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The Outcome of Sacrococcygeal Teratoma: A Multicenter Study in Iraq

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ABSTRACT

Background: Sacrococcygeal teratoma (SCT) is one of the most common solid tumors in the neonate. They are more common in females and presented as sacral masses. They require an early surgical resection with regular follow-up.
Objectives: To evaluate the SCTs regarding the perinatal variables, clinical characters, pathological features, management options, and outcome.
Materials and methods: A prospective study of 37 patients with SCTs were admitted to pediatric surgery departments in Ramadi (west of Iraq) and Mosul city (north of Iraq) during the period from December 2013 to December 2018. The detailed data of each patient were recorded and analyzed including history, physical examination, investigations, management, and follow-up.
Results: There were 23 (62%) females and 14 (38%) males. The ages were ranged from 1 day to 2 years. Fifty-one percent were diagnosed prenatally by ultrasound and 30% of them were delivered by Caesarean section. Among the neonatal age group (28 patients),75% were born at full term, their median birth weight was 3100 g. Associated anomalies were 3%. Plain X-ray of the tumors was revealed calcification in 22%. The serum α-fetoprotein level was high at presentation. According to Altman classification, there were type I 54%, type II 40%, type III 3%, and type IV 3%. Ninety-seven percent was presented as sacral mass and only 3% was presented with urinary tract obstruction. One (3%) was died a few hours after labor due to the rupture of the tumor. Surgical intervention was performed in 94% of patients. Macroscopically, 59% were mixed, 22% solid, and 19% cystic. Histologically, 92% had mature teratoma, 3% immature, and 6% malignant teratoma. Wound infection occurred in 10%, wound dehiscence in 3%, and poor cosmetic scar in 8%. One (3%) patient had a recurrence.
Conclusion: The proper management are important factors affecting the outcome in the perinatal period and early diagnosis with complete excision of the tumor. The size of the primary tumor does not affect the recurrence and the outcome.

Keywords: Sacrococcygeal teratoma; Neonatal tumors; Outcome.

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INTRODUCTION

Sacrococcygeal teratoma (SCT) is one of the most common congenital and neonatal neoplasms [1] and it is the most common extra-gonadal tumor in neonates, accounting for up to 70% of all teratomas in childhood [2, 3]. A 3-4:1 female to male ratio is generally reported [2, 3]. The estimated incidence is 1 per 35,000-40,000 live births [3]. Abdominal delivery should be considered if the external mass is greater than 5cm. SCTs are generally presenting in two distinct fashions: neonates with large predominantly external lesions, which are detected in utero or at birth, and are rarely malignant, and older infants who present with primarily hidden pelvic tumors with a much higher rate of malignancy. Altman et al. have reported that the commonly utilized classification system for tumors at this location and noted the

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higher malignancy rate in the less apparent lesions (Type III and IV) [2].

The most helpful imaging studies are consist of plain anteroposterior and lateral radiographs of the pelvis and spine, looking for calcifications in the tumor and spinal defects, and the US of the abdomen, pelvis, and spine. Further preoperative studies are unnecessary in most newborns [3]. The basic steps of surgery are excision of the primary external lesion and the tail bone. Removal of the coccyx is still considered an essential step and a drain is often placed. Follow-up should consist of serial α-fetoprotein (AFP) levels every 3 months to ensure that they are normal by 9 months of age along with rectal examinations until 3 years of age [3].

We aimed to assess the sacrococcygeal teratoma regarding the perinatal variables, clinical characteristics, pathological features, management options, and outcome at 2 pediatrics surgery departments in Ramadi and Mosul cities, Iraq.

MATERIALS AND METHODS

A prospective study of 37 (23 females and 14 males) patients with sacrococcygeal teratomas in two pediatric surgery departments in Ramadi Teaching Hospital for Maternity and Children, Ramadi city, and Al-Khanssa Teaching Hospital, Mosul city, Iraq. The study was covered the period from December 2013 to December 2018. The study was approved by the Iraqi Board for Medical Specialization and the informed consent was taken from the parents of the babies.

A detailed medical history of each patient was obtained including age, gender, mode of delivery, and prenatal history. Physical examination was performed including digital rectal examination, the weight of patients, examination of the mass with the exclusion of any associated anomalies.

A baseline serum AFP level was obtained preoperatively and rechecked after 3 months. Plain X-ray lateral view and ultrasound of the mass were performed in all cases. Ultrasound of the abdomen and urinary tract was obtained to exclude associated anomalies. The CT-scan and MRI were not used routinely because they were not available in our hospitals at the beginning of the study.

All patients were operated on, except one who died within a few hours after the vaginal delivery. The operation consists of surgical excision of the tumor and the coccyx bone by sacral or abdominosacral approach. All operated cases were sent for histopathological examination. The baby with a malignant condition was sent to a pediatric oncology center for further management. The follow-up period was ranged from 3 months to 12 months.

We classified our patients into 3 groups according to their ages neonates 1- 28 days, infants 29 days- 1 year, and patients older than 1 year. SCTs were divided into 4 types according to the Altman et al. study [1] Type I are almost exclusively exterior with minimal pelvic part; type II have a significant pelvic part (hour-glass pattern); type III has a larger proportion of intra-abdominal and intra-pelvic part than the external part, and type IV are exclusively pre-sacral with no external part. Any postoperative complications including the recurrence were recorded for every case.

RESULTS

Age and Sex of Patients

Neonates were 28 (76%), infants were 6 (16%), and $>$ 1 year was 3 (8%). There were 23 females (62%) and 14 males (38%) with a male to female ratio of 1: 1.6.

Prenatal Diagnosis and Mode of Delivery

Nineteen (51%) were diagnosed prenatally by the US at $2^{nd} - 3^{rd}$ trimester and only 11 of them had planned cesarean section (CS). Delivery of the babies was vaginally in 26 (70%) cases and by CS in 11 (30%) for those with large masses. One (3%) patient with a large tumor died due to the rupture of the tumor after vaginal delivery, as shown in Figure 1.

Gestational Age and Birth Weight

The gestational age of 28 cases was presented in the neonatal age group ranged between 35-40 weeks. Out of 28; 21 (75%) patients were born full-term and 7 (25%) at gestational age less than 37 weeks. There was no precise data about the gestational age of those presented beyond neonatal age. The birth weight of the neonatal age group was ranged between 2200-3800 gm (median = 3100). We could not collect precise data about birth weight for those presented beyond neonatal age.

Associated Anomalies

Only one patient was with congenital heart disease (3%).

Radiologic Calcification and Serum AFP Level

X-ray of the tumor was performed in 36 patients except for the one who died early after labor and calcification were detected in 8 cases (22%) as shown in Figure 2.

Serum AFP level was performed in 27 patients at presentation and rechecked in 22 patients during follow-up. The age at rechecking was variable ranging from 3-7 months depending on the visiting of the patients. Serum AFP level was noted to be high at the time of presentation and decreased at the rechecking time except in one case whose AFP level was still high when the recurrence was detected.

Altman Classification

According to Altman's classification; 20 (54%) tumors were typed I, 15 (40%) type II, 1 (3%) type III, and 1 (3%) type IV.

Clinical Findings

Out of 37; 36 patients (97%) were presented with sacrococcygeal mass, one of them was presented with a ruptured

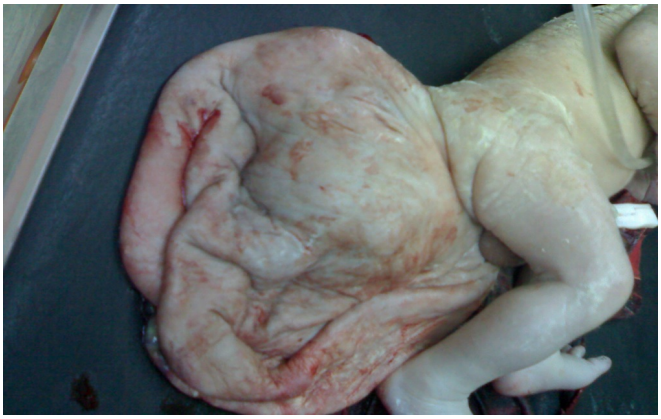

Figure 1. Ruptured sacrococcygeal teratoma. This case was reported from the pediatric surgery department in Mosul city.

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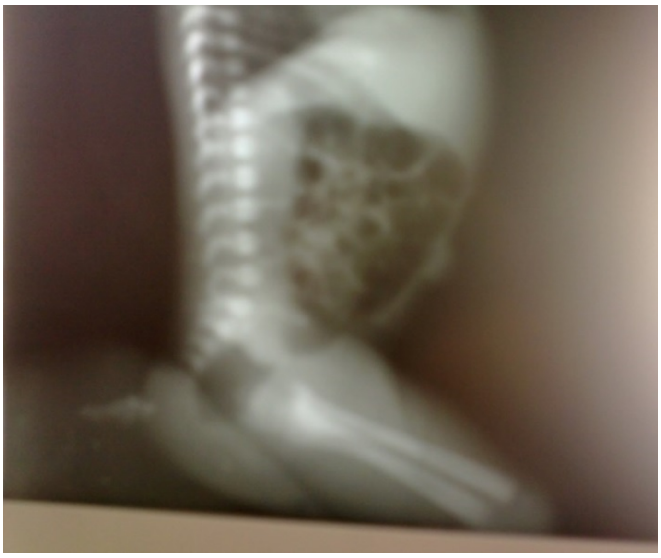

Figure 2. Plain X-ray of SCT revealing calcification.

tumor during labor, another one presented at age of 2 years with sacral mass which was diagnosed erroneously as abscess and drained surgically then referred to our department. One patient (3%) was presented with urine retention at the age of 5 months due to the pressure effect of intra-pelvic mass (type IV).

Surgical Treatment

From 36 patients who underwent surgery, 35 (97%) had complete excision of the tumor via sacral route only as shown in Figure 3. One patient (3%) with large intra-pelvic extension and urinary tract obstruction underwent a one-stage combined abdomino-sacral intervention. In all cases, the excision of the coccyx was performed.

Macroscopic Features

Twenty-two (59%) patients were mixed, eight (22%) solid, and 7 (19%) cystic lesions.

Histopathology Examination

The histopathological examination was performed in 36 cases, 33 (91%) patients were mature, 1(3)% immature, and 2 (6%) malignant teratomas (one diagnosed at the age of 8 months (type II) and the other at the age of 2 years (type III)). One of them was lost of follow-up because he went outside of Iraq for chemotherapy and the other was referred to Oncology Center for Chemotherapy. All neonates had benign mature teratomas. One patient at the age of 5 months was revealed immature teratoma. Both of the malignant teratomas were revealed as an endodermal sinus tumor (yolk sac tumor).

Postoperative Complications

Postoperative complications have occurred in form of wound infection in 4 (10%) patients that healed with local wound care and antibiotics. Wound dehiscence occurred in one (3%), and poor cosmetic scars occurred in 3 (8%) patients.

Follow-Up

One (3%) patient with type I Altman classification was revealed recurrence within 3 months after the excision of the primary benign mature teratoma at neonatal age. The recurrence was presented by the development of sinus at the scar of previous surgery and AFP level was high. Re-excision was performed and the histopathology was revealed benign mature teratoma.

DISCUSSION

Our study was showed that SCT was presented most commonly in the neonatal period (76%). Studies by Chirdan et al. [4], Swamy et al. [5], and Gabra et al. [6] have shown similar findings. They have found that the neonatal age group was represented 60.5%, 66%, and 69% of their studied cases respectively. Infants have comprised 16% of cases, which was lower than the study of Chirdan et al. (26.5%) [4], and higher than the study of Gabra et al. (6%) [6]. This difference may be related to a high percentage of patients with intra-pelvic tumor (24% type IV Altman classification) in their study resulting in a late presentation of the tumor. While those older than one-year-old were comprised of 8% of the cases, and this was similar to the study of Gabra et al. [6] in which there were 13% older than one year.

Data from this study were shown a slight female preponderance (62%). While most other studies were showed a high female preponderance ranged from 74%-92% [4–13]. These differences in male: female ratios may be related to racial variations. Fifty-one percent of patients were diagnosed prenatally, this was similar to the study of Gabra et al. [6] and Swamy et al. [5], in their studies the prenatal diagnosis have 53% and 50%, respectively. Our result is high when compared to the study of Chirdan et al. [11] in which the prenatal diagnosis was 10.5%. If the postnatal group was excluded, the detection rate would be 68%. This was similar to the study of Swamy et al. [5], and Hambraeus et al. study (74%) [12] in which the detection rate of prenatal diagnosis was increased from 50% to 63% when the postnatal group was excluded. Nine patients were presented beyond the post-neonatal group, none of these patients were detected prenatally.

The CS was performed in 11 (30%) patients. This was concomitant with the study of Gabra et al. (30%) [6] and Yadav et al. [13] (20%). While the percentage of CS in our study was lower than the study of Swamy et al. (50%) [5] and Aly et al. (73%) [10]. This difference may be attributed to obstetric problems indicating for CS. In our study, the rupture of the tumor occurred in 1 (3%) patient, this was similar to the study of Kouranloo et al. [7] who have found that rupture of the tumor occurred in 4%. Rupture of tumor in our study was because of the vaginal delivery in one case which was not diagnosed prenatally and thus the perinatal mortality in our study is 3%.

In this series, 75% of the neonatal age group were born at the term and 25% were born at gestational age less than 37 weeks. This fairly concomitant with the study of Swamy et al. [5] in which there were 40% born in gestational age less than 37 weeks. The prematurity in our study was higher when compared with the study of Chirdan et al. [4] who found that the prematurity in 3%. This difference may be related to (1) the size of the tumor which may induce early labor (2) fetal and obstetric complications related to the presence of the tumor which indicates early termination of pregnancy. The median birth weight of the neonatal age group was 3100 g. Other

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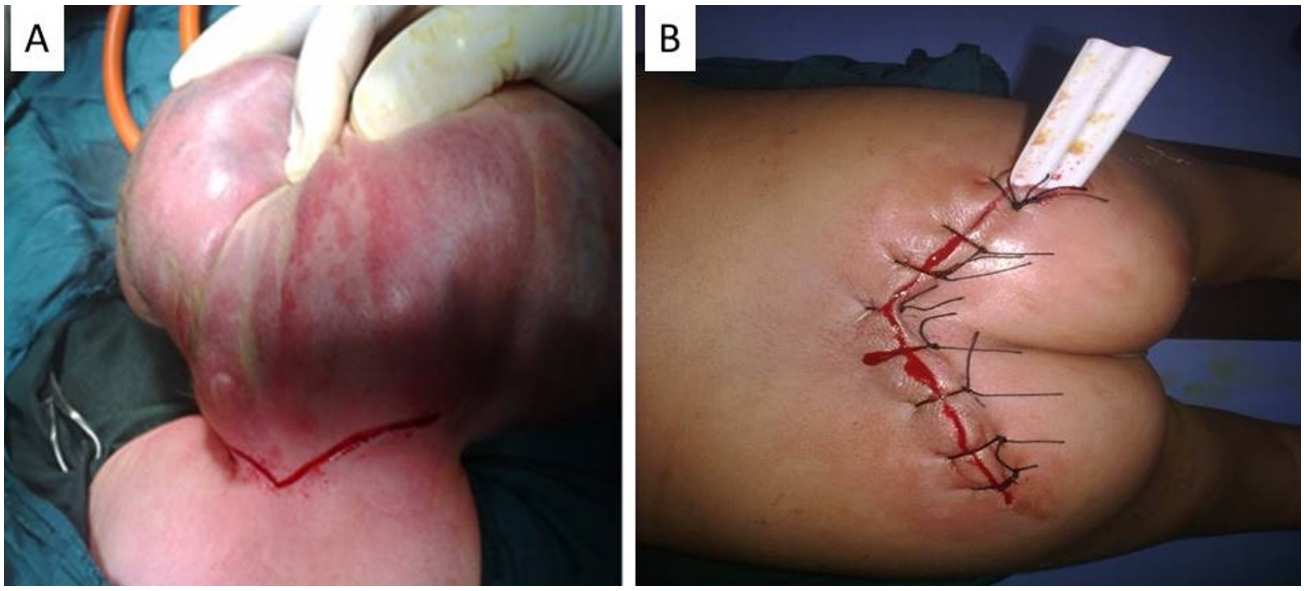

Figure 3. Chevron incision in one of the cases from the pediatric surgery department in Al-Khanssa Teaching Hospital, Mosul.

studies have been shown different data; Swamy et al.[5] (median birth weight =3420 g), Gabra et al. [6], and Hambraeus et al. [12] (median birth weight =3300 g) and Chirdan et al. [4] (median birth weight =2800 g). This difference may be related to tumor size, race, nutritional status, and economic status.

The associated anomalies were 3%. Studies of Chirdan et al. [4], Swamy et al. [5], Gabra et al. [6], Derikx et al. [9], Aly et al. [10], Shanbhogue et al. [11], Yadav et al. [13] and Kremer et al. [14] have found associated anomalies in 5%, 13%, 24%, 24.3%, 20%, 16%, and 32%, respectively. It is clear that the associated anomalies in our study were different than other studies, this possibly because (1) we received most of our patients from other hospitals, and most of the children with life-threatening associated anomalies might die before referral to our departments, (2) some of these anomalies especially urinary tract anomalies might not be detected by the US, (3) we did not use echocardiogram routinely in all cases for detection of congenital heart disease.

Twenty-two percent had calcification in their tumor and this fairly similar to the study of Ein et al. [8] who have found the calcification in 33%. Serum AFP level was high at presentation and fell following resection of the tumor, but it has stayed at the high levels in recurrent cases. This is concomitant with the study of Aly et al. [10], Kouranloo et al. [7], and Derikx et al. [9]. They have found that serum AFP levels were decreased to the normal postoperatively but were elevated again when tested in recurrent cases. Regarding Altman classification; the result in our study was compared with other studies from different locations as shown in Table 1. Our study was similar to other studies in that all have shown that type I and II are the most common. This explains the reason why the diagnosis was made in most cases at neonatal age. There was no relationship between the size of the tumor with the recurrence rate and histopathological features in the current study. This is similar to the study of Kouranloo et al. which have found that the size of the primary SCT tumor was not correlated with the high recurrence rate and outcome [7].

Most SCTs are seen as a visible mass at birth [3]. In our study, 97% of patients were presented with a sacral mass of different sizes except one (3%) who presented with urine retention at age of 5 months old. In one child, the sacral mass was diagnosed as an abscess and drained. This is similar to the study of Chirdan et al. [4] which have found that the sacral mass is comprised of 97% and the urinary obstruction 3%. The complete excision of the tumor with resection of the coccyx was performed by the sacral approach in 97% and by one stage combined abdomino-sacral approach in 3% for type IV tumor. Similar results have been found in previous studies [4, 6, 9, 10] as shown in Table 2.

Macroscopically, 59% of our patients were mixed, 22% solid, and 19% cystic. These results are similar to the results of Chirdan et al. [4] which have found that the mixed type is 53%, solid 26%, and cystic 21%. The histopathological examination was revealed mature teratoma in 91%, immature teratoma in 3%, and malignant teratoma in 6%. This is similar to the study of Gabra et al. [6] which have found that the mature teratoma comprised 79%, immature teratoma 3%, and malignant teratoma 18%. Our study is different from the study of Chirdan et al. [4] which have found that the mature teratoma in 60%, the immature in 24%, and the malignant teratoma in 16%. This difference in mature and immature type might be related to inherited, racial, or geographic differences or might be related to the histopathological difficulties in the differentiation of mature and immature teratoma.

In our study, there was no malignancy in the neonatal age group, similar to the study of Chirdan et al. [4]. However, our study concomitant with this study in that the risk of malignancy is elevated beyond the neonatal age group. Minor postoperative wound infection occurred in 10%, this was concomitant with the study of Gabra et al.[6] and Ein et al. [8] that they have recorded minor wound infection in 21% and 18%, respectively. While the wound dehiscence occurred in 3% of our patients which differ from the study of Chirdan et al. [4] and Gabra et al. [6] that they have found the dehiscence in 21% and 18% respectively. This difference might

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Table 1. Comparison of Altman classification between our study and other studies.

Altman classification	Our study	Hambraeus et al. [12]	yadav et al. [13]	Yao et al.[15]	Derikx et al.[9]
Type I	54%	57%	39%	51%	38%
Type II	40%	14%	24%	19%	30%
Type III	3%	21%	17%	15%	12.1%
Type IV	3%	7%	20%	15%	16.8%

Table 2. Comparison of the result of the surgical approach in our study with other studies.

Approach	Our study	Aly et al. [10]	Chirdan et al. [4]	Gabra et al. [6]	Yadav et al. [13]
Sacral	97%	93.3%	89%	88%	83%
Abdominosacral	3%	6.7%	11%	12%	17%

be related to using water-resistant dressing (opsite) in most of our patients using antibiotics. Our study and Aly et al. study [10] didnt record any rectal injury. Unsatisfactory cosmetic scars were observed in 8%, and this is lower than that observed by Aly et al. [10] which have found poor cosmetic scars in 40%. This possibly due to the low rate of wound infection and dehiscence in our study.

Only one (3%) patient had a recurrence. This was concomitant with other studies like the study of Chirdan et al. [4], Gabra et al. [6], Aly et al. [10], Derikx et al. [9] and Yao et al. [15], which have found that the recurrence has occurred in 5%, 9%, 6.7%, 11.1%, and 16%, respectively. Regarding the causes of the recurrence, the recurrence in our study was caused by incomplete resection of the primary tumor. Therefore, incomplete resection is considered as a risk factor for recurrence. The same finding has been noted by Derikx et al. which have found that the most important risk factor for recurrence was incomplete resection [9].

In conclusion, prenatal diagnosis and proper planning for delivery are important for minimizing the perinatal mortality. The size of primary SCT does not relate directly to the likelihood of recurrence and poor outcome. Early diagnosis and complete surgical excision are the most important factors affecting the outcome and morbidity. Beyond the neonatal period, the risk of malignancy is increasing. Regular follow-up of the patients postoperatively by physical examination and serum AFP level is important for early recurrence detection.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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