ORIGINAL ARTICLE

Pediatric Solid Abdominal Tumors: Clinical Characteristics Study

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ABSTRACT

Introduction: Pediatric solid abdominal tumors (PSAT) ranked third after leukemia and central nervous system tumors. Nowadays, there is no sufficient data about characteristics and a tumor registry of PSAT in Indonesia, especially in our institution. This study describes the clinical characteristics of PSAT in Dr. Hasan Sadikin General Hospital, Bandung. **Methods:** A retrospective descriptive study was done on children diagnosed with PSAT and admitted to our division from January 2016 to May 2021. Data were collected from medical records, including demography, diagnosis, operability, staging, and patient outcomes. **Results:** A total of 122 patients were diagnosed with PSAT. The male: female ratio was 1:1.1, and most were in the 2–5 years of age group. Wilms' tumor was the commonest type (43%), followed by benign ovarian teratoma (16%), retroperitoneal teratoma (8%), rhabdomyosarcoma (8%), neuroblastoma (7%), hepatoblastoma (7%), malignant ovarian tumor (4%), sacrococcygeal teratoma (SCT) (3%), malignant lymphoma (2%), and gastrointestinal stromal tumor (GIST) (2%). Most cases were in advanced stages (62.3%), and 63.1% were unresectable, requiring preoperative chemotherapy. Of all patients who had completed their therapy, 41% were cured, 3% had a relapse, and the mortality was 28%. **Conclusion:** Most cases had late presentations, with advanced stage and unresectable tumors associated with unfavorable patient outcomes.

Keywords: Wilms' tumor, Neuroblastoma, Teratoma, Rhabdomyosarcoma, Hepatoblastoma

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INTRODUCTION

Pediatric solid abdominal tumors (PSAT) are the most common tumor after leukemia and the central nervous system (1). Most malignant abdominal masses are found in children ages 1 to 5 years, with Wilms' tumor and neuroblastoma being the most common types (2). In Greece, 73.3% of cases occurred in children under five years of age, with the most common types being neuroblastoma (46.7%), Wilms' tumor (26.7%), hepatoblastoma (13.2%), teratoma (6.7%), and granulose cell tumor (6.7%) (3). In Indonesia, it is estimated that every year there are 4100 new cases of cancer in children, fifty percent of these cases arrived late at healthcare facilities (4). Survival rates in other countries were relatively high, 90% for neuroblastoma and Wilms' tumor, 100% for low-risk hepatoblastoma, 86%–94% for sacrococcygeal teratoma, 75%–95% for retroperitoneal teratoma, 70% for rhabdomyosarcoma, and 95% for ovarian teratoma (5). The recurrence rate was also low, 20% for neuroblastoma, 15% for Wilms' tumor, 10%–20% for sacrococcygeal teratoma, and 25% for rhabdomyosarcoma (6). Data on survival and recurrence rates for PSAT remain unavailable, so the cancer registry in Indonesia cannot show these data.

The problem in developing countries, such as Indonesia, was the inadequacy of facilities and health systems for managing patients with solid abdominal tumors. The availability of diagnostic facilities and medical specialists was unevenly distributed in several hospitals and islands. Most patients with PSAT presented late. Multiple factors, such as the referral system and National Health Insurance, take time. Also, the lack of medical specialists to make late diagnoses, available postoperative intensive rooms, chemotherapy drugs, and facilities added to the lateness of diagnostic management. Low education and economic status also had a massive impact in Indonesia for late acknowledgment in this case.

This study describes the clinical characteristics of PSAT in our institution.

MATERIALS AND METHODS

A retrospective descriptive study was done on children diagnosed with PSAT admitted to our division from

January 2016 to May 2021. Data were collected from medical records. We collected demography (age, gender), diagnosis, tumor operability and staging, and patient outcome. Demographic data were stratified for each diagnosis to analyze the differences of clinical characteristics from each tumor type. The diagnosis was based on clinical manifestations, laboratory evaluations, radiology examinations, and pathology reports. Operability was divided into unresectable and resectable tumors. The staging was stratified based on each protocol preoperatively; benign tumors were not staged. For Wilms tumor staged with COG and SIOP, neuroblastoma with INSS, Rhabdomyosarcoma with IRSG, Hepatoblastoma with Pretext, Malignant Lymphoma with Ann Arbor and Malignant ovarian tumor with TNM staging. Outcomes were assessed from all patients who completed the treatment and follow-up. Then, the data was collected, processed, and evaluated; data would be displayed in the graphs. This study was approved by Research Ethics Committee, Faculty of Medicine Padjadjaran No.LB.02.01/X.6.5/295/2021.

RESULTS

A total of 122 PSAT patients with PSAT were admitted to our hospital. The demographic data are shown in Table I. The highest peak incidence of PSAT was in the 2–5 years of age group (46.7%), followed by the 6–12 years of age group (26.2%). Most patients had PSAT in a male: female ratio of 1:1.1. The tumor types are described in Table I, Wilms' tumor (43%) was the commonest type, followed by ovarian teratoma, retroperitoneal teratoma, and rhabdomyosarcoma.

The demographic data shows that the incidence in females is greater than in males. The gender distribution of PSAT as Wilms' tumor, rhabdomyosarcoma, and hepatoblastoma showed the incidence was more common in males, as seen in Figure 1. The age distribution in figure 2 shows that most of them were diagnosed at 2–5 years of age, but benign ovarian teratoma peak incidence in 6–12 years of age. Malignant ovarian tumors mainly occur at 13–18 years of age. It shows that each diagnosis had different characteristics and risk factors.

Figure 3 shows tumor staging based on the diagnosis. All tumors were mainly diagnosed in stages III and IV (62.3%), only a few patients with PSAT presented with early stage, and none had stage I. Patients with PSAT were diagnosed with radiologic imaging. Based on this examination, 63.1% presented with unresectable tumors. Resectable tumors (36.9%) underwent surgical resection, while unresectable patients underwent preoperative chemotherapy. After completing chemotherapy, patients were re-evaluated. Of these, 15.6% of tumors converted to resectable tumors and underwent surgical resection, as shown in Table I. Resectable tumors are predominantly from benign tumors. PSAT outcomes are shown in Table I for patients who completed treatment. The cure rate was 41%, the relapse rate was 3%, and the mortality rate was 28%. We had a 17% loss to follow-up and 11% of patients with ongoing treatment. The overall 2 years of survival rate of PSAT in this study was 72%, but we still need longer follow up duration, as shown in Figures 4.

Table I: Demographic characteristics of the patients and outcome

	N	Percentage (%)
Gender		
Males	53	56.6
Females	69	43.4
Age of diagnostic		
< 2 years	19	15.6
2-5 years	57	46.7
6-12 years	32	26.2
13-18 years	14	11.5
Diagnosis		
Wilms' tumor	53	43
Rhabdomyosarcoma	10	8
Neuroblastoma	8	7
Hepatoblastoma	19	16
Malignant lymphoma	5	4
Malignant ovarian tumor	10	8
Ovarian benign teratoma	2	2
Retroperitoneal teratoma	4	3
Sacrococcygeal teratoma	3	2
Gastrointestinal stromal tumor	8	7
Operability		
Unresectable	77	63.1
Resectable	45	45
Outcome		
Cured	37	41.1
Follow up	10	11.1
Loss to follow up	15	16.7
Relaps	3	3.3
Death	25	27.8

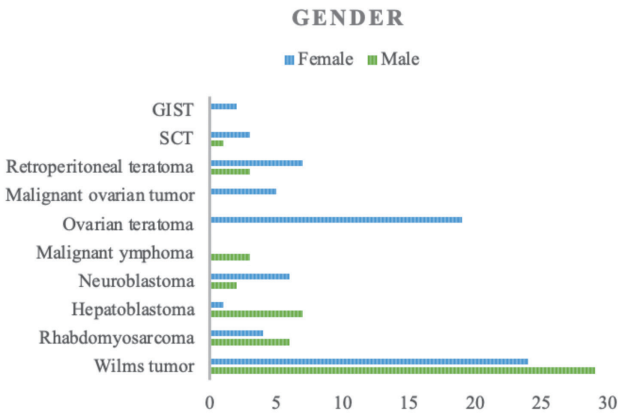

Figure 1: Gender distribution in diagnosis. Incidence in females is greater than in males. The gender distribution of PSAT as Wilms' tumor, rhabdomyosarcoma, and hepatoblastoma show the incidence is more common in males.

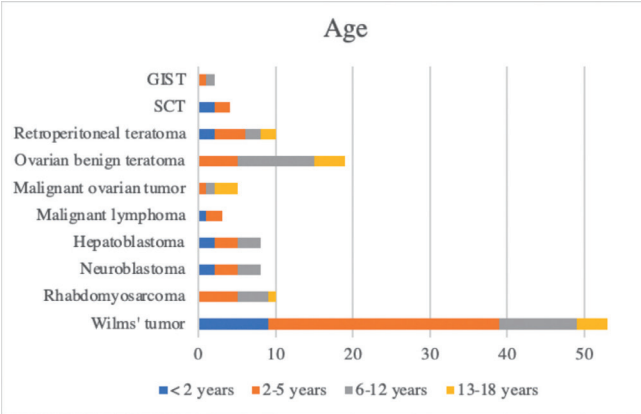

Figure 2: Age distribution in diagnosis. The age distribution shows that most of them were diagnosed in 2–5 years of age, but benign ovarian teratoma peak incidence in 6–12 years of age. Malignant ovarian tumors mainly occur at 13–18 years of age.

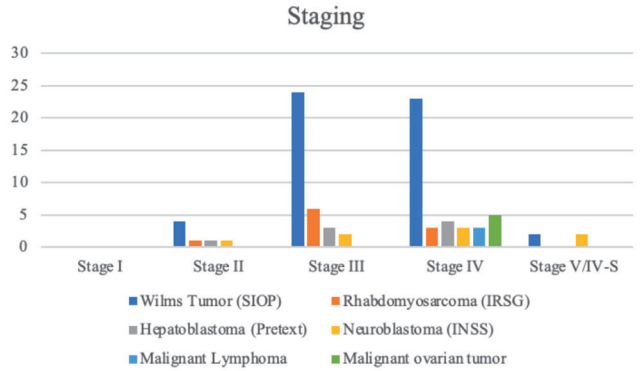

Figure 3: Staging of malignant PSAT. All tumors were mainly diagnosed in stages III and IV (62.3%).

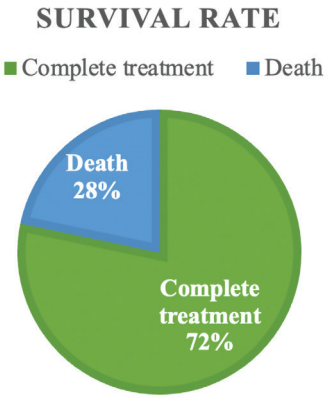

Figure 4: Survival rate. The overall 2 years of survival rate of PSAT who completed treatment in this study was 72%

DISCUSSION

PSAT are a group of nonhematologic, extracranial cancers that occur during childhood and approximately 40% of all pediatric cancers. Most malignant abdominal masses are found in children ages 1 to 5 years (2). A study in Greece showed that the PSAT incidence was 73.3% in children below five years old (3). These results were in line with this study, which also had a peak age from 2 to 5 years old. However, looking further at older age groups in our study, patients were diagnosed above five years old. These results differed from studies in other countries. For example, a study in Texas showed that the peak incidence of Wilms' tumor was below four years old, whereas that of neuroblastoma and hepatoblastoma was younger (2).

The results are not much different if we compare our incidence with studies in other developing countries. Studies in India and Iran showed that many patients were diagnosed with PSAT were over seven years old (7,8). This was because of many factors, including low education and socio-economic levels and inadequate health care in society. Not all people in developing countries view health as their priority. Also, people living in rural areas still believe in the power of spiritual and traditional treatments. These things were difficult to change and contributed to the lateness of diagnosis.

The difficulty of accessing health facilities that are capable of diagnosing and treating was another factor. This difficulty arises from Indonesia's geographical location as an archipelago state. Many islands still did not have available medical equipment and medical specialists, often requiring patients to visit hospitals on other islands to get cured. Also, our national health insurance was not be distributed evenly. Many people still did not get national insurance and could not afford medical fees. These individuals must concern us and also the governors. We need to assist them financially because tumors are one of the diseases with high medical expenses.

When we assessed by gender distribution, this study showed that the incidence of PSAT was higher in women, with a male: female ratio of 1:1.1. This was different from two studies in India where the male: female ratio for PSAT was 1.2:1 (8,9). We concluded that this result was because benign ovarian teratoma was the second most frequent cause of PSAT in our hospital. Therefore, the distribution we obtained was greater in females than in males. After gender grouping for each diagnosis, the result showed that the incidence of most tumor types is more common in men. Additional data from Yogyakarta, Wilms' tumor patients was more common in female. But for Rhabdomyosarcoma was more common in male (10,11).

The most common diagnosis was Wilms' tumor in this study, followed by ovarian teratoma 16%. Compared with other studies, this result is quite different. In a study in Greece, the most cases were neuroblastoma 46.7%, followed by Wilms' tumor 26.7%, while teratoma was only 6.7% (3). This difference can be caused by various risk factors, including genetics, lifestyle, and environmental factors. In a study in India, the commonest cases were also neuroblastoma 41.18%, followed by

Wilms' tumor 23.53%, hepatoblastoma 11.76%, and teratoma 11.76% (8). However, studies in Iran and India had similar results with this study. Wilms' tumor was the most common tumor, constituting 34%–50% of all cases, followed by neuroblastoma (7,8). A study in Africa also showed that Wilms' tumor was the commonest type of PSAT (10). First-line treatment in PSAT was surgical resection whenever possible. A study in Iran showed 47.6% of tumors were resected totally, sub-totally in 15.4%, and unresectable in 37% (9). These were not much different from our study, where the number of unresectable tumors was high. We must learn how systems work in developed countries for diagnosing and managing these cases and make some improvements in ours. Doctors, governors, and associations must have the same goal to work together and improve systems for detecting and managing the disease.

Patients who come to our hospital have disease in an advanced stage. This is similar to the stage when this condition presents at a major referral center in South Africa. Here, 65% of cases present with stage III or IV disease, contrasting with the stage distribution in North America and Europe (12). Because of this, the tumor at presentation was unresectable. So, surgical resection was not the first-line treatment. Thus, chemotherapy or radiotherapy was used instead. This condition enabled patients to prolong treatment, increase the length of hospital stay, and incur high treatment expenses. The length of hospital stay will affect the risk of secondary infections and complications, which will worsen the patient's condition and reduce the recovery rate. The patient's low economic status also dramatically affects the ability to continue the treatment because of the high cost of completing the therapy.

The late presentation has a clear association with poor outcomes. It explains why the fatality rate in this study is still very high following the low success rate. When compared with the survival rate in developed countries, it reached 80% (13). The outcome of PSAT in this study for patients who completed treatment, the cure rate remained very low in comparison to the mortality rate. However, the recurrence rate was low. There is a chance that if the tumor were diagnosed sooner, the outcome would also improve. If we can diagnose the disease sooner, it might affect many other aspects, such as lowering medical expenses, increasing the survival rate, and enabling prevention rather than treatment of PSAT.

The following circumstances warrant our consideration. Our institution is a tertiary hospital and has the highest referral rate. Many patients come from other islands. Some of them are difficult to contact when they return home, causing the number of cases lost to follow-up cases to remain high. Most patients discontinue therapy when they have difficulty paying medical fees. Even when the treatment is fully covered by national insurance, they can afford to fulfill their daily needs for the time being. Education was an essential factor contributing to successful therapy. Educating people on how to detect early signs and symptoms can help. For patients on treatment, we must assess all aspects, such as education, economic, and psychological status. We must be aware of potential problems in our patients to prevent them from discontinuing therapy.

CONCLUSION

Wilms' tumor, benign ovarian teratoma, and retroperitoneal teratoma were the most common types of PSAT in our hospital. Most cases of PSAT had late presentations, with advanced stage and unresectable tumors that were associated with unfavorable patient outcomes. PSAT mainly occurs under five years of age and is more common in males. Regarding the results, it can be concluded that the epidemiological and clinical characteristics of PSAT in our hospital are similar to other developing countries.

Limitations of this study are that many patients were lost to follow-up because they came from far away and lost contact. For improvement, patients need to be educated about the disease and the chance of a high survival rate. Follow-ups must be done regularly and continuously to develop a complete record for the cancer registry based on a sound follow-up system. We must collaborate with cancer associations in Indonesia for successful therapy. Further and more extensive studies are needed to compare the effect of tumor resection, stage, and response to treatment as prognostic factors for PSAT.

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