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ORIGINAL ARTICLE

Pediatric Germ Cell Tumors: An Experience of 7 Years in a Tertiary Hospital of Bangladesh

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Abstract

Introduction: *Germ cell tumors are a group of tumors with different clinical presentation and histological and biological characteristics. Malignant germ cell tumors occur at all ages with a trend of bimodal distribution in infancy and adolescence.*

Objective: *To evaluate the demographic characteristics, distribution of different types of germ cell tumor, treatment modalities and outcome of germ cell tumor in children in a tertiary care hospital of Bangladesh.*

Methods: *In this retrospective study, data regarding age and sex distribution, location, types of tumors, management of germ cell tumor in children were retrieved from the medical records of pediatric oncology department in NICRH, Dhaka from 2008 to 2014.*

Results: *Out of total 87 patients female were 50 and male 37. Most of the patients were up to 5 years of age. The gonadal germ cell tumors (80%) were more than extragonadal tumor (20%) in both male and female patients. The most common germ cell tumor was dysgerminoma (32%) followed by yolk sac tumor (29.8%) and teratoma (19.5%). Yolk Sac Tumor (51.4%) was the most common in male and dysgerminoma (56%) the commonest in female. Out of 87, seventy two (82.7%) received chemotherapy following surgery. Among those 72 patients who received chemotherapy 49 (68 %) patients completed their treatment. Until the last follow up 71.4% patients remained alive and tumor free.*

Conclusion: *Germ cell tumors are the most variable tumor of all childhood malignancies that has difference in age, sex, location and histological subtypes. Gonadal tumors have better prognosis than extragonadal tumors in both the sex.*

Key words: *Pediatric germ cell tumour.*

Introduction

Pediatric germ cell tumors (GCTs) are rare and heterogeneous tumors hypothesized to occur as a result of events in utero,[1,2] although the etiology is largely unknown. GCTs are grouped together due to their presumed common cell of origin, the primordial germ cell (PGC). During normal fetal development, PGCs originate in the embryonic yolk sac and migrate to the gonads.[3] GCTs typically occur in the testes or ovaries; however, extragonadal GCTs can occur and have been hypothesized to result from abnormal germ cell migration during development.[4]

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These tumors are felt to arise from a common progenitor germ cell but have a wide range of histologies. GCTs are grouped into two broad classes: seminomas comprised of the seminomas of testes and dysgerminomas of the ovaries; and nonseminomas, comprised of yolk sac tumors, teratomas, embryonal carcinomas and choriocarcinomas.[5] In this study, we analyzed the age and sex distribution, location, types of tumors, management, outcome and complications of GCTs in children.

Materials and Methods

This retrospective study was done in pediatric oncology department of National Institute of Cancer Research and Hospital, Bangladesh from 2008 to 2014 by using the data from the patients registry. Medical records of patients aged 18 years or younger, who were diagnosed with GCT and were treated between 2008 and 2014, were retrospectively reviewed. The files were reviewed in terms of age, sex, types, location, management as well as prognosis and outcomes. Histopathological subtypes and their locations were investigated. The different types of tumor according to age and gender were analyzed. The treatments of the patients and the final status were recorded in detail. All statistical analysis of the results were obtained by using window based software devised with Statistical Packages for Social Sciences (SPSS) version 22. This study had power 95%. In all statistical tests 5% level of significance that is a p value < 0.05 was considered as significant.

Results

In present study, a total of 87 patients were included. Out of them female were 50 (57.5%) and male 37 (42.5%). The age of the most of the patients were 5 years or below (47%), followed by those above 10 years (37%) and others between 5 to 10 years (16%) (Fig-1). Male were more commonly affected up to 5 years of age (p = 0.019) and after 10 years female were more affected by malignant germ cell tumors in this study (p = <0.001). The gonadal germ cell tumors (80%) were more than extragonadal tumors (20%) in both male and female patients. The most common germ cell tumor was dysgerminoma (32%), then yolk sac tumor (29.8%), followed by teratoma (19.5%). The yolk sac tumor was more common in male (51.4%) than female (14%). In male patients most common germ cell tumor was yolk sac tumor (51.4%, p =< 0.001) followed by immature teratoma (18.9%) and in female dysgerminoma (56%) followed by teratoma (20%).

Testis (83.8%) was the commonest site followed by abdomen, sacrococcygeal region and mediastinum in male patients. In case of female patients ovary (78%) was most commonly affected followed by sacrococcygeal region, abdomen and mediastinum.

Out of 87, seventy two (82.7%) received chemotherapy following surgery. Out of 72 patients who received chemotherapy 49 (68%) patients completed their treatment.

Out of 49 patients who completed their chemotherapy, 5 patients developed local recurrence (12%), 4 patients abdominal metastasis (9.5%), 3 patients had lung metastasis (7.1%) and 30 (71.4%) patients did not have any complaint about their disease (11 up to 2 years, 15 up to 3 years and 4 above 3 years). Remaining 7 patients lost to follow up.

In case of male, prognosis was good up to 5 years of age (35.7%) and in female more than 10 years of age had good prognosis (42.8%).

According to histological types, out of 42 patients who completed treatment, yolk sac tumor (33.3%) was most curable in case of male and in female dysgerminoma (33.3%) was found the most curable one.

Table I

Clinical characteristics of the patients (N=87)

Variables	Male-37 (42.5%)	Female-50 (57.5%)	P value
Age of Patient			
0 to 5 years	30(81.2)	11(22)	0.019
> 5 to 10 years	4(10.8)	10(20)	0.248
> 10 years	3(8)	29(58)	< 0.001
Histological Types			
Yolk Sac Tumor	19(51.4)	7(14)	< 0.001
Dysgerminoma	-	28(56)	-
Seminoma	3(8)	-	-
Immature Teratoma	7(18.9)	10(20)	0.898
Embryonal Carcinoma	6(16.1)	0(0)	0.003
Malignant Mixed	2(5.6)	4(8)	0.664
Germ Cell Tumor			
Teratocarcinoma	0	1(2)	0.386
Site /Location of Tumor			
Ovary	-	39(78)	-
Testis	31(83.8)	-	-
Abdomen	3(8)	4(8)	1.00
Sacrococcygeal	2(5.5)	5(10)	0.446
Mediastinal	1(2.7)	2(4)	0.742

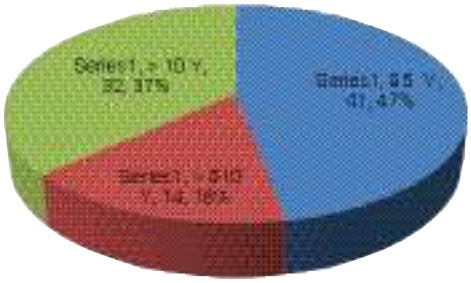

Fig 1 *Age distribution of patients*

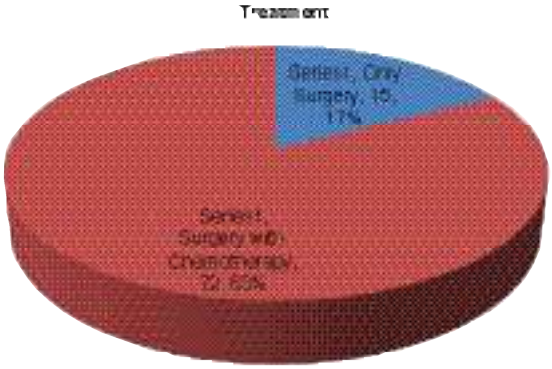

Fig 2 *Treatment modality of the patients*

Table II
Outcome of the patients (n=42)

Outcome	Number (%)	95% CI
Good	30(71.4)	65.41 - 75.54
Local Recurrence	5(12)	2.50 - 21.70
Metastasis	7(16.6)	5.31 - 27.40

Discussion

Malignant germ cell tumors are infrequent in childhood, occurring at a rate of 2.4 cases per million children and representing approximately 2% to 3% of cancers diagnosed below 15 years[6].

There are differences in distribution of tumor by location in pediatric age groups. Extragonadal tumors comprise of larger percentage in children before 4 years than in children diagnosed after the age of 10 years. It was consistent with the statement that 40% to 55% of pediatric GCTs are found in extragonadal locations[7-11].

The incidence of GCTs was similar in boys and girls in the age group of birth to 9 years, whereas the incidence was much higher in boys in the age more than 10 years reported in one study[12].

But in our study, up to 5 years of age GCTs were more common in boys and after 10 years it was found commoner in girls. Overall, female were more affected than male by GCTs, In general, the gonadal tumors occurred more than extragonadal ones regardless of the gender in this study.

The cure rate and survival are very high for pediatric GCTs, mainly due to the effectiveness of platinum based chemotherapy[13-15]. Although overall survival rate was higher, difference was observed in survival by location, histological type and staging of tumors, with gonadal tumors having more favorable prognosis than extragonadal locations. This observation is supported by numerous publications demonstrating lower survival rates in pediatric patients diagnosed in extragonadal locations.

Considering the histological types, yolk sac tumors and dysgerminoma had better prognosis than others found in this study.

The higher survival rate reported in gonadal as compared to extragonadal tumors could be explainable for having more complete tumor excision of tumors located in the gonads[14].

Conclusion

Germ cell tumors are the most varied tumors of all childhood malignancies having differences in age, sex, location and histological subtypes. Survival rate also differed depending on site and tumor types. In male prognosis was better in younger age group and that in female was better in adolescence.

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