Original Article

Management of Sacrococcygeal Teratoma in Infants and Children

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ABSTRACT

Objective: To discuss the management of SCT in infants and older patients with special attention to per operative, post operative complications like tumor rupture, GIT trauma, neurological function and recurrence.

Study Design: A retrospective descriptive

Place and Duration of Study: This study was conducted at the pediatric and neonatal surgery Bahawal Victoria Hospital from March 2017 to March 2022.

Materials and Methods: Patients under 2 months and older children (2mnths to 5 year) with SCT were included in the study. All patients were assessed for any associated anomalies. Tumour excision was done either through sacral route only or abdominosacral approach depending upon size and type of SCT. Patients were observed for any perioperative and postoperative complications and then later on follow-up for 30 months.

Results: Twenty-five patients with SCT were managed.20 patients under 2 month of age and 5 above 2months upto 5 years of age. Sacral approach (22 patients) was used in small size tumor and abdominosacral approach (3 patients). on histopathology 18 benign and 7 malignant. Patients with Sacral approach all good with fecal and urinary continence whereas sacro abdominal approach showed some form of incontinence. Survival good after chemotherapy.

Conclusion: SCT has a good prognosis if operated early with increased rate of complication and malignancy as age of patient and type of tumor increases.

Key Words: SCT, tumor, abdominosacral.

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INTRODUCTION

The commonest tumor in neonates is the Sacrococcygeal teratoma (SCT)with 1/40000 live birth incidence.[1] SCT accounts for approximately 70% of all pediatric teratomas with female to male ratio is 4: 1.the rate of malignancy increases as the age at diagnosis is increased, it is about 10% - 20%if diagnosed before the age of 6 months and after six months it is increased to 65 % approximately.[2]

The cause for the tumor or its embryological explanation is not clear. The commonest site for teratoma is the sacrococcygeal area with the 2nd most being the gonads.[3] totipotent cells of primitive knot are considered the site of origin which then settle at coccyx.

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According to anatomical location SCT is classified into four types which was described by Altman.[4]

Type I: Teratoma with little presacral component

Type II: predominantly external tumor with definite presacral component

Type III: predominantly intra abdomen (presacral) with littlel external component

Type IV: The tumor is solely presacral with no external component

Staging according to COG, stage 1tumor entirely removed with normal tumor marker, stage 11, microscopic remnants after surgery left, and tumor markers don't return to normal after surgery, stage 111, visible tumor left after treatment and lymph nodes positive stage 1V the tumor spread to other organs / sites.[4]

The close association of the SCT with surrounding sacral neurovascular and pelvic organs make it a challenging surgery as regards to hemmorrage, neurological dysfunstion of urinary bladder and fecal continence and recurrence of the tumor.[5] Some centers do a pre-operative (about 36 hours before surgery) devascularisation of the tumor by ligation of the internal iliac and middle sacral arteries and thus lessen the per operative bleeding.[6] This retrospective study was conducted to share our experience in respect of complications associated with management of SCT.

MATERIALS AND METHODS

Approval from ethical review committee was taken and study conducted at department of pediatric and neonatal surgery from March 2017 to March 2022. Age at presentation of tumor was noted and tumor was catogarised according to the type as described by Altman. Patients with any bleeding disorder were excluded from study. Digital rectal examination was performed in all patients. Clinical examination to rule out other associated anomalies and cardiac evaluation done. Neurological examination was done in all patients. Ultrasonogaphy abdomen and CT Scan abdomen /pelvis done to detect the extent of the tumor and its nature (solid , cystic or mixed). Alpha fetoprotein was performed in all patients. Surgical excision performed and mass sent for histopathology to know its nature weither benign tumor or malignant. Malignant tumors were staged according to Fredrick et al saging system into four stages.[7]all the patients were followed for 12 month to 30 months for any recurrence and evaluation of fecal and urinary continence. Malignant teratomas were sent for chemotherapy and followed up by alpha fetoprotein, USG and CT Scan abdomen.

Surgical procedure: patient is positioned in prone jackknife with buttock lifted up , a rectal tube or Hegar dilator is passed per rectal for assessment of rectum during anterior dissection . a shevron incision is made after aseptic drapping . skin flaps are reised and tumor along with coccyx is rmoved extracapsular with great care to prevent from hemmorrage , rupture of tumor and rectal injury. Pre sacral artery branch from the spinal artery is identified and ligated . levator ani muscles are carefully separated as its role in continence.this approach is used for type 1 and type 2 SCT. For patients with type 3 and type 4 combined abdominal and sacral approach is used.[8]

RESULTS

Twenty-five patients with SCT were included in the study, among these 18 were female and 7 were male (table 1). Twenty patients presented under 2 months of age of whom 18 were benign and 2 malignant. Five patients were older than 2 months up to 5 years with 4 malignant and 1 benign.(table 2) type 1 SCT were found in 12 patients (1 malignant), type II in 6 patients with 2 malignant, type III among 4 patient with 2 malignant and 2 benign, type IV in 3 patients 1 benign and 2 malignant (table 3) presented at late age with c/o constipation, abdominal distension, tumor suspected on digital rectal examination and confirmed on USG and CT Scan. The main clinical findings were mass at sacrococcygeal area which was palpable on rectal examination. Palpable abdominal mass 4 patients, bowel disfunction was found in 7 patients and urinary complaints were seen among 9 patients. 22 patients were approached through sacral incision and the remaining by abdomino sacral incision. The coccyx was removed in all patients. The excised tumor sent for histopathology shwed 18 mature/immature teratoma and 7 patients were malignant (yolk cell tumor) with stage I= 3 ,stage II= 3 stage IV =1 patient. Among these 2 were under 2 months and 5 were older. Chemotherapy (bleomycin and cisplatin) was given to all 7 patients as advised by pediatric oncologist. Wound infection was noted in 7 patients, wound dehiscence among 5 patients. all were managed conservatively. Post operative bowel and urinary dysfunction noted in 4 patients. Rectal perforation noted in 1 patient and tumour rupture in 2 patients, both large size tumor which on histopathology came out as mature teratoma 1 and immature in 1 patient.one patient expired. Follow-up was done up to 36 months. there was recurrence among 3 patients, 1 due to lost to follow-up for 1year .(table 4) the alpha feto protein level was significantly raised.

Table No. 1: %age of patients according to gender (n=25

Gender	No. of patients	%age
Male	7	28
Female	18	72
Total	25	100

Table No. 2: Age distribution with malignancy n.25

Age	malignant	benign	No. of patients	%
< 2months	2	18	20	80
>2 months to 5 year	5	0	5	20
Total	7	18	25	100

Table No. 3: %Age of patients according to anatomical type with %age of malignancy (n=25)

Type	Malignant	Benign	No.of patients	%age
I	1	11	12	48
II	2	4	6	24
III	2	2	4	16
IV	2	1	3	12
Total	7	18	25	100

Table No. 4: Outcome of patients (n.25)

Complication	No. of patients
Tumor rupture	2
Wound dehisence	5
Rectal injury	1
Bowel / urinary dysfunction	4
Recurrence of tumor	3 (2, yolk sac, 1, immature teratoma)
Total	15

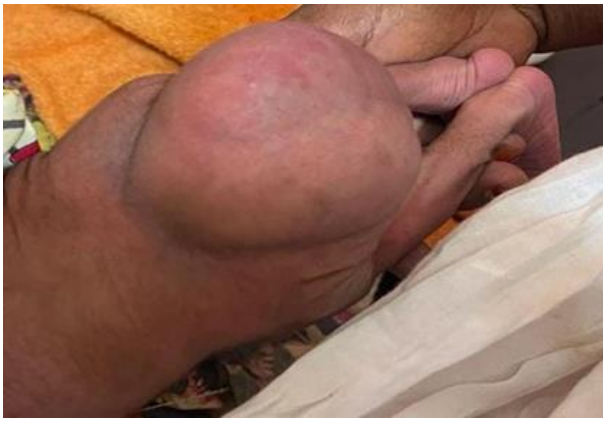

Figure No. 1: A 11-day old baby with SCT.

DISCUSSION

Pluripotent cells of Hensen's node popresemt between coccyx and anus is the most common site of extragonadal teratoma (SCT).[9]as described by Altman et al type 1, 11. 111, patients usually present early because of the tumor location at sacrococcygeal area , whereas type 1V present late as the tumor remain unnoticed being hidden in pre sacral (pelvic) area.[10] similarly in our study out of 25 patients 20(80%) presented under 2 months of age and among these only 1(5%) patient was of type 1V. Five patients presented after 2 months of age, among these 2 type 1V, 2 type 111 and 1 type 1. In our study rate of malignancy is increased with the increasing age at diagnosis. The studies conducted showed that most of the tumors(SCT) about 75%to 85% diagnosed during neonatal life were benign as compared to the late diagnosis i.e after nonatal period, where 70% of the patients were malignant.[11] the occurrence of malignancy in SCT as stated by Altman et al is related to patient's age at presentation and its excision, The rate of malignancy increases with lesion which may remain unnoticed during neonatal period due to their pre sacral presence i.e. (type IV) . They reported low rate of malignancy among patients under 2 months of age about (5–10%) which may peak upto 70% in patients above 2 months of age.[4]

A female prepondrance was noted about 2,6: 1 in our study. Yadav DK et al reported a 3.1: 1 female to male ratio in their study with a median age of 36 days(1 day to 11.6 years).[8].

Most of the patients(88%) presented with mass at sacrococcygeal area but 3 patients presented with constipation and dribbling of urine and diagnosed on DRE and USG abdomen all 3 were type 1V . Hasbay B et al reported constipation as the common symptom in their study among type 1V patients similar to our study.[4] Pediatric patients with SCT which present late usually don't have an external portion noted at birth, and their general clinical presentation is related to symptoms /signs of bladder or rectal compression.[4,8,12]

SCT are managed by surgical removal either through sacral approach or sometimes abdominosacral approach with removal of coccyx.The importance of removing the coccyx with the tumor was first emphasized by Gross et al.and then by others[5,7,9,13]. High recurrence rates of up to 37% were registered in case the coccyx had not been excised at the initial procedure[13]. In our study the recurrence rate was 3(12%) out of 25 patients. One patient who lost the follow up for upto 1 year and without proper chemotherapy and came back with recurrent mass at sacral area. A study conducted by Hasbay B showed recurrence in 2 patients out of 38 studied with death of 1 patient.[4] recurrence rate was 7.2%(3/41) in a study conducted by Yadav DK with 1 pt of imature teratoma and 2 yolk sac tumor having overall survival of 95%.[8] in our study recurrence was observed as 3(12%) out of 25 with overall 96%survival. There were injury to rectum in 1 patient and tumor rupture in 2 patients in our study which is similar to study conducted by Hasbay B et al. [4] Post operative bowel and urinary dysfunction observed in 4 patients. high recurrence rates,urinary ,fecal incontinence and neurological problems are the difficulties encountered by surgery for SCT[4,8,9] Wound infection observed in 7(28%) patients with wound dehesence among 5(20%) patients in our study. Similar results noted in a study conducted showing 23 % wound infection who underwent sacral approach.[14,15] In the early postoperative period the commonest complication noted is wound infection /de- hiscence; which may be due to close position of tumor to the anus. The rate of wound dehiscence observed was 7.3% although wound dehiscence rates as high as 90% have been reported in literature.[16]preoperative bowel preparation is claimed to be somewhat effective in decreasing the wound infection / dehesence.[17]

CONCLUSION

SCT has a good prognosisif operated early with increased rate of complication and malignancy as age and type of tumor increases. Excision only is sufficient for benign tumors whereas for malignant tumors after the improvement in chemotherapy survival is excellent with strict followup.

Author's Contribution:

Concept & Design of Study:	Muhammad Ramzan
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Data Analysis:	Soofia Mustafa
Revisiting Critically:	Muhammad Ramzan,
Final Approval of version:	Muhammad Ramzan

Conflict of Interest: The study has no conflict of interest to declare by any author.

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