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Case report

A CASE REPORT ON SOFT TISSUE SARCOMA

Peddapalli Appa Rao[1*], KV Subrahmanyam[2*], P. Sandhya Rani[2], Smitha Madhuri[2], Nafisur Rahman[1]

[1]Research Centre, IMMA Labs, Hyderabad, Telangana, India.
[2]Samskruti College of Pharmacy, Kondapur, Ghatkesar, R.R District, Hyderabad, Telangana, India.

ABSTRACT

Soft tissue sarcomas can develop from soft tissues like fat, muscle, nerves, fibrous tissues, blood vessels, or deep skin tissues. They can be found in any part of the body. Most of them develop in the arms or legs. They can also be found in the trunk, head and neck area, internal organs, and the area in back of the abdominal cavity (known as the retroperitoneum). Sarcomas are not common tumors, and most cancers are the type of tumors called carcinomas. Soft tissues include muscles, tendons, fat, fibrous tissue, synovial tissue, vessels, and nerves. Approximately 60% of soft tissue sarcomas arise in the extremities, with the lower extremities involved three times as often as the upper extremities. Thirty percent arise in the trunk, the retroperitoneum accounting for 40% of all trunk lesions. The remaining 10% arise in the head and neck. Malignant transformation of a benign soft tissue tumor is extremely rare, with the exception that malignant peripheral nerve sheath tumors (neurofibrosarcoma, malignant schwannoma) can arise from neurofibromas in patients with neurofibromatosis. The most common presentation is an asymptomatic mass. Mechanical symptoms referable to pressure, traction, or entrapment of nerves or muscles may be present. All new and persistent or growing masses should be biopsied, either by a cutting needle (core-needle biopsy) or by a small incision, placed so that it can be encompassed in the subsequent excision without compromising a definitive resection. Lymph node metastases occur in 5%, except in synovial and epithelioid sarcomas, clear-cell sarcoma (melanoma of the soft parts), angiosarcoma, and rhabdomyosarcoma, where nodal spread may be seen in 17%. The pulmonary parenchyma is the most common site of metastases. Exceptions are GISTs, which metastasize to the liver; myxoid liposarcomas, which seek fatty tissue: and clear-cell sarcomas, which may metastasize to bones. Central nervous system metastases are rare, except in alveolar soft part sarcoma. One such case of soft tissue sarcoma has been treated by Dr. Appa Rao's Immunonutritive therapy and the result was beneficial to the patient. Patient was followed for a year and was found to be healthy.

Key Words:- Soft tissue sarcoma, Immunonutritive therapy.

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Corresponding Author

Peddapalli Appa Rao
Research Centre, IMMA Labs, Hyderabad, Telangana, India.

Email:- vatavapparao@gmail.com

INTRODUCTION

Soft tissues include muscles, tendons, fat, fibrous tissue, synovial tissue, vessels, and nerves. Approximately 60% of soft tissue sarcomas arise in the extremities, with the lower extremities involved three times as often as the upper extremities. Thirty percent arise in the trunk, the retroperitoneum accounting for 40% of all trunk lesions. The remaining 10% arise in the head and neck. Malignant transformation of a benign soft tissue tumor is extremely rare, with the exception that malignant peripheral nerve sheath tumors (neurofibrosarcoma, malignant schwannoma) can arise from neurofibromas in patients with neurofibromatosis.

Several etiologic factors have been implicated in soft tissue sarcomas.

Environmental Factors : Trauma or previous injury is rarely involved, but sarcomas can arise in scar tissue resulting from a prior operation, burn, fracture, or foreign body implantation. Chemical carcinogens such as polycyclic

hydrocarbons, asbestos, and dioxin may be involved in the pathogenesis.

Iatrogenic Factors : Sarcomas in bone or soft tissues occur in patients who are treated with radiation therapy. The tumor nearly always arises in the irradiated field. The risk increases with time.

Viruses : Kaposi's sarcoma (KS) in patients with HIV type 1, classic KS, and KS in HIV-negative homosexual men is caused by human herpes virus (HHV) 8 (Chap. 175). No other sarcomas are associated with viruses.

Immunologic Factors : Congenital or acquired immune deficiency, including therapeutic immunosuppression, increases the risk of sarcoma.

Genetic Factors : Li-Fraumeni syndrome is a familial cancer syndrome in which affected individuals have germ-line abnormalities of the tumor-suppressor gene p53 and an increased incidence of soft tissue sarcomas and other malignancies, including breast cancer, osteosarcoma, brain tumor, leukemia, and adrenal carcinoma. Neurofibromatosis 1 (NF-1, peripheral form, von Recklinghausen's disease) is characterized by multiple neurofibromas. Neurofibromas occasionally undergo malignant degeneration to become malignant peripheral nerve sheath tumors. The gene for NF-1 is located in the pericentromeric region of chromosome 17 and encodes neurofibromin, a tumor-suppressor protein with GTPase-activating activity that inhibits Ras function. Germ-line mutation of the Rb-1 locus (chromosome 13q14) in patients with inherited retinoblastoma is associated with the development of osteosarcoma in those who survive the retinoblastoma and of soft tissue sarcomas unrelated to radiation therapy. Other soft tissue tumors, including desmoids tumors, lipomas, leiomyomas, neuroblastomas, and paragangliomas, occasionally show a familial predisposition. Ninety percent of synovial sarcomas contain a characteristic chromosomal translocation t(X;18) (p11;q11) involving a nuclear transcription factor on chromosome 18 called SYT and two breakpoints on X. Patients with translocations to the second X breakpoint (SSX2) may have longer survival than those with translocations involving SSX1. Insulin-like growth factor (IGF) type 2 is produced by some sarcomas and may act as an autocrine growth factor and as a motility factor that promotes metastatic spread. IGF-2 stimulates growth through IGF-1 receptors but its effects on motility are through different receptors. If secreted in large amounts, IGF-2 may produce hypoglycaemia (Kasper *et al*., 2005).

CLASSIFICATION

Approximately 20 different groups of sarcomas are recognized on the basis of the pattern of differentiation toward normal tissue. For example, rhabdomyosarcoma shows evidence of skeletal muscle fibers with cross-striations; leiomyosarcomas contain interlacing fascicles of spindle cells resembling smooth muscle; and liposarcomas contain adipocytes. When precise characterization of the group is not possible, the tumors are called unclassified sarcomas. All of the primary bone sarcomas can also arise from soft tissues (e.g., extraskeletal osteosarcoma). The entity malignant fibrous histiocytoma includes many tumors previously classified as fibrosarcomas or as pleomorphic variants of other sarcomas and is characterized by a mixture of spindle (fibrous) cells and round (histiocytic) cells arranged in a storiform pattern with frequent giant cells and areas of pleomorphism. For purposes of treatment, most soft tissue sarcomas can be considered together. However, some specific tumors have distinct features. For example, liposarcoma can have a spectrum of behaviors. Pleomorphic liposarcomas and dedifferentiated liposarcomas behave like other high-grade sarcomas; in contrast, well-differentiated liposarcomas (better termed atypical ipomatous tumors) lack metastatic potential, and myxoid liposarcomas metastasize infrequently but, when they do, have a predilection for unusual metastatic sites containing fat, such as the retroperitoneum, mediastinum, and subcutaneous tissue. Rhabdomyosarcomas, Ewing's sarcoma, and other small-cell sarcomas tend to be more aggressive and are more responsive to chemotherapy than other soft tissue sarcomas. Gastrointestinal stromal cell tumors (GISTs), previously classified as gastrointestinal leiomyosarcomas, are now recognized as a distinct entity within soft tissue sarcomas. Its cell of origin resembles the interstitial cell of Cajal, which controls peristalsis. The majority of malignant GISTs have activating mutations of the c-kit gene that result in ligand-independent phosphorylation and activation of the KIT receptor tyrosine kinase, leading to tumorigenesis.

DIAGNOSIS The most common presentation is an asymptomatic mass. Mechanical symptoms referable to pressure, traction, or entrapment of nerves or muscles may be present. All new and persistent or growing masses should be biopsied, either by a cutting needle (core-needle biopsy) or by a small incision, placed so that it can be encompassed in the subsequent excision without compromising a definitive resection. Lymph node metastases occur in 5%, except in synovial and epithelioid sarcomas, clear-cell sarcoma (melanoma of the soft parts), angiosarcoma, and rhabdomyosarcoma, where nodal spread may be seen in 17%. The pulmonary parenchyma is the most common site of metastases. Exceptions are GISTs, which metastasize to the liver; myxoid liposarcomas, which seek fatty tissue: and clear-cell sarcomas, which may metastasize to bones. Central nervous system metastases are rare, except in alveolar soft part sarcoma (Kasper *et al*., 2005).

Radiographic Evaluation Imaging of the primary tumor is best with plain radiographs and magnetic resonance imaging (MRI) for tumors of the extremities or head and neck and by computed tomography (CT) for tumors of the chest, abdomen, or retroperitoneal cavity. A radiograph and CT

scan of the chest are important for the detection of lung metastases. Other imaging studies may be indicated, depending on the symptoms, signs, or histology.

STAGING AND PROGNOSIS The histologic grade, relationship to fascial planes, and size of the primary tumor are the most important prognostic factors. The current American Joint Commission on Cancer (AJCC) staging system is shown in Table 1. Prognosis is related to the stage. Cure is common in the absence of metastatic disease, but a small number of patients with metastases can also be cured. Most patients with stage IV disease die within 12 months, but some patients may live with slowly progressive disease for many years (Kasper *et al*., 2005).

TREATMENT

AJCC stage I patients are adequately treated with surgery alone. Stage II patients are considered for adjuvant radiation therapy. Stage III patients may benefit from adjuvant chemotherapy. Stage IV patients are managed primarily with chemotherapy with or without other modalities.

Surgery : Soft tissue sarcomas tend to grow along fascial planes, with the surrounding soft tissues compressed to form a pseudocapsule that gives the sarcoma the appearance of a well-encapsulated lesion. This is invariably deceptive, because "shelling out" or marginal excision of such lesions results in a 50 to 90% probability of local recurrence. Wide excision with a negative margin, incorporating the biopsy site, is the standard surgical procedure for local disease. The adjuvant use of radiation therapy and/or chemotherapy improves the local control rate and permits the use of limb-sparing surgery with a local control rate (85 to 90%) comparable to that achieved by radical excisions and amputations. Limb-sparing approaches are indicated except when negative margins are not obtainable, when the risks of radiation are prohibitive, or when neurovascular structures are involved so that resection will result in serious functional consequences to the limb.

Radiation Therapy : External beam radiation therapy is an adjuvant to limb-sparing surgery for improved local control. Preoperative radiation therapy allows the use of smaller fields and smaller doses but results in a higher rate of wound complications. Postoperative radiation therapy must be given to larger fields, as the entire surgical bed must be encompassed, and in higher doses to compensate for hypoxia in the operated field. Brachytherapy or interstitial therapy, in which the radiation source is inserted into the tumor bed, is comparable in efficacy (except in low-grade lesions), less time consuming, and less expensive(Kasper *et al*., 2005).

Adjuvant Chemotherapy : Chemotherapy is the mainstay of treatment for Ewing's primitive neuroectodermal tumors (PNET) and rhabdomyosarcomas. Meta-analysis of 14 randomized trials revealed a highly significant improvement in local control and disease-free survival in favor of doxorubicin-based chemotherapy. Overall survival is improved only for extremity sarcomas, however. A chemotherapy regimen including an anthracycline and ifosfamide with growth factor support improved overall survival for high-risk (high-grade, _5 cm primary, or locally recurrent) extremity soft tissue sarcomas.

Advanced Disease : Metastatic soft tissue sarcomas are largely incurable, but up to 20% of patients who achieve a complete response become long-term survivors. The therapeutic intent, therefore, is to produce a complete remission with chemotherapy and/or surgery. Surgical resection of metastases, whenever possible, is an integral part of the management. Some patients benefit from repeated surgical excision of metastases. The two most active chemotherapeutic agents are doxorubicin and ifosfamide. These drugs show a steep dose-response relationship in sarcomas. Gemcitabine and dacarbazine also have some activity. Taxanes have selective activity in angiosarcomas, and vincristine, etoposide, and irinotecan are effective in rhadomyosarcomas and Ewing's sarcomas. Imatinib mesylate targets the KITtyrosine kinase activity and is standard therapy for advanced/ metastatic GISTs (Kasper *et al*., 2005).

Table 1. AJCC Staging system for sarcoma

Histologic Grade (G)	*Tumor Size (T)*	*Node Status (N)*	*Metastases (M)*
Well differentiated (G1) Moderately differentiated (G2) Poorly differentiated (G3) Undifferentiated (G4)	≤5 cm (T1) >5 cm (T2) Superficial fascial involvement (Ta) Deep fascial involvement (Tb)	Not involved (N0) Involved (N1)	Absent (M0) Present (M1)

Disease Stage	*5-Year Survival, %*
Stage I A: G1,2; T1a,b; N0; M0 B: G1,2; T2a; N0; M0	98.8
Stage II A: G1,2; T2b; N0; M0 B: G3,4; T1; N0; M0 C: G3,4; T2a; N0; M0	81.8
Stage III G3,4; T2b; N0; M0	51.7
Stage IV A: any G; any T; N1; M0 B: any G; any T; any N; M1	<20

DISCUSSION

Soft tissue sarcomas can develop from soft tissues like fat, muscle, nerves, fibrous tissues, blood vessels, or deep skin tissues. They can be found in any part of the body. Most of them develop in the arms or legs. They can also be found in the trunk, head and neck area, internal organs, and the area in back of the abdominal cavity (known as the retroperitoneum). Sarcomas are not common tumors, and most cancers are the type of tumors called carcinomas (Anonymous 1).

Dr. Appa Rao's Immunonutritive therapy focuses mainly on Inflammation. In this patient Immunonutritive therapy was initiated, when the patient was advised to undergo surgery i.e., amputation of leg. Immunonutritive therapy has shown beneficial results in this patient.

CONCLUSION

A 67 year old male patient is a known case of soft tissue sarcoma. He was operated for his tumor in the mid of 2005. Patient had no fresh complaints for six years after surgery, then in the mid of 2011, his sarcoma relapsed again. He visited an oncologist again and was advised for reoperation. His lump continued to increase in size. He was advised to go to TMC, Mumbai, for the surgery. He went to TMC in December 2011 and got MRI done. Then he was given date for surgery in January 2012. He again went to TMC in January 2012 and got MRI done. But somehow the patient decided not to go for surgery and came back to Hyderabad. Then he came to know about Dr. Appa Rao and visited him. He took his treatment and improved a lot. Patient was found comfortable and lump was reduced in size with Dr. Appa Rao's immunonutritive therapy.

Treatment schedule and follow up :

Injection Human normal immunoglobulin (12 mg) and histamine dihydrochloride (0.15 mcg), (Belonging to any manufacturer). Two vials once in three days (3 doses) followed by two vials once in a week until 8 weeks. Aceclofenac 100mg twice a day for one month. Prednisolone tapered and maintained 5 mg per day. Ranitidine 150 mg once a day in the morning. Tomato, Banana fruit, Prawns and milk were restricted in nutrition.

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