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Sacrococygeal Teratomas Experience with 32 Patients

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✔ Case records of 32 patients (22 female and 10 male) with sacrococygeal teratoma who were treated in our department between 1984 and 1994 were reviewed. 27 patients had benign sacrococcygeal teratoma and the majority of the benign sacrococygeal teratomas were newborns. Total excision with coccygectomy was performed in this group. In contrast, only one of the patients with malignant tumor was a newborn. In malignant group excision and multiagent chemotherapy were used for the treatment and two recurrences were noted. The average survival was increased and early mortality was decreased for all patients in comparison with past decades.

Key words: Teratoma, sacrococygeal

✔ 1984 ile 1994 yılları arasında servisimizde takip edilen 32 Sakrokoksigeal teratomlu hasta (22 kız ve 10 erkek) yeniden gözden geçirildi. Hastaların 27 tanesi benign teratomdu ve bunların çoğu yenidoğan döneminde başvurmuştu. Bu gruptaki hastalara total eksizyonla beraber koksigektomi uygulandı. Tersine, malign tümor grubunda yalnız br hasta yenidoğan döneminde başvurmuştu. Bu grupta ise tedavi amacıyla ekzisyon ve multiajan kemoterapi uygulandı ve iki rekürens gözlendi. Geçen dekadlarla karşılaştırıldığında, son dekadda hastaların yaşam sürelerinin uzadığı ve erken mortalitenin düştüğü saptandı.

Anahtar kelimeler: Teratom, sakrokoksigeal

Sacrococygeal teratoma (SCT) is the most common tumor in the neonatal period occurring one in 35000 to 40000 livebirths[1,2]. The first recorded case was found on a Chaldean cuneiform tablet dated approximately 2000 BC[3]. The prognosis for the malignant SCT has been considered poor until 1980's, but has improved considerably in recent years by the use of multiagent chemotherapy[4,5,6].

This paper presents a clinical retrospective study in order to evaluate the diagnostic and therapeutic approaches to SCTs in our clinic.

MATERIAL AND METHOD

A retrospective review of SCT was made in our department archives for the 1984–1994 period. The number of the cases and clinical characteristics including sex, age at presentation, prenatal and perinatal histories, the physical status and the appearance of the tumor were noted. X-ray appearance and serum alpha-fetoprotein (AFP) levels were documented, if possible. The diagnostic modalities for each patient was noted. The therapy for each case was analysed. Tissue diagnosis including benign and malignant forms and if malignant the details of the chemotherapy and/or radiation therapy were obtained.

The tumors classified according to the Altman's anatomical classification[4]. Type I SCT had no presacral component. Type II SCT was predominantly external but had a definitive presacral component extending to the abdomen. Type III SCT was mostly in the abdomen

but had a small external component. Type IV SCT was entirely presacral with no external component.

RESULTS

Of the 32 SCTs, 27 (84.4%) were benign and 5 (15.6%) were malignant. Girls were affected more often than boys (22 vs 10; 68.8% vs 31.2%). The female–male ratio was 2.08:1 for benign lesions and 1.5:1 for malignant lesions. For all children with SCTs, the age at presentation ranged from newborn to 2.5 years. With benign lesions, almost all the children were newborns at the time of diagnosis (26 of 27). On the other hand, only one patient of five malignant lesions was newborn. The average age at diagnosis of benign lesions was significantly younger with 3.1 days (range 1–45 days) compared with malignants which was 229 days (range 1–890 days) (table 1).

There were 22 type I SCTs (68.7%). Of these, 20 were benign and two were malignant. Four patients presented with type II lesions, of which all were benign. Type III lesions were also found in 4 patients. Of these, 3 were benign and 1 was malignant. Finally, 2 patients presented with type IV lesions, of which all were malignant (table 2).

In cases originally diagnosed as benign lesions, total excision of the mass and coccygectomy were performed in all patients and no recurrence was noted in the follow up group. Morbidity in benign SCTs included 7 wound infections, and 2 septicaemias. One death has been recorded in this group. This neonate was lost preoperatively and, had multiple congenital anomalies including spina bifida, pes eqinuovarus and potter face in addition to SCT. It is notable that exstrophy vesicale and SCT were seen together in 2 patients. One neonate had a second retroperitoneal benign teratoma.

In malignant group, total excision and coccygectomy were performed initally in 3 of 5 patients and two recurrencus were noted. One of the patients had been initially operated in a local hospital with the diagnosis of gluteal abscess without performing coccygectomy. This patient was admitted to our hospital with recurrence in the fourth month following the initial therapy. The chemotherapy and radiotherapy were performed following biopsy (case 1). One of the patients died intraoperatively because of massive haemorrhage (case 5). In the remaining of group, one recurrence was noted following one year after initial therapy (case 2). After recurrence, biopsy was performed and radiotherapy in addition to the chemotherapy was initiated. The malignant patients are detailed in table 3. Morbidity in malignant lesions included only wound infection.

Clinical significance of results of late complications and long–term survivals of these patients are difficult to asses because 11 of 27 benign SCTs and 4 of 5 malignant SCTs were available for follow up over 6 months after operation.

Beginnnig in 1985, alpha–feto protein (AFP) levels have been determined in our clinic for 12 patients. But, no significant change in this marker for any patient because of therapy is identified.

As previously reported, 50% of all teratomas have calcification with no special relation to malignancy[7]. In our series, calcification was found in eight of the benign SCT patients (25% of total).

DISCUSSION

Teratomas are tumors with a wide spectrum of clinical presentation. In 1974, the largest series of patients with SCT was published by Altman et al. describing 405 cases[6]. After this, serial authors have reported great

Table 1: Age at presentation

AGE	benign SCT	malign SCT	TOTAL
0-30 days	26 (96.3 %)	1 (3.7 %)	27 (84.39 %)
30day-3 months	1 (33.3 %)	2 (66.7 %)	3 (9.37 %)
3-6 months	0	1 (100 %)	1 (3.12 %)
> 6 months	0	1 (100 %)	1 (3.12 %)
TOTAL	27 (84.38 %)	5 (15.62%)	32 (100 %)

Table 2: Types of SCT

TYPES	benign SCT	malignant SCT	TOTAL
I	20 (90 %)	2 (10 %)	22 (68.75 %)
II	4 (100 %)	0	4 (12.50 %)
III	3 (75 %)	1 (25 %)	4 (12.50 %)
IV	0	2 (100 %)	2 (6.25 %)
	27	5	32

numbers of patients with this common neonatal tumor[1,5,7,8]. Girls are affected more often than boys particularly in benign lesions and the female-male ratio is higher in benign lesions compared with malignant lesions[9]. In our series, 68.8% of patients was girl and the female-male ratio was higher in the benign group.

It is generally believed that most of the benign tumors are seen in the newborns and the malignant form occurs predominantly beyond the first year[7]. The classical benign tumor presents as a mass in the neonatal period. In our series, 96.7% of the benign SCTs were newborns and all the patients in this group presented with a mass in sacrococcygeal region.

Total excision of the mass with coccygectomy is the main procedure for the treatment. Morbidity in the present series still remains notable particularly with 25.92% wound infection and with 7.4% septicemias. However, the high survival rate of our series of benign lesions remarkably depends on the modern antibiotic therapy in septicaemia and multidisciplinary care of these patients.

The necissity of removing the coccyx to avoid recurrence was evident before this decade[10,11]. In our series, there is one patient who did not undergo coccygectomy at initial therapy. This patient had recurrence in the fourth month following therapy. Therefore, we agree that coccygectomy should be performed routinely at the time of tumor excision.

Table 3: Malignant SCTs

CASES	Year of Diagnosis	Type	Age at diagnosis	Surgery	Chemotherapy	Radiotherapy	Outcome
Case 1	1984	I	4–months	Excision Without coccygectomy	(-)	(-)	reccurence at the 4. year Chemotherapy + radiotherapy
Case 2	1984	III	3–months	Excision With occygectomy	(+)	(+)	reccurence at the 1. year Chemotherapy + radiotherapy
Case 3	1986	I	2–days	Excision With occygectomy	(+)	(-)	no reccurence in 3 months
Case 4	1990	IV	2.5–years	Excision With occygectomy	(+)	(-)	no reccurence in two years
Case 5	1993	IV	1.5–years	intraoperative death			

According to Altman's classification, malignant lesions are commonly presented with type III and IV lesions. In our series all type IV lesions were malignant. The rate of malignancy increased significantly in our series with the increased age and Altman's types. Surgical resection for malignant tumours is a prerequisite for care but most malignant tumours are unresectable at the time of diagnosis[5]. So that, before the use of multiagent chemotherapy, the prognosis for malignant SCT was considered to be poor. However, since the introduction of multiagent chemotherapy the prognosis seems to have been improved[1,6].

We have to emphasise that it is impossible to definitively attribute the decrease in mortality only to adjuvant chemotherapy; because of the improvement in the treatment of paediatric patients including better ventilators, parenteral nutrition, advanced antibiotics and the technological advence in the diagnosis of malignant SCT[7,8,9]. Computed tomography (CT) and magnetic resonance imaging (MRI) are excellent techniques for imaging the sacrum and presacral area[10]. MRI adds multiplanar capacity and improved soft tissue contrast. With the use of these diagnostic modalities, the success of the operative management can be increased.

In some cases, early ultrasonograhic screening during pregnancy has made the diagnosis of SCT possible in the antenatal period. Polihydramniosis and large-for-days uterus are the main ultrasonographic indications[11]. The advantage of US is that it permits early correct diagnosis of the tumor with subsequent caesarean section, however false negative results with US have been reported [13,14]. We believe that prenatal tumor diagnosis is very important and this will lead to better preoperative management of these patients.

The risk of early delivery, immaturity of the child and massive haemorrhage are the important risks of fetal teratomas. The high occurrence of immature teratoma in fetal groups of tha series of Sheth et al., Flake et al. and Havranek et al is suprising[1,2,12]. Here, in our series there was not any patient diagnosed prenatally.

Several authors have reported on AFP as a marker of malignant teratoma[14,15,16]. The value of AFP in the newborn period has been discounted becuse of the physiological elevation of this marker due to its production in the fetal liver and to a lesser extend in the gastrointestinal tract[10]. In 1983, Tsuchida and Hasegawa distinguishd the hepatic and yolk sac original types of AFP[17]. The sensitivity of serum AFP levels in relation to tumor recurrence makes AFP very valuable in monitoring the effect of chemotherapy as well.

In 1992, Murphy et al. noted that the frequency of coagulopathy and haemorrhage in neonates with large SCTs might be higher than previously appreciated[18]. It is believed that prompt surgical excision of the lesion should be undertaken immediately after birth because the coagulopathy appears to worsen by time. In the last decade the average time of excision for the neonatal period patients decreased remarkably. Maybe, in the future fetal surgery will play role in the treatment of these large SCTs.

We believe that greater survival rates may be attainable even in children having metastatic disease, with early diagnosis by US, accurate diagnosis with CT nd MRI and ability to screen for recurrences with AFP as well as the multidisciplinary intensive care.

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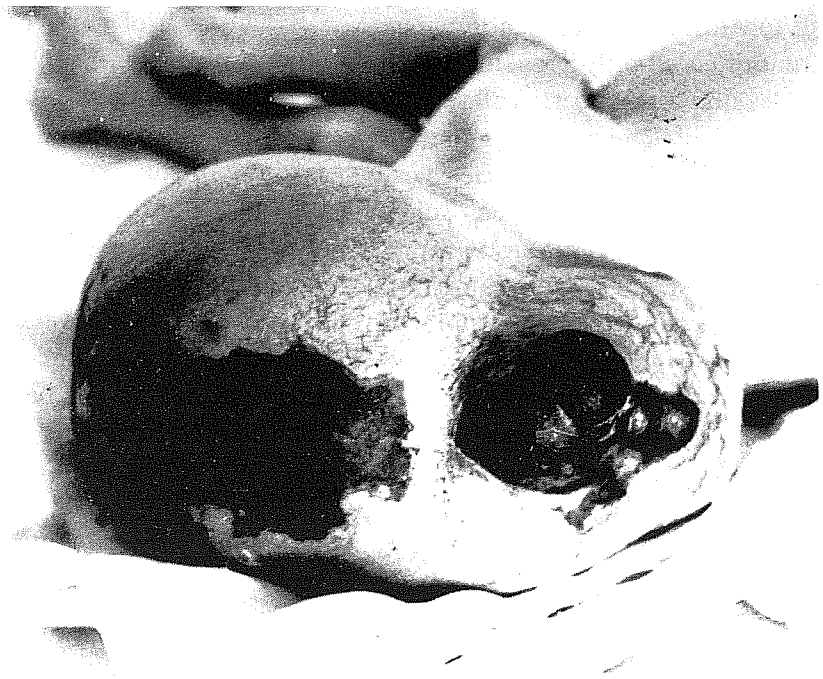

Figure 1– Preoperative apprearence of a giant Sacrococygeal teratoma

Figure 2– Postoperative view of the same patient

KAYNAKLAR

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