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Original Research Article

FUGITIVE PROBLEM OF SACROCOCCYGEAL TERATOMA IN CHILDREN OF KASHMIR, A DETAILED CLINICAL STUDY AND ROLE OF SURGERY WITH SPECIAL EMPHASIS ON COMPLICATIONS

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ABSTRACT

Background: Sacrococcygeal teratoma (SCT) is the most common congenital germ cell tumor, with an incidence of 1 in 35,000-40,000 live births and a female predominance. At birth, the great majority of SCTs are benign. Approximately 70% of SCTs are malignant at the patient age of nine months. Therefore, prompt surgical resection is required and imaging studies play an important role in confirming the diagnosis, demonstrating intra-abdominal extension and effects on adjacent structures.

Material and Methods: a) Study was conducted in the Department of Pediatric surgery, Sheri Kashmir Institute of Medical Sciences, Srinagar. All children of less than 14 years of age and presenting to the department with sacrococcygeal teratoma were included. The ultrasonography delineated the size, extent, the shape, consistency solid and cystic components, intralesional calcifications as well as the status of other pelvic and abdominal organs. All patients were subjected to Magnetic resonance imaging (MRI) abdomen and pelvis.

Results and Conclusion: A total of 25 children less than 14 years of age with post natal diagnosis of Sacrococcygeal teratoma admitted. The age of patients included in this study ranged from 0.03 (1 day) to 168 months (14 years), with a mean age of 23.68±1.47 months (median age 8 months). Majority (64%) of the patients were aged less than 12 months. The mean age of presentation was 16.48±6.27 months (median 4 months) and in 48% (12 cases) age was less than 12 months. A prenatal diagnosis was found in 8% patients. Among 25 patients' included in this study, sacral swelling was the most common clinical sign observed in 88% patients (n=22), followed by constipation which was observed in 12 % cases (n=3) and intra-abdominal extension in 8% cases (n=2). The alpha-fetoprotein (AFP) level of the patients ranged between 4.9 to 2479ng/ml. Females are more likely to suffer from sacrococcygeal teratoma. Most of patients had Altman type I, and mature teratoma. Surgery was the mainstay of treatment for Sacrococcygeal Teratomas. Collection at wound site was the most common complication observed in current study (38.46%), followed by wound site infection (23.08%). Serum alpha-fetoprotein is a good diagnostic indicator for recurrence. Patients treated early in life have a better prognosis.

Keywords: Sacrococcygeal teratoma (SCT), alpha-fetoprotein (AFP).

INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common congenital germ cell tumor, with an incidence of 1 in 35,000-40,000 live births and a female predominance (F:M=3:1-4:1 ratio). At birth, the great majority of SCTs are benign. Approximately 70% of SCTs are malignant at the patient age of nine months. Therefore, prompt surgical resection is required and imaging studies play an important role in confirming the diagnosis, demonstrating intra-abdominal extension and effects on adjacent structures.[1,2,3] The tumor has been classified according to the system developed by Altman and colleagues based on the location and degree of intrapelvic extension. The prognostic factors include age at diagnosis, Altman type, gender of the child and histopathological type. The mass may be as small as a few centimeters in diameter or as massive as the size of the infant. Lesions with growth predominantly into the presacral space often present later in childhood. Patients in whom SCT is diagnosed postnatally typically do well after early surgical resection. The preferred approach to a small SCT is through the perineum. The cure rate is excellent if the tumor is excised completely. The endeavor of our study is to evaluate these aspects in our institute, thus filling this gap in knowledge.

MATERIALS AND METHODS

Study was conducted in the Department of Pediatric surgery Sheri Kashmir Institute of Medical Sciences, Srinagar. Total study duration was five years (2015 to 2020). All children of less than 14 years of age and presenting to the department with sacrococcygeal teratoma were included. Detailed history & clinical examination was followed by laboratory investigations, tumor markers and radiological investigations. Among tumor markers Alpha-fetoprotien (AFP), Beta- Human Chorionic Gonadotropin (β-hCG), lactate dehydrogenase (LDH) levels were measured and noted which were compared with post-operative levels. The USG delineated the size, extent, the shape, consistency solid and cystic components, intralesional calcifications as well as the status of other pelvic and abdominal organs. All patients were subjected to Magnetic resonance imaging (MRI) abdomen and pelvis. Fine needle aspiration cytology (FNAC) was done in some of the patient whereas others were diagnosed on strong clinical suspicion and radiological findings. Chevron incision was made in most cases. Standard surgical technique was adopted for resection. Tumor was removed enbloc followed by coccygectomy. After satisfactory hemostasis, pelvic floor muscles were reapproximated in midline behind the rectum. Drain was kept in tumor bed getting out of edge of wound, which was removed subsequently within few days in most cases. Early complications were managed appropriately, and patients were discharged to be followed up by a gap of one week. The patients were followed up for two years. The findings noted included in all patients were clinical examination findings, AFP levels and USG findings. In patients with suspicion of recurrence, Contrast Enhanced Computed Tomography (CECT) was done on need basis. The data was collected and analysed.

RESULTS

A total of 25 children less than 14 years of age with post natal diagnosis of Sacrococcygeal teratoma admitted.The age of patients included in this study ranged from 0.03 (1 day) to 168 months (14 years), with a mean age of 23.68±1.47 months (median age 8 months). Majority (64%) of the patients aged less than 12 months. The mean age of presentation was 16.48±6.27 months (median 4 months) and in 48% (12 cases) this was less than 12 months. Among the studied patients majority (80%) were females and rest (20%) were males with a male to female ratio of 1:4. A prenatal diagnosis was found in 8% patients. Among 25 patients' included in this study, sacral swelling was the most common clinical sign observed in 88% patients (n=22), followed by constipation which was observed in 12 % cases (n=3) and intra-abdominal extension in 8% cases (n=2).

The radiological findings of the patients conformed to the published literature. It also revealed that the lesion in most (8 out of 13) of the patients with Altman type I sacrococcygeal teratoma was heterogenous in nature.

Table 1: Radiological findings

Altman Type	No. of patients	Lesion (on USG)	Lesion (MRI)
I	13	Solid and cystic mass lesion in pelvis arising from spine	Heterogenous mass in presacral area with large external component
II	7	Solid with Cystic component mass lesion in pelvis with extra pelvic component	Heterogeneously enhancing mass with intrapelvic and extra pelvic component
III	2	Complex mass in pelvis with abdominal extension	Intrapelvic mass extending to abdominal cavity anterior to the sacrum and coccyx
IV	3	Solid cystic mass in presacral area	Large presacral mass with septated cystic and solid components arising from coccygeal area
Total	25	-	-

The alpha-fetoprotein (AFP) level of the patients ranged between 4.9 to 2479ng/ml. For simplification of results the patients were categorized into two groups based on whether AFP was less than 100ng/ml or more than 100ng/ml. Among patients with AFP count of more than 100, the mean was 2479ng/ml including 6, 1, 1 and 1 patient from age groups' 0 to 6 months, 6 to 12 months, 18 to 24 months and 24 to 30 months respectively. Similarly among patients with AFP count of less than 100, the mean was 16.71ng/ml, with 6,3,3,1 and 3 patients from age groups' 0 to 6 months (mean 20.16ng/ml), 6 to 12 months (mean 17.66ng/ml), 18 to 24 months (mean 14.3ng/ml), 24 to 30 months (mean 12ng/ml) and more than 30 months (mean 19.46ng/ml) respectively.

Table 2: Alpha-fetoprotein level in patients

Age (months)	Total No. of patients	No. of patients with raised AFP	Reference value (Blohm *et al*., 1998)[35]
<2 Months	10	0	9500 ng/ml
3-7 Months	2	1	33-88ng/ml
≥8 Months	13	11	8.5ng/ml
Total	25	12	-

The lactate dehydrogenase (LDH) levels of studied patients ranged from 6.2 U/L to 1166U/L with a mean of 200.49±58.98U/L.

Table 3: Lactate Dehydrogenase level in patients

Age (months)	Total No. of patients	No. of patients with raised LDH	Reference value (U/L)
<1 Months	10	1	125-765
1-12 Months	6	1	170-450
≥12 Months	9	1	135-225
Total	25	3	

Beta-Human Chorionic Gonadotropin (β-hCG) level of the studied patients ranged from 0.01 mIU/ml to 33mIU/ml.

Table 4: Beta- Human Chorionic Gonadotropin level in patients

Age (months)	Total No. of patients	No. of patients with raised β-hCG	Reference value (mIU/mL)
<3 Months	10	0	<50
>3 Months	15	4	<5
Total	25	4	-

Among 25 patients upfront surgery was done in 23 (92%) cases out of which 19 were females and 4 were male children. Fine needle aspiration cytology was done in two cases with suspicion of malignancy which revealed malignant germ cell tumor (GCT) in one patient and yolk sac tumor (YST) in another patient. Both these patients were subjected to NACT followed by surgery (Table 12).

Table 5: Treatment modalities used in patients

Treatment modality	No. of patients	Percentage
Surgery	23	92%
NACT followed by surgery	2	8%
Total	25	100%

There were a total of 13 complications recorded in patients following surgery. Collection at wound site was the most common complication observed in current study (38.46%), followed by wound site infection (23.08%). Two instances of bleeding and flap necrosis (each) were observed (15.38%), and only one instance of sepsis (7.69%) was seen in one of the studied patients.

Table 6: Intra and post-operative complications

Complication	No	%age
Bleeding	2	15.38
Collection	5	38.46
Wound infection	3	23.08
Flap necrosis	2	15.38
Sepsis	1	7.69
Total	**13**	**100**

Findings of clinical examination and serum AFP levels were monitored at one month, three months, six months, one year and two years. Up to one year uneventful course was observed in all but two patients which had elevated AFP level at one year of surgery (Table 14). These patients were subjected to Contrast Enhanced Computed Tomography (CECT) of abdomen and pelvis which revealed recurrence.

FNAC in both these patients revealed malignant transformation. Both these patients were referred to Department of Medical Oncology, SKIMS for adjuvant therapy.

Table 7: Post-operative course of patients

Condition	Recurrence		Expired	
	No.	%age	No.	%age
Yes	2	8	2	8
No	23	92	23	92
Total	25	100	25	100

DISCUSSIONS

Age of patients included in this study ranged from 0.03 (1 day) to 168 months (14 years), with a mean age of 23.68±1.47 months (median age 8 months). Majority (80%) of patients were females with a male to female ratio of 1:4. Contrararily Barakat et al., (2011) reported that 82% patients admitted for SCT were in the first 2 months of life.[4] Chirdan et al., (2009) also reported a lower that median age (7 days) and body weight (2.8 kg) among children with SCT; and a male to female ratio of 1:5.[5] A prenatal diagnosis was found in 8% patients. The most common clinical presentation was sacral swelling (88% patients) followed by constipation (12% cases) and intra-abdominal extension (8% cases). Ashcraft et al., (1974) reported the commonest mode of presentation is sacrococcygeal swelling with /without constipation and urinary retention.[6] The alpha-fetoprotein (AFP) level of the patients ranged between 4.9 to 2479ng/ml. The lactate dehydrogenase (LDH) levels ranged from 6.2 U/L to 1166U/L (mean 200.49±58.98U/L) and beta-hCG level ranged from 0.01mIU/ml to 33mIU/ml with a mean of 9.74±9.25 mIU/ml. The AFP levels were raised in 12 (48%) patients; LDH in (12%) patients and β-hCG in 4 (16%) patients. Three patients had all three markers raised that included one case of immature teratoma, one case of yolk sac tumor and one case of malignant germ cell tumor. Another patient with immature teratoma had elevated AFP and β-hCG levels. Agreeing with finding of this study, Havránek et al., (1992) reported that AFP levels were raised only in 45% patients with sacrococcygeal teratomas and β-hCG level was raised only in malignant testicular teratomas.[7] Contradictorily Göbel et al., (1998) and Harms et al., (1986) reported that evaluation of AFP in serum aids in establishing diagnosis of teratoma or germ,cell tumors and therefore, evaluation of serum AFP prior to surgery is mandatory in all children with suspected teratoma or malignant GCT.[8,9] Two patients with germ cell tumor and yolk sac tumor had elevated β-hCG levels.[9] In agreement with this were the findings of Frazier et al., (2008) who reported that in case of pediatric germ cell tumors (GCT) elevated serum β-hCG concentrations represent a good diagnostic criterion.[10] Out of the studied patients more than half were of Altman type I (52%), followed by Altman type II (28%). The number of cases with Altman type III and IV were 12% and 8% respectively. In agreement with this study Dirix et al., (2015) in a study on 205 patients with SCT reported that most (35.1%) were Altman type I followed by type II (23.9%), 13.7% were type III and 23.4% were type IV) (26). De Backer et al., (2006) also reported that majority patients in his study had Altman type I (36%) followed by type II (27%).[11] Histopathology was done in all the patients, which revealed mature teratoma in 21 (84%) patients followed by immature teratoma which was reported in 2(8%) patients. Malignant germ cell tumor and yolk sac tumor was reported in one patient each. Gabra et al., 2006 in concurrence to this reported that in 79% sacrococcygeal teratomas, histopathology is benign.[12] Among 25 patients upfront surgery was done in 23(92%) cases out of which 19 were females and 4 were male children. In two cases (one case each of yolk sac tumor and malignant germ cell tumor) neo-adjuvant chemotherapy (NACT) followed by surgery was offered. Smith et al., (1961) also reported that excision at first instance should be the intervention of choice in mature teratomas.[13] There were a total of 13 complications recorded in patients following surgery. Collection at wound site was the most common complication observed in current study (38.46%), followed by wound site infection (23.08%). Two instances of bleeding and flap necrosis (each) were observed (15.38%), and only one instance of sepsis (7.69%) was seen in one of the studied patients. Yadav et al., (2020) reported that early complications included wound infection (12.2%), superficial wound dehiscence (4.9%), complete dehiscence (2.4%), copious or prolonged serosanguinous drain output (2.4%), and urinary infection (2.4%) (34 Among 25 patients recurrence was seen among two patients. The serum AFP level was raised in both of these two patients by 1 year of surgery.

Pertinently both of these patients aged more than 2 years. Two patients (age >2.5 years) expired before the end of this study. One of these patients had immature teratoma, while other had yolk sac tumor on FNAC. Isaacs (1997) reported that there is a relationship between age of the patients at diagnosis and outcome of treatment of SCT and incidence of malignancy at the neonatal period is approximately 10%, against almost 100% at the age of 3 years.[14]

Havránek et al., (1992) also reported that serum AFP level is a sensitive marker for tumor recurrence and can be valuable for monitoring the chemotherapy.[15]

CONCLUSION

Females are more likely to suffer from sacrococcygeal teratoma. Most of patients had Altman type I, and mature teratoma. Surgery is the mainstay of treatment for sacrococcygeal teratomas. Serum alpha-fetoprotein is a good diagnostic indicator for recurrence. Patients treated early in life have a better prognosis.

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