Original Article Journal of Bone and Soft Tissue Tumors 2018 July-Dec;4(2):7-10

Spectrum of Bone and Soft Tissue Tumors in A Tertiary Cancer Institute in Eastern India

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Abstract

Introduction: Bone and soft tissue tumours are uncommon tumours that can affect any age group. Soft tissue tumors are said to be heterogeneous group of mesenchymal malignancies. Primary bone sarcomas are rare tumors, comprising approximately 0.2% of all cancers. Their true incidence is difficult to estimate because of their rarity.[1]The outcome of the disease depends on the age and time at diagnosis.

Material and Methods: A study was carried out in department of pathology in tertiary cancer institute in East India from December 2015 to September 2017. A total of 60 cases were included in the study. Clinical and radiological details of patients were noted along with gross specimen findings and microscopic examination of H &E stained slides. IHC was also carried out for confirmatory diagnosis.

Aims and Objecctives: 1- To study the histological spectrum of bone and soft tissue tumors in a tertiary cancer institute in Eastern India
2- To study gender distribution and site distributionof bone and soft tissue tumors

Results: Out of the total 60 cases studied 24 cases were of bone and cartilaginous tumours and 36 cases included were of soft tissue tumors. Benign tumors comprised of 08 cases (13.3%) and malignant tumors accounted to 52 cases (86.7%). Age of the patients ranged from 10 to 80 years. Male female ratio was 1.4:1.

Conclusion: Multimodal therapies of treatment is practiced for bone and soft tissue tumors. Early detection and treatment is essential for diagnosis of these malignant tumors. Molecular studies are most important in diagnosing, classifying and also prognosticating bone and soft tissue tumors.

Keywords: bone tumors, soft tissue tumors, histology.

Introduction

Bone and soft tissue tumors can affect any age group.Soft tissue tumors are defined as mesenchymal proliferation that occurs in extraskeletal non epithelial tissue of the body, excluding the viscera, covering of the brain and lymphoreticular system[2]. A core biopsy, an excisional biopsy and an incisional biopsy are more useful for diagnosing as IHC and special stains can be easily carried out on the material. Extremities were the commonest site for malignant soft tissue tumors, followed by trunk, abdominal cavity and head and neck region.[3]

Aims and Objectives

1. To study the histological spectrum of bone and soft tissue tumors in a tertiary cancer institute in Eastern India
2. To study gender distribution and location wise distribution of bone and soft tissue tumors

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Materials and Methods

A study was carried out in the department of pathology in a tertiary cancer institute in eastern India for the period of December 2015 to September 2017. A total of 60 cases of bone and soft tissue tumors sent to histopathology department were included in the study. The sections were stained with H& E and also IHC was done wherever feasible.

Results

Out of the total 60 cases studied 24 cases were of bone and cartilaginous tumours and 36 cases included were of soft tissue tumors. Benign tumors comprised of 08 cases (13.3%) and malignant tumors accounted to 52 cases (86.7%).[Table 1] As per the study Ewings sarcoma was the most common bone sarcoma accounting to 12 cases (50%) followed by chondrosarcoma comprising of 5 cases (20.8%) osteosarcoma comprising of 3 cases (12.5%) and chondroma comprising of 3 cases(12.5%) and 1 case (4.2%) of chordoma. Amongst the soft tissue sarcomas rhabdomyosarcoma predominated with 8 cases (22.2%) followed by synovial sarcoma 6 cases (16.6%), and Malignant peripheral nerve sheath tumor 4 cases (11.1%). 3 cases (8.3%) each of undifferentiated pleomorphic sarcoma and angiosarcoma were studied. Other malignant tumors included 1 case (2.8%) each of leiomyosarcoma, liposarcoma, dermatofibrosarcoma protuberance and 2 cases (5.6%) of fibrosarcoma. Other rare malignant soft tissue tumors included in this study were extraskeletal ewings sarcoma comprising of 1 case (2.8%) and a case (2.8%) of clear cell sarcoma of soft part. Amongst the benign soft tissue tumors we received 4 cases (11.1%) of benign fibrous histiocytoma and 1 cases (2.8%) of fibroma.[Table 2] Age of the patients ranged from 10 to 80 years. Male female ratio was 1.4:1 (35 male, 25 female).[Table 3]Lower limb was the most common location in our study (26 cases), followed by upper limb (16 cases), chest wall (7 cases) and pelvis (5 cases). Other sites of involvement were retroperitoneum, scalp

Table 1: Incidence of benign and malignant bone and soft tissue tumors.		
	BENIGN	MALIGNANT
BONE TUMORS	-	15
CARTILAGENOUS TUMOR	3	6
SOFT TISSUE TUMORS	5	31
TOTAL	08(13.3%)	52(86.7%)

and jaw.[Table 4]

Discussion

Soft tissue tumors are a heterogenous group of tumors which are classified on histogenetic basis. Benign soft tissue tumors outnumber malignant tumors by a margin of about 100: 1 in hospital population[4]. Our study was conducted in a tertiary cancer institute which was mostly dealing with referred cases hence in our study malignant cases predominated. Bone and soft tissue tumors range from benign self-limited lesions to highly aggressive malignancies with significant metastatic risk and mortality. Classifications of soft tissue tumors continue to evolve as new molecular genetic abnormalities are identified. Among all sarcomas, irrespective of adult or pediatric patients, the most frequently observed specific subtypes are Ewing sarcoma/PNET, followed by synovial sarcoma; both the entities require IHC for confirmation in all cases. Over the years, with applications of immunohistochemical stains, usage of the histopathological term "spindle cell sarcoma" has reduced, and is being replaced by rather specific histopathological subtypes.Bone sarcomas constitute as the third most common cause of mortality in adolescentsOsteosarcoma (OGS), chondrosarcoma, and Ewing sarcoma/primitive neuroectodermal tumor (PNET) are the common bone sarcomas, as per Western data, with rare tumors such as fibrosarcomas, chordomas, and undifferentiated pleomorphic sarcoma (UPS) constituting as the remaining subtypes. The most common location of soft tissue tumor and bone tumors in our study was extremitis. The studies observed by lazim, Beg and Zhi et al[5,6,7] also stated the commonest site to be extremeties for maliganant soft tissue tumor mainly involving lower extremities followed by trunk and abdomen. In our study Male preponderance was observed in almost all tumours (Male: Female 1.4:1). A study by Batra et al[8] reported a ratio of 2.1:1(M:F).

Few of the interesting cases are discussed below.

Table 2: Histological distribution of bone and soft tissue tumors				
BONE	BENIGN			
			Ewings sarcoma	12
	MALIGNANT		Osteosarcoma	3
	BENIGN		Chondroma	3
CARTILAGENOUS	MALIGNANT		Chondrosarcoma	5
			Chordoma	1
SOFT TISSUE TUMORS				
SKELETAL MUSCLE TUMORS		BENIGN	-	-
		MALIGNANT	RMS	8
SMOOTH MUSCLE TUMOR		BENIGN	-	-
		MALIGNANT	Leiomyosarcoma	1
		BENIGN	-	-
VASCULAR		MALIGNANT	Angiosarcoma	3
NERVE SHEATH TUMOR		BENIGN	-	-
		MALIGNANT	MPNST	4
		BENIGN	-	-
ADIPOCYTIC		MALIGNANT	Liposarcoma	1
		BENIGN	Fibroma	1
FIBROBLASTIC			Fibrosarcoma	2
		MALIGNANT	DFSP	1
FIBROHISTIOCYTIC		BENIGN	BFH	4
			Synovial sarcoma	6
UNCERTAIN DIFFERENTIATION		MALIGNANT	Undifferentiated pleomorphic sarcoma	3
			Extra skeletal Ewings sarcoma	1
			Clear cell sarcoma	1

Ewing sarcoma family of tumors

Ewing sarcoma/PNET is an undifferentiated small blue round cell tumor of bone and soft tissues, predominantly seen in children and young adults. Ewing sarcoma family of tumor (EWSFT) includes Ewing sarcoma of the bone, extraosseous Ewing sarcoma, also referred to as extraskeletal Ewing sarcoma (tumor growing outside of the bone), PNET, peripheral neuroepithelioma, Askin's tumor (Ewing sarcoma of the chest wall), and atypical Ewing sarcoma. It is characterized by recurrent balanced translocations such as EWS-FLI and EWS-ERG that play key role in its pathogenesis. This tumor is diagnosed by histopathology with demonstration of positive immunohistochemical staining of antibody markers, such as CD99/MIC2 and FLI1 in most cases. Radiological imaging plays an important role in the diagnosis and assessment of response in EWSFT. This tumor has a wide clinicopathological spectrum and in cases with equivocal immunohistochemical results and in tumors occurring at unusual locations or in older age group patients, should be ideally confirmed with molecular testing[9]. We received 1 case of extraskeletal ewings sarcoma where in an adolescent boy presented with mass lesion over the lower back. CT scan showed a soft tissue mass in left chest wall confined to the muscle and underlying bone was unremarkable. Biopsy revealed small round cell tumor and IHC and molecular studies were confirmative. Patients with EES have a higher mean age, but also a bimodal distribution with EES more commonly found in those older than 35 and less than 5 years compared with skeletal tumors[10]. Two cases of Askin's tumor were also included in the study where

Table 3: Gender distribution

	NUMBER OF CASES	PERCENTAGE
MALE	35	58.30%
FEMALE	25	41.7

Table 4: Site distribution of bone and soft tissue tumors

	NUMBER OF CASES	PERCENTAGE
LOWER LIMB	26	43.30%
UPPER LIMB	16	26.70%
CHEST WALL	7	11.70%
PELVIS	5	8.30%
OTHERS	6	10%

in both the patients were young male in the age group of 20-30 years who presented with mass over the chest wall. Histological examination revealed small blue round cell tumor. IHC studies were confirmative. Askin's tumor is a primitive neuroectodermal tumor (PNET) of the thoracopulmonary region described first time in 1979 by Askin et al[11]. It develops from the soft tissues of the chest wall, particularly in the paravertebral region. Askin's tumor occurs in young Caucasian adults and is associated with poor prognosis. It is presented as a painful wall mass, often associated with dyspnea, cough, weight loss, Horner's syndrome, or regional lymphadenopathy. A chest wall soft-tissue density mass, sometimes associated with rib erosion and/or pleural effusion, is the commonest radiographic manifestation[12]. The most important role of the CT scan is to confirm the presence of a solid chest wall and to demonstrate their possible intrathoracic extension and/or direct lung invasion[12,13].

Clear cell sarcoma of soft part

We report a rare case of clear cell sarcoma of soft part. A 35 year female presented with soft tissue mass over the left thigh. MRI revealed an ill defined soft tissue mass involving the quadriceps of left lower thigh. Wide local excision of the mass was done and histopathology showed tumor composed of spindle cells arranged in fascicles having pleomorphic nuclei and mitosis was 9-10/HPF. On IHC tumor cells were positive for HMB 45, MITF and S-100. Therefore the diagnosis of clear cell sarcoma of soft part was confirmed. Margins were free of tumor and there was no lymph node metastasis. Post of follow up was uneventful. Clear cell sarcoma of tendons and aponeuroses was first described in 1965, as a rare malignant tumor originating from tendons and aponeurosis, with histological clear cell appearance due to the accumulation of glycogen[14]. In 1973, melanocytic differentiation was recognized by the presence of cytoplasmic melanosomes15. Clear cell sarcoma accounts for less than 1% of all soft tissue tumors[14,15,16,17]. Currently, clear cell sarcoma is a well documented, distinct clinicopathological entity, resembling melanoma and soft tissue sarcomas. Like melanoma, clear cell sarcoma shows melanocytic differentiation in almost all instances as a result of the translocation t(12;22)(q13;q12), a genetic event not seen in melanoma. Like soft tissue sarcomas, it shows deep soft tissue primary location, lacks cutaneous invasion, and has preference for lymph node and pulmonary metastasis[18].The treatment of choice for clear cell sarcoma is wide surgical resection.[19] If complete excision is achieved, adjuvant treatments are not necessary[20].

Conclusions

Multimodal therapies including surgery, chemotherapy and radiotherapy have improved clinical outcomes of patients with bone and soft tissue sarcomas there by plateauing the prognosis over the recent years. Molecular classification has emerged as a newer mode for specifically diagnosing the malignant tumors and also pre determining their prognosis. The above study describes the various bone and soft tissue tumors that were commonly encountered in a tertiary cancer institute and also highlights few of the rare soft tissue sarcomas. We conclude that bone and soft tissue tumors can affect a wide range of age group and show male preponderance. Early detection and treatment is essential for diagnosis of these malignant tumors. Molecular studies are most important in diagnosing, classifying and also prognosticating bone and soft tissue tumors.

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Conflict of Interest: NIL
Source of Support: NIL

How to Cite this Article

Ashwini Natekar A, Gupta G, Basu S. Spectrum of Bone and Soft Tissue Tumors in A Tertiary Cancer Institute in Eastern India. Journal of Bone and Soft Tissue Tumors July - Dec 2018;4(2): 7-10.