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CLINICOPATHOLOGICAL PROFILE OF SOFT TISSUE SARCOMA IN A PEDIATRIC AGE GROUP AMONG THOSE WHO ATTENDED HEALTH INSTITUTIONS IN NORTH INDIA

Surgery

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ABSTRACT

Background: Soft tissue sarcomas are a rare group of heterogeneous tumours that consist of several neoplasia which differentiate into different cell lines. They could arise from the mesodermal or ectodermal embryonic germ layers. Rhabdomyosarcoma is the most common soft tissue tumour in the paediatric age group in most studies, while in adults, malignant fibrous histocytoma, liposarcoma and fibrosarcoma are seen to be more predominant. The study aims to highlight the histological patterns, clinical presentation, associated risk factors and the patients' performance status at presentation.

Methods: This is a retrospective Study of all cancer patients diagnosed with soft tissue sarcoma at a tertiary health centre Data collected included socio-demographic, histological type, clinical features, and performance status.

Results: A total of 64 patients were reviewed during the study period and 4 patients were diagnosed suffering from soft tissue sarcoma.(6.25%).50% cases seen in 2-4 year age group with equal gender preponderance(male:female;50:50).Rural and urban also show 50:50 incidence. Abdominal mass weight loss were main symptoms.Pallor was the main sign (75%).Investigations like USG reveal distortion of architecture & Cct Showed either the disease is localized /extent demarcated.50% CASES WERE IN Stage I Disease as per International soft tissue sarcoma study group.75% cases were operable showing the tumour was located retroperitoneally or arised from bladder wall.Complete excision was done in those cases with localized retroperitoneal disease followed by histopathology.

Conclusions: Rhabdomyosarcoma was the most predominant histological type of soft tissue sarcoma seen over the review

KEYWORDS

Sarcoma, Soft tissue, Tumour

INTRODUCTION

Soft tissue sarcomas are a heterogeneous group of mesenchymal tumours which consists of 1% of all adult malignancies and about 12% of pediatric cancers.(1,2) It can arise from both the mesodermal and ectodermal layers and has the capacity to mature into several adult cell lines/ tissues which include striated and smooth muscles, adipose tissues, fibrous tissues, among others. There are also those whose line of differentiation are not clearly defined.(3,4).The classification of soft tissue tumours includes adipocytic tumours, fibroblastic/ myofibroblastic tumours, so called fibrohistiocytic tumours, smooth muscle tumours, pericytic tumours, gastrointestinal stromal tumours, nerve sheath tumours, tumours of uncertain differentiation and undifferentiated sarcomas.(3) Malignant fibrous histiocytoma is the most common soft tissue sarcoma globally, though there are other studies where liposarcoma was seen as the commonest histological type.(5,6)A pathological grouping of soft tissue sarcomas in a study showed malignant fibrous histiocytomas to be 34.2% of the histological type seen, followed by synovial sarcoma (17.1%), liposarcoma (16.3%) and rhabdomyosarcoma (12.6%). Fibrosarcoma was however the commonest soft tissue sarcoma in another study followed by malignant fibrous histiocytoma.(7) Rhabdomyosarocma is the commonest paediatric soft tissue sarcoma seen but was also the commonest type in a study in Niger delta region of Nigeria in both the adult and paediatric age group.(8,9) A review of soft tissue sarcoma over a 20 year period showed the peak incidence of age occurred in the third and sixth decades of life, while another study showed the mean age of presentation of soft tissue sarcoma to be 37.4±12.6 years and the age range from 18 to 85 years among adult population but with inclusion of paediatric age group in another study, the youngest patient seen was 3 years and the oldest was 73 in another study.(8)There is a slight gender predilection seen in soft tissue sarcoma in males compared to females. This is also seen in a number of studies in Nigeria that showed soft tissue sarcoma to be commoner among the males than females in all age groups.(10-12)Risk factors of soft tissue sarcoma include environmental exposure to carcinogens which include vinyl chloride, herbicides and pesticides. It also includes genetic susceptibility, chronic lymphoedema, previous radiation exposure and interaction between these factors. Immunosuppression is also a risk factors for soft tissue sarcomas.(13,14) HIV associated malignancy have increased risk for patients having soft tissue sarcoma. Kaposi sarcoma is a soft tissue sarcoma seen in patients with AIDS.(15,16) However, most cases of soft tissue sarcoma do not have an identifiable cause.(13)The most affected site of the body of presentation of soft tissue sarcoma are the extremities with the lower limbs constituting most of the cases seen, followed by the lower limb while the least affected site was the retroperitoneum.(17)There are varied predilection site based on the different classes of soft tissue sarcoma.(8) A similar finding was seen in another study where the extremities was the commonest site which was closely followed by the trunk but the retroperitoneum constituted 15%, while the head and neck region was 9%.(18,19)The performance status of cancer patients at presentation affects the prognosis of the disease as patients with poor performance status have worse survival, due to poor tolerance to treatment.(20,21). Due to the wide heterogeneity of soft tissue sarcoma, characterization of this group of malignancy to know the pattern, identify the risk factors and highlight the presenting symptoms of patients which could be easily identified for prompt referral, diagnosis and treatment.

METHODS

The study was conducted in the teaching hospital in North India .This is a retrospective study of all patients with soft tissue sarcoma seen in the teaching hospital .Data was collected from cases notes, histology reports and treatment cards. Data collected included; socio-demography, presenting symptoms, site of soft tissue sarcomas, and performance status of the patients at presentation

Inclusion criteria

All patients seen with soft tissue sarcoma.

Exclusion criteria

- All patients with bone sarcoma.
- All patients without a histological confirmation of soft tissue sarcoma.

Data analysis

Data was analyzed using SPSS version 21.0. All continuous variables were assessed for normality and presented as means ±standard deviations (SD). Qualitative variables will be summarized as tables and charts.

RESULTS

TABLE 1: Shows The Distribution Of Soft Tissue Tumours Among The Study Group

PRIMARY ABDOMINAL TUMOURS DETECTED	64
SOFT TISSUE SARCOMA	04 (6.25%)

TABLE 2:shows Age-distribution Of Soft Tissue Tumours In Study Group

AGE IN YEARS	SOFT TISSUE SARCOMA CASES	PERCENTAGE(%)
2-4	2	50
4-6	1	25
10-12	1	25

TABLE 3: Showsthe Sex Distribution Of Soft Tissue Sarcoma Cases Among Study Group

GENDER	NUMBER OF CASES	PERCENTAGE(%)
MALES	2	50
FEMALES	2	50

TABLE4 : Shows Rural V/s Urban Distribution Among Study Group

LOCALITY	NUMBER OF CASES	PERCENTAGE(%)
RURAL	2	50
URBAN	2	50

TABLE 5: Shows Incidence Of Various Symptoms In Soft Tissue Sarcoma Cases

SYMPTOMOLOGY	NUMBER OF CASES	%
ABDOMINAL MASS	2	100
PAIN ABDOMEN	1	50
VOMITTING	2	100
ANEMIA	2	100
WEIGHT LOSS	2	100
PALLOR	2	100
JAUNDICE	1	50

TBLE 6- shows Incidence Of Physical Signs Among Cases

PHYSICL SIGN	SOFT TISSUE SARCOMA CASES
PALLOR	3(75%)
INGUINAL LYMPHADENOPATHY	1(25%)

TABLE 7: Shows Findings On Local Examination Among Cases

PHYSICAL SIGN	SOFT TISSUE SARCOMA CASES
DISTENSION	2(50%)
VISIBLE LUMP	4(100%)

TABLE 8:showing Hb Level Among Cases

HB LEVEL	SOFT TISSUE SARCOMA CASES
< 6 gm%	1 (25%)
6-10 gm%	2 (50%)
>10 gm%	1 (25%)

TABLE 9: Shows Usg Evaluation Of Study Cases

TUMOUR CHARACTERISTIC	SOFT TISSUE SARCOMA CASES
MIXED	2(50%)
DISTORTION OF ARCHITECTURE	2 (50%)
LYMPH NODE STATUS	1 (25%)
ASCITES	1(25%)

TABLE 10: Shows The Results Of Ct Scan Among Study Cases

CT CHARACTERISTIC	SOFT TISSUE SARCOMA CASES (n=2)
DISEASE LOCALIZED	1 (50%)
EXTENT DEMARCATED	1(50%)

TABLE 11: Shows Incidence According To Staging Of International Soft Tissue Sarcoma Study Group

STAGE	NUMBER OF CASES	%
STAGE I	2	50
STAGE II	1	25
STAGE III	-	-
STAGE 1V	1	25

TABLE 12: Shows Distribution Of Patients Who Underwent Surgery In Our Study

SOFT TISSUE SARCOMA (RHABDOMYOSARCOMA)	NUMBER OF CASES INVESTIGATED	NUMBER OF PATIENTS OPERATED	%	NOT-OPERATED	%
	04	03	75	01	25

TABLE 13: shows Operative Findings And Histopathological Staging Of Soft Tissue Sarcoma(n=3)

TUMOUR SITE	EXTENT	NODES	SPLEEN	ASCITES	OPERATION	HISTOPATHOLOGY
RETROPERITONEUM	LOCALISED TO RETROPERITONEUM				TOTAL EXCISION OF TUMOUR WITH WIDE MARGIN	EMBRYONAL RHABDOMYOSARCOMA
RETROPERITONEUM	LOCALISED TO RETROPERITONEUM				EXCISION OF TUMOUR WITH WIDE MARGIN	D0
URINARY BLADDER	ARISING FROM ANTERO LATERAL WALL OF BLADDER				ONLY Bx TAKEN & SENT FOR CHRMOTHERAPY	DO

DISCUSSION

Soft tissue sarcoma epidemiology differs slightly among several regions around the world. There are studies that showed a 6.25 % and above prevalence of soft tissue sarcoma in North India which is higher than global incidences.(1,13,22) There is a predominance of the male gender in patients with soft tissue sarcoma in most studies but differs from our study which show equal preponderance between males & females.

Majority of the patients had no identifiable risk factors. This correlates with in a study in France and other studies where most patients prospectively studied did not have any identifiable risk factors.(6,13)

Rhabdomyosarcoma was the commonest soft tissue sarcoma seen in the total patients reviewed with which was similar with a study in the south western part of Nigeria were is made up 13.1% of all soft tissue sarcoma but this differ from a study in Kano where Kaposi sarcoma was the predominant soft tissue sarcoma seen but only 4.9% of the patients seen in this study had Kaposi sarcoma.(12)

Rhabdomyosarcoma was also the commonest histological type seen in both the paediatric age group.

This is in keeping in other studies conducted in Nigeria and other countries.(23-25) Malignant peripheral nerve sheath tumour comprise of 2% of all soft tissue sarcoma Also malignant fibrous histocytoma was 3.3%, however other reports showed malignant fibrous histocytomas the commonest soft tissue sarcoma.(26-29).

Most patients presented with a swelling followed by pain pain .This is in keeping with findings in most studies.(19)

CONCLUSION

Soft tissue sarcomas are rare tumours that consist of various histological types. Its epidemiological characteristics differs globally in several literatures. Rhabdomyosarcoma is seen to be the predominant histological type in both paediatric and adult age groups, but this is not

the case in other reports in most other studies. The extremities remain the commonest site of involvement as seen in both this study and most reports. Most patients still present late in our environment and thus have a poor performance status at presentation, which has been noted to affect their survival.

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