Distribution of pediatric bone and soft tissue tumors in the central Black Sea region

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This study aimed to evaluate pediatric cases treated surgically for an initial diagnosis of bone and soft tissue tumor between January 1987 and January 2012. This retrospective study evaluated 328 patients with pathologically confirmed tumor from a total of 374 patients hospitalized with an initial diagnosis of tumor. The cases were analyzed with respect to frequency, age, gender, and localization. One hundred and eighty (54.8%) males and 148 (45.2%) females, with a mean age of 13 years (range: 1-18 years), were included in the study. The tumors were determined as 258 (78.6%) bone tumors and 70 (21.4%) soft tissue tumors. The most common benign bone tumor was observed to be osteochondroma (n=61, 30.7%), and the most common malignant bone tumor was osteosarcoma (n=31, 52.6%). The most common benign soft tissue tumor was hemangioma, which is a vascular tumor (n=28, 43.8%), and the most common malignant soft tissue tumor was rhabdomyosarcoma (n=5, 83.3%). It is thought that similar studies will serve to form larger series and facilitate inter-regional comparisons by collecting data from centers that surgically treat bone and soft tissue tumors, thereby benefitting both pediatric and public health.

Key words: bone tumor, soft tissue tumor, pediatrics.

Turkey has a population of approximately 73 million, one-fourth of which are under 15 years of age and one-third of which are under 19 years of age. Pediatric cancers are the fourth most common cause of pediatric mortality following infection, cardiological pathologies and accidents[1,2]. Approximately 2000 new cases of pediatric cancer are reported each year. Of all the cancer cases in our country, bone tumors represent 6.1% and soft tissue tumors 6%[3]. With respect to the diagnosis, monitoring and treatment, pediatric bone and soft tissue tumors form a complex and difficult patient group. Therefore, just as for adult patient groups, it is appropriate that they be evaluated by a multidisciplinary and experienced team. Cases should be taken into centers that have orthopedics, pediatric oncology, radiology, pathology, radiation oncology, and nuclear medicine departments working together in collaboration.

In this study, 328 patients of 374 admitted to the Orthopedics and Traumatology Department between January 1987 and January 2012, who were reported to have a tumor based on results of the pathology examination, were evaluated with respect to tumor frequency, age, gender, and whether the localization showed any regional characteristics.

Material and Methods

The department records and pathology archives were examined for the period January 1987 – January 2012. From the department records, 374 cases with an initial diagnosis of tumor were determined to have been hospitalized. Pathology examination results confirmed 328 cases of tumor. A total of 46 cases were excluded from the study: 39 cases were discharged for reasons other than the tumor, and in 7 cases, the family changed their mind regarding the surgery. The tumors were

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separated into groups as bone tumors and soft tissue tumors and were examined with respect to frequency, age, gender, and localization.

Statistical analysis was conducted using the Statistical Package for the Social Sciences software (version 15.0; SPSS Inc., Chicago, IL, USA), which is a descriptive study.

Results

Of the 328 cases, 180 (54.8%) were male and 148 (45.2%) were female. The mean age of all cases was 13 years (range: 1-18 years). The localization of the lesions was determined as 95 (28.9%) in the femur and thigh, 80 (24.3%) in the knee joint, 57 (17.3%) in the tibia and calf, 52 (15.8%) in the humerus and arm, 41 (12.5%) in the wrist and hand bones, and 19 (5.7%) in the pelvis and gluteal region. The tumors were determined as 258 (78.6%) bone tumors and 70 (21.3%) soft tissue tumors.

One hundred and ninety-nine (77.1%) bone tumors were benign, and 59 (22.9%) were malignant, with 137 seen in males and 121 in females. The lesions were observed to be around the knee in 76 (29.4%), in the fingers in 26 (10.1%) and in the humerus in 26 (10.1%) cases.

When the benign bone tumors were examined, 100 were determined to be in males and 99 in females, the mean age was 12.9 years, and the most common benign bone tumor was osteochondroma (n=61, 30.7%), followed by solitary bone cyst (n=32, 16.1%), enchondroma (n=23, 11.6%), osteoid osteoma (n=21, 10.6%), and aneurysmal bone cyst (n=21, 10.6%) (Table I).

When the commonly seen benign bone tumors were examined, 29 (47.5%) of the osteochondroma cases had localization around the knee, 12 (19.6%) were in the fingers and toes, and 7 (11.5%) were in the humerus. Solitary bone cyst was seen in 11 cases (34.4%) in the proximal humerus, and a total of 20 cases (62.5%) were observed to have lesion localization in the humerus. Of the enchondromas, 20 cases (87%) were in the fingers. Osteoid osteoma lesions were observed in the femur in 7 cases (33.3%) and in the proximal and distal tibia in 5 cases (23.8%); a total of 13 cases (61.9%) had tibial localization of the lesion. Aneurysmal bone cyst was observed in 6 cases (28.5%) in the proximal humerus and in 3 cases (14.3%) in the femur.

Malignant bone tumors were seen in 37 males and 22 females, with a mean age of 14.3 years. The most frequently seen malignant bone tumor was osteosarcoma (n=31, 52.6%), followed by Ewing sarcoma (n=25, 42.4%) (Table II).

When the malignant bone tumors were examined, 21 cases (67.7%) of osteosarcoma were seen to be around the knee, and of the Ewing sarcomas, 7 cases (28%) were around the knee and in the iliac wing, 4 cases (16%) in the tibia, and 3 cases (12%) in the scapula.

When the soft tissue tumors were examined, 64 (91.4%) were benign and 6 (8.6%) were malignant, with 43 seen in males and 27 in females. The lesions were observed to be in the thigh in 14 cases (20%), in the forearm in 13 cases (18.6%), and in the wrist in 10 cases (14.3%).

Benign soft tissue tumors were observed in 41 males and 23 females, with a mean age of 12.2 years. The most common benign soft tissue tumors was observed to be hemangioma, which included vascular tumor (n=28, 43.8%), followed by lipoma (n=10, 15.6%) and cystic hygroma (n=9, 14.1%) (Table III).

The localization of the most frequently seen soft tissue tumors included hemangioma in the forearm (12 cases, 2.9%), lipoma in the thigh (8 cases, 80%), and cystic hygroma in the wrist (5 cases, 55.6%) and ankle (3 cases, 33.3%).

Malignant soft tissue tumors were seen in 2 males and 4 females, with a mean age of 10.5 years. The most common malignant soft tissue tumor was observed to be rhabdomyosarcoma (n=5, 83.3%).

When the most frequently seen malignant soft tissue tumors were examined, rhabdomyosarcoma was observed in the arm in 3 cases (60%) and in the thigh in 2 cases (40%) (Table IV).

When reasons other than tumor were examined, 17 cases (43.5%) were reported to have infection and 7 cases (17.9%) had normal tissue (Table V).

Discussion

Knowledge of tumor incidence, age, gender, and localization provides important clues to reach a diagnosis. There are very few studies in our country showing the distribution of bone and

soft tissue tumors. Studies on tumor patient series have been published by Gülman et al.[4], (1982-1986), Kösem and Bayram[5] (1994-2000), Solakoğlu and Benzer[6] (1990-2000), Güngör et al.[7] (2000-2007), and more recently by Yücetürk et al.[8] (1989-2009). In the current study, it was aimed to create a regional data source by examining the data of pediatric cases over a 25-year period.

Turkey has a population of approximately 73 million, one-fourth of which are aged below 15 years, and one-third of which are aged below 19 years. At a rate of 7.2%, pediatric cancers are the fourth most common cause of pediatric mortality following infection, cardiological pathologies and accidents[1,2]. Every year in Turkey, 2500-3000 new pediatric cancer cases are expected, although according to the records of the Turkish Pediatric Oncology Group (TPOG) and the Turkish Pediatric Hematology Association (TPHA), approximately 2000 new pediatric cancer cases are reported each year. Of the total pediatric cancer cases in our country, 6.1% are seen to be bone tumors and 6% soft tissue tumors[3].

The total bone and soft tissue tumors of this study comprised 263 benign tumors and 65 malignant tumors, respectively, with an approximately four-fold difference between them. Benign tumors are seen more often than malignant tumors in the general population. In a study by Yücetürk et al.[8], the number of malignant tumors was determined to be greater, which was reported to be due to benign tumors being treated at external healthcare centers. In this respect, our clinic can be considered as a center for pediatric tumor patients in the region.

Although there is no greater rate for sarcomas in the extremities among all malignancies in the body, the malignant tumors determined are nearly all sarcomas[5]. In pediatric cases, the most feared and most commonly seen malignant bone and soft tissue tumors are osteosarcoma, Ewing sarcoma and rhabdomyosarcoma[9]. These tumors represent 10% of all newly diagnosed pediatric cancer cases[10]. In the current study, these three tumors were seen to account for approximately 94% of the malignant tumors. In this respect, the importance of the subject is obvious.

Bone tumors are seen more often in males, benign tumors are more common than malignant tumors, and the most common location is around the knee[7]. The findings of the current study conformed with this, in that they were seen slightly more in males than females, benign tumors were seen more often, and they were most often around the knee.

The most commonly seen benign bone tumor is osteochondroma[6]. Osteochondromas represent 10-15% of all bone tumors and 20-25% of all benign bone tumors[11]. In children, osteochondroma is the most commonly seen lesion in the distal femur and proximal tibia[12]. Güngör et al.[7] reported that there was most

Table I. Distribution of Benign Bone Tumors (n=199)

Benign bone tumors	Number	%
Osteochondroma	61	30.7
Solitary bone cyst	32	16.1
Enchondroma	23	11.6
Osteoid osteoma	21	10.6
Aneurysmal bone cyst	21	10.6
Chondroblastoma	12	6
Non-ossified fibroma	12	6
Giant cell tumor	7	3.5
Fibrous dysplasia	3	1.5
Eosinophilic granuloma	2	1
Fibroma	2	1
Simple cyst	2	1
Osteoma	1	0.5
Total	199	100

Table II. Distribution of Malignant Bone Tumors (n=59)

Malignant bone tumors	Number	%
Osteosarcoma	31	52.5
Ewing sarcoma	25	42.3
Lymphoid origin tumors	3	5
Total	59	100

Table III. Distribution of Benign Soft Tissue Tumors (n=64)

Benign soft tissue tumors	Number	%
Hemangioma (vascular tumor)	28	43.8
Lipoma	10	15.6
Cystic hygroma	9	14.1
Neural origin tumors	4	6.2
Giant cell tumor of the tendon sheath	4	6.2
Lymphangioma	3	4.6
Villonodular synovitis	2	3.1
Desmoid tumor	2	3.1
Hamartoma	1	1.5
Fibroma	1	1.5
Total	64	100

often humerus proximal involvement of solitary bone cyst, while femoral proximal involvement was seen most often for osteoid osteoma. Enchondromas are often located in the short bones of the hand and foot[13-15]. Aneurysmal bone cyst often occurs in the metaphysis of the long bones[14]. Of the benign bone tumors of the current study, the most frequent was osteochondroma (61 cases), followed by solitary bone cyst (32 cases), enchondroma (23 cases), osteoid osteoma (21 cases), and aneurysmal bone cyst (21 cases). Osteochondromas were seen at a rate of 47% in the area around the knee; solitary bone cyst at 62.5% in the humerus, especially in the proximal part; enchondroma at 87% in the fingers; osteoid osteoma at 33.3% in the proximal femur; and aneurysmal bone cyst at 28.5% in the proximal humerus.

Osteosarcoma is the most frequently seen malignant tumor in children with an annual incidence of $4.4/10^6$, peaking in the second decade of life and showing no gender or race characteristic[10,16]. Ewing sarcoma is the second most frequently seen malignant tumor in children, with an annual incidence of $2.9/10^6$, and that is also seen most often in the second decade, but is not seen often in Asia and Africa[17-19]. The incidence in America has been reported as $2.1/10^6$. There are studies stating that osteosarcoma and Ewing sarcoma occur more often in males than females[20,21]. The localization of osteosarcoma is most often around the knee, whereas Ewing sarcoma has been reported at rates of 60% in the pelvis and lower extremity long bones[6,22,23]. In the current study, osteosarcoma was the most prevalent of the malignant bone tumors, with 31 cases, and Ewing sarcoma was seen in 25 cases. Both of these tumors were seen more in males than females. Osteosarcoma was observed most frequently around the knee, and the majority of Ewing sarcomas were in the pelvis and lower extremities.

Benign tumors are seen more often than malignant tumors in the general population. Of the soft tissue tumors examined in the current study, 64 were benign and 6 were malignant, they were observed more in males than females, and localization was observed to be mostly in the thigh and forearm.

Hemangioma is the most frequently seen benign tumor in childhood, encountered 2-5 times more in females, with localization in the extremities at a rate of 15%[24]. Lipomatous tumors represent 6% of all soft tissue tumors

Table IV. Distribution of Malignant Soft Tissue Tumors (n=6)

Malignant soft tissue tumors	Number	%
Rhabdomyosarcoma	5	83.3
Malignant mesenchymal tumor	1	16.7
Total	6	100

Table V. Distribution of Reasons Other than Tumor (n=39)

Reasons other than tumor	Number	%
Bone and soft tissue infection	17	43.5
Normal tissue	7	17.9
Acute-chronic inflammation	7	17.9
Callus tissue	4	10.3
Myositis ossificans	2	5.1
Synovial hypertrophy	1	2.6
Metabolic disease	1	2.6
Total	39	100

in children, and 94% of these are benign lesions[25]. Lipomas are rarely seen in the first 20 years of life[26]. Cystic hygroma seen in the hand, wrist and ankle is the most commonly encountered type of soft tissue tumor and is the most common soft tissue tumor seen in the upper extremities, most frequently occurring in females aged 30-50 years[27,28]. In the current series, hemangioma was the most common soft tissue tumor, with 28 cases, followed by lipoma in 10 cases and cystic hygroma in 9 cases. Hemangioma was observed in the forearm at a rate of 42.9%, lipoma in the thigh at 80%, and cystic hygroma in the wrist at 55.6%.

Among solid tumors in children, rhabdomyosarcoma is the fourth most frequently seen, accounting for more than half of malignant soft tissue tumors, with an annual incidence of 4.5/10^6. In the United States, the incidence for those below the age of 15 years has been reported as 4-7/10^6 [30]. It is seen more often in those of African ethnicity and in Asian children than in Caucasians. In conformity with the literature, the most frequently seen malignant soft tissue tumor in the current series was rhabdosarcoma, with 5 cases (3 in the arm, 2 in the thigh).

When reasons other than tumor were examined, 17 cases were determined to have infection, and seven cases were determined to have normal tissue in the lesion. This may be related to the radiological and clinical findings of infection mimicking a tumor[8]. In this respect, in the differential diagnosis of tumors, particularly those reported to be Ewing sarcoma, infection must certainly be borne in mind[31,32]. It has also been observed that the reactional process of normal tissue may result in findings similar to a tumor.

In general, it was observed that some data showed similarities while other data showed differences. In this regard, it can be said that although tumors have a specific character, regional differences may be shown. Therefore, by collecting data from centers that treat bone and soft tissue tumors surgically, these types of studies can form larger series and facilitate inter-regional comparisons, thereby benefitting both pediatric and public health.

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