Sacrococcygeal teratoma in children: Single center experience

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Abstract

Introduction: Sacrococcygeal teratoma (SCT) represent the most common benign and malignant germ cell tumors (GCT) in newborns and infants developing from primordial germ cells of the human embryo. Most of them are benign, however those presenting later in life and having major intra-pelvic component have greater malignant potential. Early and complete excision of the SCT has been the mainstay of successful management. The aim of this study is to describe the patient's details, management done, histopathology of the tumor, recurrence and the over all outcome of patients with SCT.

Materials and Methods: Patients with histological diagnosis of sacrococcygeal teratoma during the period of January 2014 and October 2016 were included in this retrospective observational study. Data was collected regarding age, gender, mode of presentation, associated anomalies, Altman's classification, investigations, treatment modality, tumor histology according to type of tumour with margin of resection, and outcome of management. All the diagnosed cases of SCT presenting during the above duration were included for study.

Results: Twenty eight patients were included in the study. Out of them 75% were females and 25% were males. 32.14% presented during neonatal period. 78.57% presented with external mass. The tumour was completely resected by sacral approach in 22 patients and by combined abdominosacral approach in 06 patients. Recurrence was detected in one patients (3.57%); with Altman types 1 mature teratoma. Patients were followed for a duration ranged between 1 months and 3 years.

Conclusion: Early diagnosis and early complete enblock excision of the tumor with the coccyx was mainstay of treatment with good prognosis. Late presentation and the presence of malignant changes are associated with poor prognosis. Intraoperative spillage of the tumor should be avoided. Overall survival of SCT is high.

Keywords: Sacrococcygeal tumors, Germ cell tumor, Immature tissue.

Introduction

Germ cell tumors (GCT) are neoplasms that develop from primordial germ cells of the human embryo. Sacrococcygeal tumors (SCT) represent the most common benign GCT in children accounting for up to 70% of all GCTs.[1] Most germ cell tumors in the neonatal period are benign. They are divided into mature and immature teratomas.[2,3] World wide incidence of SCT is one in 35,000 to 40,000 live births.[4] During neonatal period mostly they present with a visible external sacral mass but some patient with intrapelvic tumors may present later. Females are affected more commonly. Associated congenital anomalies are seen in18%.[5-7]

SCTs was classified by Altman et al. into four types.[4] Type I tumors are almost external with minimal pelvic component; type II tumors have a significant intrapelvic component; type III tumors have a larger proportion of intra-abdominal and intrapelvic component than the external component; and type IV tumors are completely intrapelvic presacral with no external component. This study was a retrospective observational study to analyze the patient's details, surgical procedure, histopathology of the tumor, recurrence and the outcome of patients with SCT.

Materials and Methods

A retrospective analysis of neonates, infants and children who presented with SCTs between January 2014 and October 2016 to the Department of Paediatric Surgery, SPMCHI, Sawai Man Singh College, Jaipur was performed. Data was collected including age, gender, mode of presentation, associated anomalies, Altman's classification, investigations, treatment modality, tumor histology according to type of tumour with margin of resection, and outcome of management.

The initial diagnosis depended on clinical and radiological evidence of SCT and AFP level. Surgical types of the tumor as proposed by Altman were assessed. Surgical excision was performed through sacral or combined abdomino-sacral approaches according to tumor extension. Coccygectomy was performed for all patients.

The data collected from hospital records included findings on clinical examination like wound complications, any resection site mass and per rectal examination. Ultrasonography and serum AFP measured in every three months.

Results

A total of 28 patients with SCT were evaluated. There were 21 (75%) females and 7 (25%) males with a male to female ratio of 1:3. Age at presentation ranged between 1 days and 3 years. The diagnosis was made antenatally in one patient. 9 (32.14%) patients in neonatal and 13 (46.42%) patient during infantile period presented with visible external mass and diagnosed clinically (Type 1 & type 2). 6 (21.42%) patients presented after one year of age among them 3 had constipation and 1 had retension of urine (Type 3 & type 4) [Table 1]. None of the patient presented with ruptured tumour but one patient presented with mass which was biopsied outside.

On the basis of radiological investigations thirteen (46.42%) patients were Altman type 1, nine (32.14%) were type 2, three (10.71%) were type 3 and three (10.71%) were type 4 [Table 2]. No patient had intraspinal extension.

Associated congenital anomalies were found in two (7.14%) patients. One had small ASD and another had PDA.

One patient was biopsied outside which revealed yolk sac tumour were received three cycle of neo-adjuvant cisplatin, etoposide and bleomycin. Twenty two (78.57%) patients had complete tumor excision via the sacral route only (all type 1 and 2 patient). Six (21.42%) had combined abdomino-sacral resection of tumor (type 3 and 4). The coccyx was resected en block with tumour in all patients. Intraoperative rupture of the tumor with spillage during dissection occurred in four (14.28%) patients.

Tumour were solid in four (14.28%) cases, cystic in eleven (39.28%) cases, and in thirteen (46.42%) cases it was mixed solid and cystic on gross examination.

Histopathological reports showed that 20 (71.42%) patients had grade 0 pathology, four patients (14.28%) had grade I, two patients (7.14%) had grade II, and two (7.14%) had grade III [Table 3]. Platinum based drugs along with etoposide and bleomycin adjuvant chemotherapy was given in two patients with malignant immature component.

Patients were followed for a period ranged between 1 months and 3 years. Immediate postoperative complications; in the form of complete (n = 1) and partial (n = 4) wound dehiscence; were reported in 5 of 28 patients. All were successfully managed. One patient who had tumor excision suffered with transient urinary and faecal incontinence, again managed successfully. No patient required stoma for faecal diversion.

In most of cases AFP level measures decreased within 3 to 9 months.In recurrent cases it raised again. Recurrence seen in one patients (3.57%); which was Altman types 1 with mature teratoma. Recurrence detected after six months of initial resection in follow up by raised serum AFP. Re excision was done in type 1 with mature teratoma recurrence and histopathological finding of re excised specimen revealed yolk sac tumour. This patient received platinum based multidrug adjuvant chemotherapy and was well till 8 months follow up.

Table 1: Age of presentation of sacrococcygeal teratomas

Age of Presentation	No. of Patients (Percentage)
Neonates (<1 month)	09 (32.14%)
Infants (<1 year)	13 (46.42%)
Children (>1 year)	06 (21.42%)

Table 2: Altman classification of patients

Type	No. of Patients (Percentage)
Type 1	13 (46.42%)
Type 2	9 (32.14%)
Type 3	3 (10.71%)
Type 4	3 (10.71%)

Table 3: Gonzalez-crussi histopathologic grading

Grade	No. of Patients (Percentage)
Grade 0 (No immature tissue)	20 (71.42%)
Grade 1 (<10% immature tissue)	04 (14.28%)
Grade 2 (10%-50% immature tissue)	02 (7.14%)
Grade 3 (>50% immature tissue)	01 (7.14%)

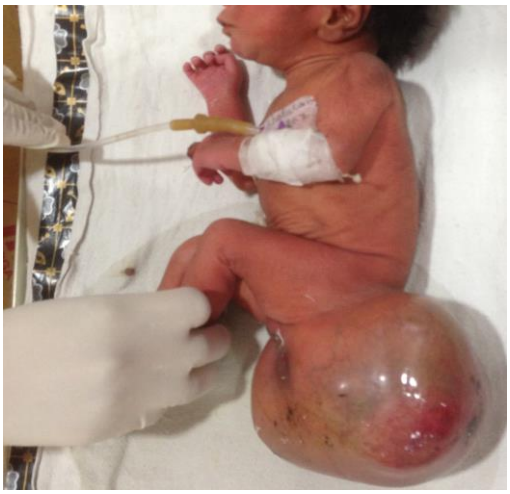

Fig. 1

Discussion

SCT is the most common tumor diagnosed in the neonatal period. Females are affected more commonly than males with female-to-male ratio of approximately 4:1.[6] In our series, female comprised of 75% of cases. This slight difference in our study may be due to relatively small number of patients. Teratomas in children originate most commonly in the sacrococcygeal region. Other sites are in the gonads, retroperitoneal region and rarely in the brain or liver.[8] In this study, associated congenital anomalies were found in 7.14% of our cases. Bittman S and Bittman V in their series of 25 patients reported 20% incidence of associated congenital anomalies.[9]

Antenatal diagnosis is important for a safe delivery and to avoid complications. In the current study, only one (3.57%) patients were diagnosed antenatally. In a previous series 20% of cases were diagnosed antenatally.[10] The reason for low antenatal diagnosis in our study may be due to low socio-economic status, lesser availability of tertiary are facilities at door step and ignorance regarding antenatal care. According to anatomical location and extension sacrococcygeal teratoma is classified by the American Academy of Pediatric Surgical Section. In the literature, Altman type I is present in 47%, type II in 35%, type III in 8% and type IV in 10% of cases.[4] In the current study, the incidence of the four types was 46.42%, 32.14%, 10.71%, and 10.71% respectively. This is comparable to that reported in the American Academy of Pediatrics survey.[4]

Previous studies have shown that there is significant relationship between outcome of treatment of SCT and age of the patients at the time of diagnosis. The incidence of

malignancy at the neonatal period is approximately 10%, against almost 100% at the age of 3 years.[11] Before 2 months of age, mostly the lesions are benign, and complete surgical excision with coccygectomy is associated with relatively low morbidity and mortality.[12] The use of alpha-fetoprotein (AFP) as a tumor marker is well established in literature, and persistent elevated level may be seen with residual tumor, recurrence, or malignant degeneration.[13] However, there is physiological rise of AFP levels till 8 months of age and it should be interpreted with caution in infants.[14] Only significantly higher levels are to be considered for further evaluation. After SCT resection the mean time required for AFP to be normalized is about 9 months.[15]

Type 1 and type 2 SCT patients present early with external mass, while delayed diagnosis may occur in type 3 and 4 patients, as the presacral mass may be unnoticed. In the current study, 22 patients presented with external mass. These patients had either type I, II or III SCT. Six patients presented late and all of them had type III or IV SCT. Rescorla et al reported that all children presenting after the age of one year had malignant tumors.[12] Gabra et al reported that after the neonatal period 71% of their patients presented with malignancy.[16] In our study, 7.14% of the cases were malignant and all of them presented after the neonatal period.

Gonzalez-Crussi graded SCTs as follows: grade 0, tumor contains only mature tissue; grade I, tumor contains rare foci <10% of immature tissues; grade II, tumor contains moderate quantities 10%-50% of immature tissues; and grade III, tumor contains large quantities>50% of immature tissue with or without malignant yolk sac elements.[17] Unlike of ovarian teratomas grading of SCTs, doesnot seem to correlate directly with prognosis.[18] In our study, two patients had malignant SCT, one was grade II and another was a grade III histopathology with yolk sac tumour. The primary treatment of SCTs is early complete surgical resection with excision of the coccyx.[13,19] Adjuvant chemotherapy in the form of combination of cisplatin, etoposide and bleomycin is used in malignant cases.[20] Early surgical intervention is associated with better prognosis.[12,19] In our study, the patient with malignant disease presented late and one patient developed recurrence.

The surgical approach depends on the size of mass and degree of pelvic extension.[4] Surgical excision was done by using a posterior sacral approach in 22 (78.57%) patients (all type I and II cases) or combined abdominal and posterior sacral approach in the remaining six patients (types III and IV). Extension into the spinal canal may cause paraplegia but intradural invasion of a SCT is a rare.[21] There was no case of SCT with intraspinal extension in any of our patient.

Depending on the amount of immature elements; mostly neuroepithelium, the teratomas are divided into immature and mature tumors.[22] Tumor recurrence was reported in 2- 35% of patients in the various studies. Incomplete surgical excision with the presence of microscopic residual tumour, failure of the en-block removal of the coccyx along with the tumor, tumor spillage or the presence of immature component are the main reasons for tumour recurrence. Outcomes after surgical resection during the neonatal period are generally good, but malignant transformation can occur in 10–20% of patients.[23] Early and complete excision of the SCT has been the mainstay of successful management.[24,25] A multimodal approach involving surgery and platinum-based chemotherapy has resulted in a significant improvement in prognosis in both localized and metastatic malignant tumors.[13]

After complete surgical excision and coccygectomy mature teratoma does not recur.[26] De Backer et al reported 0-26% recurrence rate for mature teratomas and 12-55% for immature teratomas.[26] In our study, Recurrence of the tumor occurred in 1 patients (3.57%). This patient had mature teratoma presented with recurrence in which re-resection was done and biopsy revealed yolk sac tumour. In our study, recurrence was diagnosed by clinical examination, radiological investigations and increased AFP. Many authors have recommended the use of AFP as a tumor marker in the follow up for these patients.[27] Residual or recurrent tumor may be indicated by persistently elevated serum AFP levels. On the other hand, Brewer and Tank reported that the level of AFP is normally elevated in the first 8 months of life so high levels should be interpreted with caution in infants.[14]

Conclusion

We conclude that most cases of SCT present in early life with benign nature and malignant lesions are seen beyond infantile period. This study stressed on the importance of early diagnosis, complete enblock resection of the tumor and postoperative management based on histological findings. Close follow-up with clinical examination, radiological investigations and serum AFP should be done. Compliance with follow up visits requires proper parental counselling and tertiary are services at doorsteps.

Conflict of Interest: Nil

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