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Dysfunctional Elimination in Children Operated for Sacrococcygeal Teratoma

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ABSTRACT :

Introduction: *Sacrococcygeal teratoma is a rare condition (1:35.000-40.000 for the newborn). We report single-center experiences about it focusing on functional sequelae.*

Material Method: *From December 1997 to November 2007, 2 boys and 14 girls were treated in our department for Sacrococcygeal teratoma. Their hospital records were analysed retrospectively. Only 12 of these cases were evaluated prospectively, the others were not assessed because they were not available for follow-up. They were evaluated by a physical exam, an uroflowmetry-EMG and cystometry. A questionnaire was administered about bowel and voiding habits.*

Result: *9 out of 12 were operated at an average age of 8.4 days and were diagnosed in the neonatal period, their histopathology proving benign. Tumor manifestation occurred in 2 months, 11 months and 19 months of age in the remaining 3 children. Histopathology was mature in 8, immature in 3, malignant in 1 case. Surgery comprised tumor excision with coccygectomy. The child with the malignant tumor received chemotherapy after colostomy. During follow-up AFP levels were normal for all patients. None of children had reccurrences. Overall survival rate was 100%.*
5 children had voiding dysfunction. 3 children was voiding in urgency and also squatting down. 3 children had been reported for enuresis nocturna and 1 for enuresis diurna. Functional bladder capacity was low in 2 cases. One patient with a high functional bladder capacity had strained and interrupted voiding. Abnormal EMG potentials were observed in 4 patients. Detrussor instability was seen in 3 children. None of children had anal stenosis and fecal soiling, 4 had constipation. Satisfaction levels of parents about operation cavity and scar of their children were found 5 out of 12 null, 4 medium and 3 mildly high.

Conclusions: *Follow-up after surgery for SCT should include a careful examination of voiding and anorectal dysfunctions because of neuorourological and/or anorectal dysfunction. Detailed information about cosmetic reconstruction after teen age should be made available to the parents.*

Key Words: *Sacrococcygeal teratoma, Dysfunctional elimination, Child, Sequelae.*

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INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common tumor seen in the newborn occuring in 1 out of every with a 35.000 to 40.000 live births [1, 2]. The embryological origin of these tumors is the pluripotent cells in Hensen's node of the primitive streak and have components from endoderm, mesoderm, and ectoderm layers. The arrest or aberrant migration of the primordial germ cells can explain their midline distribution [3]. These pluripotent cells differentiate into embryonic (mature and immature teratomas) and extraembryonic (choriocarcinoma or yolk sac teratoma). The latter has increased risk of malignant transformation [4]. Metastasis is rarely found in the regional and abdominal lymph nodes, liver, lungs. Metastatic spread to the vertebrae and bone marrow is very rare [4]. The extragonadal teratomas most common site is the sacral region, which is followed by the anterior mediastinum, pineal, retroperitoneum, neck, stomach, and vagina [5].

According to the AAPS's (American Academy of Pediatrics, in Surgical Section) classification (Altman criteria); type I is primarily external and has only a small presacral component; type II is predominantly external

having a significant intrapelvic portion; type III is partially external but is predominantly intrapelvic with abdominal extension; type IV is entirely located within the pelvis and abdomen. The utility of this classification arises from the ease of surgical resection, antenatal detection and the probability of malignancy [6]. Compression or infiltration of sacral nerves by the tumor, intradural extension of the tumor, or surgical trauma to the pelvic splanchnic or hypogastric nerves acquired during resection of the tumor can damage the innervation of the lower urinary tract in SCT [7]. This article is designed to understand the functional sequelae in children operated for SCT. An assessment of cosmetic results was also made.

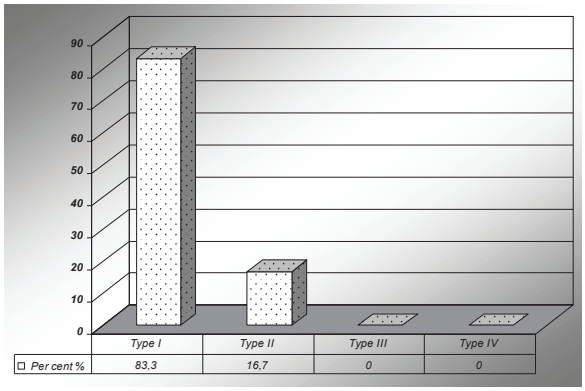
MATERIALS AND METHODS

From December 1997 to November 2007 a total of 16 children with sacrococcygeal teratoma were treated in the Pediatric Surgery Department at our hospital. Their hospital records were analysed retrospectively, considering the age at the time of operation, AFP values, operative findings, histopathology, recurrences and survival. Only 12 of these cases were evaluated prospectively for this study, the others were not assessed because they were not available for follow-up. They were evaluated by a physical exam, an uroflowmetry-EMG and cystometry. The uroflowmetry-EMG and cystometry of the patients were performed and evaluated by the same urologist. The questionnaire about bowel and voiding habits was developed by researchers taking into consideration present literature and used in face to face interviews. Analog scale graduated from 0 to 10 was used to determine scar tissue satisfaction levels (0-3 low, 4-7 medium, 8-10 high).

RESULTS

16 children comprising 2 male (12.5%), 14 female (87.5%) were treated for SCT in our department between December 1997 and November 2007. The male to female ratio was found approximately 1:7. Only 12 of these cases were evaluated prospectively for this study, the others were not assessed because they were not available for follow-up, so these 4 cases were excluded from this study. 9 patients of 12 were diagnosed in neonatal period and operated in 8.4±1.8 days of age. Tumor manifestation occurred in 2 months, 11 months and 19 months of age in the remaining 3 children, these patients were operated after the admittion.

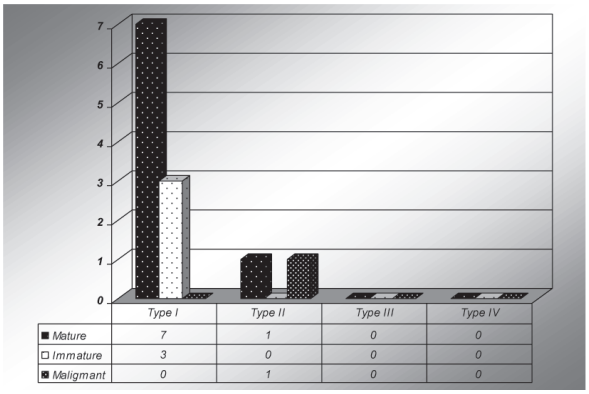
The tumors were subdivided and classified by the Altman classification according to their location 83.3% (n=10) showed Altman I tumor and 16.7% (n=2) had Altman II tumor [Figure I].



Şekil 1: . Staging of our SCTs cases according to Altman classification (type I to type IV).

Surgical resection was performed via a sacroperineal approach in all cases. After chevron incision, skin flaps were mobilised. Anorectal muscle sling was preserved. The wound reconstruction resulted in a predominantly transvers scar with buttock configuration. Surgery comprised tumor excision with coccygectomy. Only one child with the malignant tumor received chemotherapy after colostomy followed by tumor resection. During follow-up AFP levels were normal for all patients. Histopathology was mature in 66.7% (n=8), immature in 25.0% (n=3) and malignant (95% endodermal sinus tumor, 5% mature teratoma) in 8.3% (n=1) [Figure II]. An analysis covering all patients demonstrated following mean values: 6.41±1.5 for miction and 2.25±0.4 for defecation, as well as 8.3% (n=1) recurrent UTIs. Bowel and voiding function features were evaluated for only 9 patients. 3 patients having received no toilet training or being too young. Urinary function analysis showed 33.3% (n=3) urgency, 33.3%

(n=3) squatting down, 33.3% (n=3) enuresis nocturna and 11.1% (n=1) enuresis diurna. Bowel habits examination revealed 66.7% (n=6) normal bowel habits, 33.3% (n=3) constipation. One of these patients with constipation had hypothroidism also. Soiling or anal stenosis were not observed [Table II].



Şekil 2: Histopathologic findings of our cases according to Altman classification

Functional sequelae	n	%
Bladder dysfunction		
Urgency	3	33.3
Squatting down	3	33.3
Enuresis nocturna	3	33.3
Enuresis diurna	1	11.1
Straining	1	11.1
Bowel dysfunction		
Normal	6	66.7
Constipation	6	33.3
Soiling	-	-
Diarrhea	-	-

Tablo 2: Prevalence of bowel and bladder functional disturbances

%41.6 of cases (n=5) had voiding dysfunction. Functional bladder capacity was low in 2 cases. One patient with high functional bladder capacity had strained and interrupted voiding. Abnormal EMG potentials were observed in 4 patients. Detrussor instability was seen in 3 patients.

Mean points of analog scale (graduated from 0 to 10) used to measure scar tissue satisfaction levels of parents was 3.75. Satisfaction levels distribution was as follows: low (41.7 %), medium (33.3 %) and high (25 %) . The follow-up period ranged from 6 to 139 months. None of the children had recurrence. Overall survival rate was 100%.

DISCUSSION

SCT is the most common tumor observed in the newborn with an incidence of 1 in 35.000 to 40.000 live births. Approximately %75-80 of the cases had female origin [1, 2, 6 , 8, 9]. A male/ female ratio of 1/7 was found in our study.

Makin et al. series of 41 SCT cases showed 80% mature teratoma [10]. Rescorla et al. reported a large series of 126 SCTs from 15 centers seen over a 22-year period. 80 out of 126 patients had mature teratoma [11]. In our study, histopathology was mature in 66.6% (n=8), immature in 25.0% (n=3) and malignant (95% endodermal sinus tumor, 5% mature teratoma) in 8.3% (n=1). Tumor size does not affect malignancy. According to Altman criteria, SCT frequency is as follows: Type I 47%, type II 35%, type III 9%, type IV 9% [1]. Our study found the most frequent SCT being type I 83.3 % and type II 16.7 % . Results arising from our study proved consistent with the literature [Table 1].

Patient number	Gender	Operation age	Dimension	Altman classification	Histopathogy
Patient 1	Female	22/365	10*8*6 cm	Type I	Mature
Patient 2	Female	2 /365	12*9*7 cm	Type I	Mature
Patient 3	Female	2/365	11*8*4 cm	Type I	Mature
Patient 4	Female	1/365	11*10*8 cm	Type I	Immature
Patient 5	Male	2/365	15*11*9 cm	Type I	Immature
Patient 6	Female	3/365	10*7*5 cm	Type I	Mature
Patient 7	Male	2/365	8*5*3 cm	Type I	Immature
Patient 8	Female	2/12	4*2.5*2 cm	Type I	Mature
Patient 9	Female	19/12	7*6*5 cm	Type II	Malignant (95% endodermal sinus tumor, 5% mature teratoma)
Patient 10	Female	11/12	5*3*2 cm 4.5*2.5*2 cm	Type II	Mature
Patient 11	Female	16/365	5*4.5*3 cm	Type I	Mature
Patient 12	Female	26/365	6*4.5*3 cm 4*2*1.5 cm	Type I	Mature

Tablo 1: Distrubition of our cases according to gender, operation age, dimension of the tumor, classification type and hystopathology.

On the other hand, according to literature the re is a correlation between age and malignancy. Malignant tumor incidence was 5-10% below 2 months of age while it was 48-67% for older children [6, 8, 12]. Our study showed average ages as follows: 57.62 days for mature group (n=8), 1.66 days for immature group (n=3) and 562 days for malignant group (n=1). Despite limited number of

cases followed in our study malignancy was not observed under 2 months of age, which is consistent with existing literature [Table 1]. Bittmann's study demonstrates that poor cosmetic results in the buttock region were the most common long-term complication after surgery for SCT, possibly leading to more corrective operations in later life [13]. The study of Derikx also found low scar tissue satisfaction levels in 40% of cases. In our study, we witnessed low parent satisfaction about operation cavity and scar tissue (41.7 %) which confirms Bittmann and Derikx studies' related data.

The postoperative functional sequelae of SCT resection have been previously reported, although only a few publications deal with associated or subsequent impairment of bladder and anorectal functions. More than half of the surviving children with SCT had functional bowel disturbances. In addition, more than half of those with bowel disturbance had associated bladder-voiding disorders [14, 15]. The incidence of anorectal dysfunction in the literature was found in up to 40% [13, 16, 17, 18, 19]. Several studies found a wide range of 8% to 35% of patients with constipation [20, 21]. According to the study of Derikx designed to determine long-term complications of SCT, the constipation rate is 16.7% [22]. In our study, 6 patients (66.7%) reported normal bowel habits, whereas 3 patients (33.3%) complained about constipation [Table 2]. Frequencies of postoperative urologic sequelae ranging from 20% to 50% have been described [18, 23, 24]. Hedrick et al. reported urologic sequelae occurred in 43% of their patients. In our study we found that the patients had urgency (33.3%), squatting down (33.3%), enuresis nocturna (33.3%) and enuresis diurna (11.1%) [Table 2]. Retrospective study of a large series made by Rescorla et al. demonstrated 11% local recurrence rate for fully resected mature teratomas [11]. During our study, any of the children had recurrence. None recurrence may be explained by the inexistence of type III and type IV tumors in our series, and coccyx was removed in all patients. As a conclusion neurourological and/or anorectal dysfunction may expect in children after surgery for SCT. Follow-up after surgery for SCT should include a careful examination and detailed questionnaire for voiding and anorectal dysfunctions. Urodynamic study is necessary postoperatively. A better surgical procedure for closing the buttock region in the treatment of SCT should be considered. Parent should be made aware of reconstruction possibilities after teen age.

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