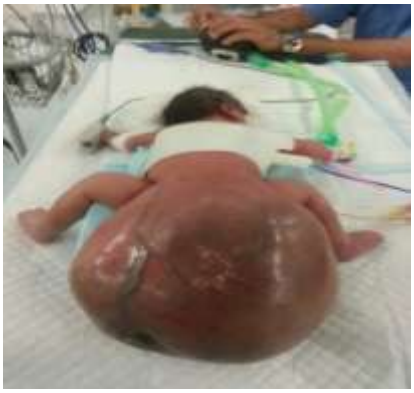

Figure (4)

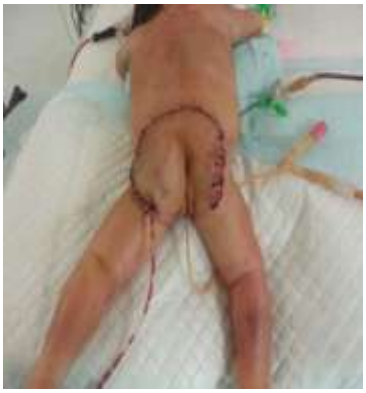

Figure (5)

Figure 4: Patient is placed in a prone position.
Figure 5: Perineal reconstruction.

DISCUSSION:

Sacrococcygeal teratoma is the most common tumor in the newborn. There is an unexplained female preponderance of 3:1[(1)]. In this study, female to male ratio was 1.75:1 which is less than that reported in other studies [(3,4)]. This might be a result of the small sample size of this study.

8 (72.7%) patients in this study were delivered by cesarean section. This is similar to what is reported in other studies [(4, 5)]. Table.2

Four of the cesarean sections were planned electively based on available antenatal diagnosis by prenatal ultrasound, and 4 were done on an emergency basis because of obstructed labour and fetal distress.

Antenatal diagnosis of sacrococcygeal teratoma is often made on prenatal ultrasonography, especially if the examination is performed in the second trimester[(1)].

It is recommended that a fetus with sacrococcygea lteratoma larger than 5 cm should be delivered by cesarean section to avoid the risk of intratumor hemorrhage[(1,6)] . Besides, antenatal ultrasonographic follow up can predict fetuses with poor prognostic signs [(1, 2)]. 36.3% of cases in this study were diagnosed antenatally. This result reflects the fact of irregular antenatal follow up of pregnant women in our locality.

In this study there were 8(72.7%) tumors with type I extent and 3 (27.3%) tumors with type II extent according to the Altman classification. These results vary slightly from those reported by Perrelli et al, who had 75% type I and 18.75% type II [(5)]. Table .5

Statistical analysis using chi-squared test in our study revealed no significant association between sex and tumor type according to Altman classification. Table.3

Macroscopically, 63.6% of the tumors were of the mixed type (partly solid and partly cystic). This result is consistent with the result reported by Keslar et al, who reported 62% of tumors in their study as mixed tumors [(7)].

Approximately 90% of sacrococcygealteratomas excised during the neonatal period are benign. The risk of malignant degeneration increases with age, therefore early surgical intervention is the standard approach of treatment to avoid this risk. All of tumors in this study were excised during the neonatal period(Table.1). Histopathological examination of the excised tumors revealed the diagnosis of benign mature teratoma in all cases. This result is similar to that reported by Hashish A. et al [(3)].

Most of the tumor-related mortalities are attributed to the hyperdynamic state caused by the arteriovenous shunting within the extremely large vascular tumor, which results in high output cardiac failure [(2,3)].All of the newborns operated upon in this study survived with the exception of one patient who had a giant sacrococcygeal mass (22X16 cm) .Death had occurred on the second postoperative day because of heart failure

CONCLUSION:

Although rare, Sacrococcygeal teratoma is the most common tumor in the newborn. Early detection including antenatal diagnosis is essential to define fetuses with poor prognostic signs and deciding the appropriate mode of delivery, and in some centers even fetal intervention. Early excision of these

tumors during the neonatal period is essential to avoid the risk of malignant transformation.

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