

Clinicopathological study in a case series of soft tissue sarcomas in a tertiary care hospital of north-east India and their management

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How to citation this article: Dr. Nilanjan Paul, Dr. Sajeed Ahmed, Dr. Dilip Killing, “Clinicopathological study in a case series of soft tissue sarcomas in a tertiary care hospital of north-east India and their management”, IJDSIR - April - 2023, Volume – 6, Issue - 2, P. No. 370 – 374.

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Type of Publication: Original Research Article

Conflicts of Interest: Nil

Abstract

Introduction: Soft tissue sarcomas are malignant tumors of connective tissues. These are very rare and can occur in any anatomical site but commonly involve the extremities. The incidence of different sarcomas differs in children and adults. A number of benign conditions can resemble soft tissue sarcoma resulting in unplanned and incorrect excision. Histopathological examination with tissue biopsy is considered the best modality for diagnosing such tumors along with radiological imaging. Planned surgical removal is the standard of care and is usually followed with adjuvant treatment. These tumors are very notorious for recurrence and are prone to metastasize. Due to the variety of these tumor entities along with their rare incidence, this study was conducted for evaluating the clinical and pathological profile of soft

tissue sarcomas in our region thereby enhancing our knowledge for proper management.

Methods: The study was conducted in the department of Surgery in Assam medical college and Hospital from the period of January 2021 to January 2023. It was a prospective case series of 15 patients of soft tissue sarcoma. The history, clinical and radiological findings were recorded and tabulated along with the treatment received according to the grade and histological type.

Results: Out of 15 patients, 10 were males. Most of the patients were in the age group of 50-60 years with primary tumors. There were 9 cases of tumors in the abdomen, 5 in the extremities and 1 in chest wall. 5 cases were unresectable with metastasis present in 3 cases. All unresectable tumors underwent palliative treatment while the resectable tumors underwent Wide local excision. 2 patients died.

Conclusion: Soft tissue sarcomas are rare tumors with a variety of presentations. They should be identified and referred early to tertiary healthcare facilities for definitive diagnosis and treatment to reduce morbidity and mortality.

Keywords: Biopsy, excision, management, soft tissue sarcoma.

Introduction

Soft tissue sarcomas are heterogeneous group of malignant tumors, arising from connective tissues which are embryologically derived from the mesenchyme.^{1,2} These are rare type of cancers that can occur in any part of the body - internally inside the body cavities or externally involving the trunk or limbs.³ The tissue of origin of such tumors include skeletal muscle, smooth muscle, adipose cells, blood and lymphatic vessels or other connective tissues.²

They usually constitute about 1% of all malignant neoplasms.⁴ The incidence of different sarcomas differs in children and adults and by geographical areas. The median

Age of diagnosis is 59 years with a bimodal distribution peaking in the 5th and 8th decades.⁴

Soft tissue sarcomas have more than 100 different tumour entities that make these complex and diagnostically challenging.⁴ These most commonly occur in the extremities, with 60% of all sarcomas.⁴ Also there are certain soft tissue sarcomas that commonly are found in particular sites.

The pathology of soft tissue sarcoma is diverse, so is their biological activity from being locally aggressive to potentially metastatic.³ Though they are grouped under an umbrella term of soft tissue sarcoma, the different entities under it have different presentations with varied outcomes and thereby their management is different.^{5,6}

A number of common nonneoplastic conditions like lipoma, hematoma or cyst can mimic soft tissue sarcoma which can increase the chance of mismanagement and recurrence due to unplanned excision.⁶ Histopathological examination in the form of tissue biopsy is considered to be the gold standard for diagnosing soft tissue sarcoma.⁷ Immunohistochemistry also plays a major role in assisting diagnosis of such cancers along with the regional radiological imaging of the affected part.⁷ Planned surgical removal is considered the standard of care for the treatment, but depends on the resectability.⁸ It is followed by adjuvant chemotherapy (Doxorubicin, Ifosfamide, etc) or radiotherapy depending on histopathology for preventing recurrence or metastasis in high-grade tumours.⁸

Though there are different studies and reports of cases of soft tissue sarcoma, there has not been any considerable clinical and pathological data from northeastern part of India on account of their rarity. Thus a clinical prospective case series study has been tried in Assam medical college and hospital for documenting the rare cases and enhancing knowledge on patient management and outcome along with future research activities.

Methods

The study has been conducted in the department of Surgery in Assam medical college and hospital, Dibrugarh from the period of January, 2021 to January, 2023. Considering the rarity of cases with varied presentations, a prospective clinical case series was done on patients of age more than 12 years coming with soft tissue lumps which are increasing in size, firm to solid in consistency and/or deep seated. The patients who had FNAC and/or biopsy reports conclusive of sarcoma or cellular atypia were included in the study. Patients with benign reports were excluded. The biopsy approach is

planned so that the entire needle trajectory can be easily incorporated into the forthcoming surgical resection volume to avoid any recurrence.

The following were recorded for each patient in the study: age at presentation, gender, disease status at presentation, prior treatment, lump characteristics (size, location, depth, consistency, fixity to skin and deep structures, involvement of neurovascular and vital structures), radiological features (USG of concerned area complemented with CECT or MRI for local and regional spread depending on abdominal or peripheral tumor respectively), details of treatment (preoperative and postoperative chemo or radiotherapy, with or without surgery) and follow up within the study period. Sarcomas were staged according to AJCC staging. All the resectable tumors were planned for wide local excision including the biopsy tract for assuring negative margins and preventing recurrence. The excised area in case of body wall or extremity tumors was then filled up with tissue flaps from surrounding areas. The features were tabulated and studied. In the end a conclusion derived from the study findings regarding proper evaluatory and management processes.

Results

In the study period, 131 patients reported to the hospital with a palpable painless mass in the extremities or abdomen. Following the history and clinical examination with radiological study and biopsy, 116 cases were excluded as they were benign tumors or cysts. Out of 15 (11.45%) patients with soft tissue sarcoma, 10 were males. There were patients of varied age groups but most of them were elderly. 7 patients were in the age group of 50-60 years followed by 3 each in 30 – 40 years and 40- 50 years age group. Depending on the location, there were 9 (60%) cases of malignant tumors in the

abdomen, 5 (33.33%) were in the extremities and 1 (6.67%) in chest. Among all the soft tissue sarcomas, majority (6;40%) were gastrointestinal stromal tumors, followed by retroperitoneal sarcomas in 2 cases (13.33%). There were 1 (6.67%) case each of desmoid fibromatosis, liposarcoma, synovial sarcoma, rhabdomyosarcoma, alveolar soft part sarcoma, myxofibrosarcoma and liposarcoma.

All but 1 (6.67%) case of soft tissue sarcoma were having primary presentation of soft tissue sarcoma. One patient presented with recurrent mass in abdomen due to incomplete resection in previous surgery somewhere else. Upon histological grading and radiological imaging, 5 (33.33%) of the total cases were unresectable due to higher grades and tissue invasion, including the case with recurrent presentation. Majority (80%) of these aggressive tumours were present in the age group 50-60 years with late presentation. Out of all high-grade tumors, 4 (80%) were of intraabdominal origin (3 GIST and 1 retroperitoneal sarcoma) and 1 (20%) tumor was of lower limb (liposarcoma). Metastasis was present in 3 cases (20%). All high-grade GIST who was unresectable underwent palliative treatment with Imatinib. Metastatic liposarcoma of lower limb got palliative chemoradiation. Most of the extremity sarcomas were resectable with wide

Local excision, 80% (4 of 5) including one in the chest wall. There was 2 (13.33%) death in our study due to metastatic spread.

Figures

a) Excised Gastrointestinal stromal tumor



b) Excised soft tissue sarcoma of thigh



c) Soft tissue sarcoma of chest wall



Table

| Patient details | Presentation | Tumour site | Diagnosis | Stage | Treatment | Progression | Outcome | Cases |
|-----------------|--------------|-----------------------------------|----------------------------|-------|-----------------------------------|---------------|---------|-------|
| 53y/F | Primary | Abdomen | GIST | IV | Palliative CT | Metastasis | Died | 1 |
| 30y/M | Primary | Left chest wall with rib invasion | Liposarcoma | IV | WLE + rib resection + adjuvant CT | No recurrence | Alive | 2 |
| 60y/M | Primary | Abdomen | GIST jejunum | III | Tumour resection + CT | No recurrence | Alive | 3 |
| 50y/M | Recurrence | Abdomen | GIST | IV | Palliative CT | Unresectable | Alive | 4 |
| 60y/M | Primary | Left leg | Liposarcoma | IIIB | Palliative RT + CT | Metastasis | Alive | 5 |
| 59y/M | Primary | Abdomen | GIST | IV | Palliative CT | Metastasis | Died | 6 |
| 40y/M | Primary | Abdomen | Retro-peritoneal sarcoma | IIIB | Palliative CT | Inoperable | Alive | 7 |
| 58y/M | Primary | Abdomino-pelvis | GIST | IIB | Resection +CT | No recurrence | Alive | 8 |
| 54y/M | Primary | Right leg | Rhabdomyosarcoma | III | WLE + CT | No recurrence | Alive | 9 |
| 35y/F | Primary | Abdomen | GIST | IIB | Resection + CT | No recurrence | Alive | 10 |
| 41y/M | Primary | Abdomen | Retro-peritoneal sarcoma | IIB | Resection | No recurrence | Alive | 11 |
| 56y/F | Primary | Left thigh posterior | Synovial sarcoma | III | WLE + RT | No recurrence | Alive | 12 |
| 31y/F | Primary | Abdominal | Mesenteric Desmoid | IIB | WLE | No recurrence | Alive | 13 |
| 29y/F | Primary | Right Buttock | Alveolar soft part sarcoma | IIA | WLE | No recurrence | Alive | 14 |
| 42y/M | Primary | Right thigh | Myxofibrosarcoma | IIB | WLE | No recurrence | Alive | 15 |

[M: Male. F: Female, GIST: Gastrointestinal stromal tumor, CT: Chemotherapy, WLE: Wide local excision, RT: Radiotherapy]

Discussion

The study focused on different presentations of soft tissue sarcoma in a period of 2 years at AMCH, Dibrugarh. The incidence of soft tissue sarcoma was 11.45% in our study which was comparable to the available literature.⁷ The male to female ratio was found to be 2:1 with majority (46.67%) of patients falling in the age group of 51-60 years.^{7,8} All (100%) patients presented with a painless palpable mass either in abdomen or limbs. These findings were in accordance with available literature.⁹

60% tumors in our study were present in abdomen followed by 33.33% in the lower extremities and 6.67% in the thorax. In the study by Shukla NK et.al, 48% of soft tissue sarcoma was from lower extremities followed by 21% in chest and trunk.⁸ Jenna P et.al in their study found 44% tumors in the lower extremities, 17% abdomen and 6% in chest.⁹

In our study, majority (40%) of the tumours were Gastrointestinal Stromal Tumours followed by 13.33% of retroperitoneal sarcoma, 6.67% of each of liposarcoma, rhabdomyosarcoma and synovial sarcoma. Synovial sarcoma was the most commonly encountered tumour (15%) followed by malignant fibrous histiocytoma (13.9%) in the study by Shukla NK et.al.⁸ In study by Kransdorf MJ et.al, proportion of sarcoma were malignant fibrous histiocytoma (24%), liposarcoma (14%) and leiomyosarcoma (8%).¹⁰ 66.67% tumours in our study were treated by surgery with Wide local excision. 33.33% tumours received adjuvant chemotherapy and 1 case (6.67%) of synovial sarcoma received adjuvant radiotherapy. 26.67% cases received palliative chemotherapy for being unresectable and 6.67% received palliative chemoradiation for being metastatic liposarcoma which was a rare finding. Limb salvage surgery was possible in all of the sarcomas in extremities. There was no recurrence of tumours within our study period. But 2 patients who succumbed to death due to metastatic spread were under palliative treatment already. In the study by Jenna P. Et.al, surgery was the main modality of treatment with 2 patients required limb amputation.⁹ Recurrence was seen in 12% of patients.⁹

Conclusion

Treatment of Soft tissue sarcoma depends primarily on presenting features, the duration of illness, tumor location, resectability with respect to the surrounding tissue invasion and also operability with reference to the performance status of the patient. Tissue diagnosis plays the forefront for eventual treatment and prognosis. As soft tissue sarcoma is a rare and challenging entity, they should be referred early for definitive treatment by surgery and adjuvant chemoradiotherapy to reduce recurrence and metastasis.

Ethical approval

The study was approved by the Institutional Ethics Committee.

9. References

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