BENIGN SACROCOCCYGEAL TERATOMA: A FIFTEEN-YEAR RETROSPECTIVE STUDY

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Abstract- In spite of being histologically benign, sacrococcygeal teratoma (SCT) may recur either as a benign or malignant tumor. A total of 26 patients with benign neonatal SCTs were treated in Taleghanee and Mofid medical centers from 1986 to 2000. Investigations included radiography, abdominal ultrasound and computed tomography (CT) scan and measurement of tumor marker (α-fetoprotein). Initial surgical removal of the SCT (including the coccyx) was carried out during the first two weeks of life. One patient died on the first day of life following tumor rupture due to hemorrhagic shock before undergoing surgical intervention. Eight children had recurrences. Two were benign and six malignant teratomas, the latter having been benign on histology of the primary tumor. Five patients with malignant lesions required abdominosacral excision, two had a preliminary colostomy and chemotherapy followed by excision of the residual tumor and colostomy closure at a later stage, but in last one tumor was excised at the sacrococcygeal area. The overall follow-up ranged from 3 months to 13 years. There have been no complaints of functional neurological deficits after the operation. We conclude that SCT, although histologically benign, has an alarming potential to recur either as a benign or malignant tumor during the first 3 years of life, therefore, a close follow up for at least 3 years (physical examination serum α-fetoprotein and diagnostic imaging) is recommended for all patients who have undergone excision of SCT.

Acta Medica Iranica, 44(1): 33-36; 2006

Key words: Sacrococcygeal teratoma, yolk sac, α-fetoprotein

INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common congenital neoplasm (1), occurring in 1 in 40000 infants. Approximately 75% of the affected infants are female. This neoplasm is composed of a wide variety of tissues form all three germ cell layers alien to the anatomic site in which it arises (2). It often occurs near the coccyx. It is assumed to be derived from the pluripotent cells of Hensen's node located anterior to the coccyx. The teratomas form and grow during intrauterine life, and can become quite large with the growth of most sacrococcygeal teratomas paralleling the growth of the fetus.

In spite of being histologically benign, these tumors may recur either as a benign or malignant tumor (3, 4). This study was performed to evaluate potential of recurrence of these tumors.

MATERIALS AND METHODS

A retrospective study was carried out on the records of 26 patients with benign SCT who were admitted at the two university hospitals affiliated to Shaheed Beheshtee University School of Medicine. All patients were operated during the first two weeks of life except one who died on the first day of life following tumor rupture with hemorrhagic shock before surgical intervention. Each patient was

Received: 19 Oct. 2005, Revised: 19 Dec. 2005, Accepted: 22 Dec. 2005

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operated by inverted Chevron incision with its apex directed superiorly. Skin flaps at the superior margin of the tumor were elevated first and the tumor mass drop caudar. The mass was mobilized close to its capsule, widely displaced retrorectally. Muscles were carefully identified and preserved. The tumor was dissected free from rectal wall and removed with coccyx. The anorectal muscles were reconstructed. All excised tissues were histologically examined. Multiple sections were routinely cut. This was done at the time of the original resection.

RESULTS

Of 26 patients, 20 (76%) were female. The distribution of the tumor according to Altman's classification can be seen in table 1.

Seventy-two percent of the SCTs contained cystic components and 28% (7 SCTs) contained solid elements. The mean period of follow up was from 3 months to 13 years in 25 patients. Eight children had tumor recurrence, that were detected by physical examination or laboratory tests by means of α-fetoprotein evaluation or urinary or anorectal functional impairment. One child with a patulous anus presented with fecal soiling and two reported nocturnal enuresis. The rest of the patients presented with a sacral or an abdominal mass.

DISCUSSION

Teratomas in infants and children most commonly appear in the sacrococcygeal region. Less common sites are the mediastinum (5), testes, retro peritoneum, neck and stomach, as reported in the literature. Most of the infants are female and more than 60% are present at birth (6, 7). Our series correlate with these published data.

The surgical approach to SCT consists of complete removal of the tumor through the sacral area or a combined abdominosacral approach with the coccyx always being removed. The importance of removing the coccyx with the tumor was emphasized first by Gross (6) and then by others (4, 8, 9). High recurrence rates of up to 37% were registered if the coccyx had not been excised at the initial procedure (6, 10, 11). In all our cases the coccyx was removed.

Table 1. Altman distribution of SCT

Altman classification	No of patients	Percent
I	15	57.6%
II	10	38.4%
III	1	3.8 %
IV	0	0

Eight patients (30%) in our series developed recurrences in contrast with Waldhausen's series of 93 patients where a lower recurrence rate (7.5%) was reported (10). The size of the primary SCT was not found to correlate with a high recurrence rate by Altman *et al.* nor by Carney *et al.* (12). Also Gross did not consider the size of the primary SCT to be related directly to the likelihood of recurrence or poor outcome (6).

No postoperative death occurred in our series regardless of the size of the primary SCT. All the recurrent SCTs in our series occurred in patients whose primary tumors contained both cystic and solid elements, and no recurrent SCT occurred in the totally cystic SCT group in our series. Six patients in our study had been diagnosed benign as having mature SCT at the primary tumor resection but malignant components were documented in the recurrences. This phenomenon has been documented also by other authors (6, 10, 13). Attempts to explain this malignant transformation of mature and benign SCT were made by Willis who believed that late malignancy might be due to the retained capacity for continued growth at the embryonic level (14). Although multiple histological sections of these large primary tumors were read as mature and benign SCT, such large tumors may well harbor small foci of malignant endodermal sinus cells, which may delay its detection (3, 4).

In our study, the Altman staging of the primary SCT was not found to be helpful in predicting tumor recurrence in terms of benign or malignant status. All eight cases that recurred were initially staged as Altman stage II and I. Only one case was in stage III. Serum α-fetoprotein in the first month of life was not found to be of any prognostic signification in patients with SCT (15).

The mean serum α- fetoprotein level in this group was 13 ± 1 mg/ml. After total resection of the primary SCT together with the coccyx, an elevated

serum α-fetoprotein level has been found to be a reliable marker for recurrence of a poorly differentiated yolk sac tumor (16, 17).

In our series the mean serum α-fetoprotein level in the malignant recurrent group was markedly elevated despite the apparent absence of a clinically detectable tumor. The mean serum α- fetoprotein level was 7320 mg ± 4510 mg/ml. Radiographic investigations were promptly undertaken to locate the presence of a possible recurrent tumor. Every recurrence of SCT should be regarded as being potentially malignant. CT scan and MRI imaging are both reliable and helpful diagnostic modalities, which can add to the initial preoperative assessment in determining the anatomic relation of the tumor and the degree of trans-spinal tumor extension. We recommend that a preoperative CT is unnecessary in the neonate, but it is recommended in the recurrent tumor and to rule out the presence of distant metastases. We believe that every 3 to 6 months for at least the first 3 postoperative years, routine physical examination is essential and indeed the best means of detecting early recurrences. Prognosis of sacrococcygeal teratoma is improving due to prenatal detection, planned intra-partum management, prompt surgical resection, histological examination, routine physical examination and regularly serum α-fetoprotein levels measurement with multimodal chemotherapy. Overall survival for germ cell malignancies has improved from 84% to 94% (18, 19). Factors reportedly associated with a worse prognosis in malignant germ cell tumors include: 1. An extragonadal location; 2. Age greater than 11 years; 3. Extent of disease; 4. Inability to perform a complete resection; and 5. Germinoma or mixed germ cell histology (20).

Radiation therapy has been used in some situations when complete resection was prevented by involvement of vital structures. There is no conclusive evidence that radiation significantly improves overall survival for malignant germ cell tumors outside the CNS (21).

We conclude that SCT, although histologically benign, has an alarming potential to recur either as a benign or malignant tumor during the first 3 years of life; therefore, a close follow up for at least 3 years (including physical examination, serum α-fetoprotein and diagnostic imaging) is recommended for all patients who have undergone excision of SCT in the newborn period.

Conflicts of Interests

We have no conflicts of interest.

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