Original Article

Juxtacortical Chondrosarcoma: Analysis of 52 Cases

Jukstakortikal Kondrosarkom: 52 Vakanın Analizi

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ABSTRACT

Introduction: Juxtacortical (surface) chondrosarcoma (JCC) is a very rare subtype of chondrosarcoma, mostly seen in young adults. In this study, we aimed to report demographic data and average survival rates of JCC.

Methods:In this study we used the latest version of the Surveillance, Epidemiology and End Results (SEER) database, patients were examined under the titles of gender, age, race / ethnic origin, lesion location in the body, degree of tumor differentiation, applied surgeries and follow-up periods. Descriptive statistics were given as mean ± standard deviation, frequency and percentage.

Results: A total of 52 patients were included in the study, of which 16 were female and 36 were male (69%), with a mean age of 41.3 ± 20.0 years (range: 8-84 years). When examined for ethnicity, it was found that it was seen most in white race (84.6%).The most common localization of tumor lesions was the lower extremity long bones (50%).Low grade tumor (45% of all tumors) was the most common tumor differentiation. The mean follow-up was 125,5 months (range 4 to 358 months).

Discussion: JCC is a very rare type of chondrosarcoma, and often seen as lower grade compared to other types of chondrosarcoma. It's a little more common in men. It tends to settle on the long bones, most commonly on the lower extremity, especially the femoral diaphysis.

Keywords: Juxtacortical chondrosarcoma, SEER, demographic, survive

ÖZET

Giriş: Jukstakortikal (yüzey) kondrosarkom (JKK), kondrosarkomların daha çok genç erişkinlerde görülen çok nadir bir alt tipidir. Bu çalışmada JKK'ye ait demografik verilerin ve ortalama sağ kalım oranlarının raporlanması amaçlandı.

Yöntemler: Sürveyans, Epidemiyoloji ve End Results (SEER) veritabanının en son versiyonu kullanılarak yapılan bu çalışmada, hastalar, cinsiyet, yaş, ırk/etnik köken, lezyonun vücutta yerleşim yeri, tümörün diferansiasyon derecesi, uygulanan cerrahiler ve takip süreleri altbaşlıklarında incelendi. Tanımlayıcı istatistikler ortalama ± standart sapma, sıklık ve yüzde olarak verildi.

Bulgular: Çalışmaya toplam 52 hasta dahil edildi, bu hastaların 16'sı kadın 36'i (%69) erkekti ve yaş ortalamaları 41,3±20,0 std (8-84 yaş arası) idi. Etnik köken açısından incelendiğinde en fazla beyaz ırkta (%84,6) görüldüğü saptandı. Tümöral lezyonların en sık görüldüğü lokalizasyon alt ekstremite uzun kemikleri ve ilişkili eklemleri (%50) idi. Tümör diferansiasyonları açısından incelendiğinde en sık düşük dereceli tümörler (tüm tümörlerin %45'i) görüldü. Ortalama takip süresi 125,5 ay (4-358 ay arası) idi.

Sonuç: JKK çok nadir bir kondrosarkom türüdür ve sıklıkla diğer kondrosarkom tiplerine göre daha düşük grade'li olarak görülür. Erkeklerde biraz daha sıktır. En sık alt ekstremiteye, özellikle de femur diafizine olmak üzere, uzun kemiklere yerleşim eğilimindedir.

Anahtar Kelimeler: Jukstakortikal kondrosarkom, SEER, demografi, sağkalım

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Introduction

Chondrosarcomas are the third most common malignant bone tumors and contain about 16% of all bone tumors. Chondrosarcomas have many subtypes such as classical, mesenchymal, myxoid, dedifferentiated, clear cell and secondary chondrosarcoma, and it is difficult to differentiate between osteochondromas and grade I chondro-sarcoma [1]. Juxtacortical chondrosarcoma (JK), also known as periosteal chondrosarcoma, is a rare chondrosarcoma variant originating from the outer surface of the bone [2,3]. This tumor accounts for about 2% of all chondrosarcomas [4].

Periosteal chondrosarcomas tend to be seen most frequently between the second and fourth decades and are more common in men [2,4,5].

These tumors most often settle in long bones, such as the femur and humerus. Clinically, as with most other malignant tumors, the most common admission symptoms are pain and / or swelling. Generally, it is a long-term and slowly progressing tumor [3,4,6].

Periosteal chondrosarcomas are often seen as a soft tissue mass with a juxtacortical location on radiographs. They may include features such as peripheral calcification, mottled appearance, popcorn appearance, which are features of cartilaginous tumors. As with many tumors in MRI, they show hypo-intense in T1 sequences and hyper-intense in T2 sequences. Some patients may have punctate mineralization foci. They show peripheral and septal enhancement after gadolinium injection [2,5,6].

The gold standard treatment method of JKs, as with many other malignant tumors, is the resection of the tumor with wide limits. Since it is a periosteal located tumor, it is vital to determine whether the tumor contains the medullary cavity before surgery, because if there is medullary involvement, it should be treated like central chondrosarcoma [2,3,6].

To our knowledge, there is no study on SEER database data and juxtacortical chondrosarcoma that analyzes the epidemiology and survival rates of the tumor. In this study, we studied demographic data, surgical treatments, and survival rates for this disease.

Materials and Methods

In this study, we used the data of the database Surveillance, Epidemiology and End Results (SEER), which was released in 2018. We evaluated juxtacortical chondrosarcoma cases retrospectively. The cases included in the study in this version include patients diagnosed between 1983 and 2016.

As it is known, the SEER database is the most comprehensive registry of tumor patients in the United States. The SEER database contains data on patients' demographic data, cancer incidence and survival times [7]. This database has been used in many surgical subfields and cancer-related studies today and has been used in many studies in the past. In the United States, all cancer data of the country age has been collected through this database since 1973, and the information for this database currently represents data for about 28% of the entire US population [7,8]. This data base is a valuable resource especially for the examination of very rare diseases. It is a database where statistical inferences can be made about these diseases thanks to its ability to provide extensive data. Juxtacortical chondrosarcoma is also one of the very rare subtypes of chondrosarcoma.

In this study, we included cases with Juxtacortical chondrosarcoma (ICD O3 code 9221/3) among the cases with Chondrosarcoma in the International Classification of Diseases for Oncology, Third Edition (ICD-O 3) morphology [9] coding system. The patients were evaluated in terms of age, gender, race (ethnicity), marital status, the direction of the disease, tumor location, tumor grade, follow-up time and follow-up results. In addition, we classified all patient ages in 5 slices. The races of the patients were examined in 3 groups, white races, black races and other races (Asian Pacific, American Indian etc.). Their marital status was grouped as single or married, divorced patients were included in the single group. Histological grades of tumors were examined at four degrees.

As it is known, the studies written with the use of SEER database data are exempt from the approval of the Ethics Review Board. Therefore, no ethics committee application was made for our study.

Statistical Analysis

All statistical analyzes were done using IBM SPSS 22.0 statistical software (IBM Corp, Armonk, NY, USA). Descriptive statistics were expressed as mean ± standard deviation, frequency and percentage.

Results

A total of 52 patients, 36 (69%) males and 16 females, with an average age of 41.3 ± 20.0 std (8-84years), were included in this study.44(84.6%) of the patients were from the white race, 4 were from the black race and 4 were from the other races group.

When patient ages are examined in groups of 5 years, 1 patient is in the 5-9 age group, 5 patients are in the 10-14 age group, 1 patient is in the 15-19 age group, 6 patients are in the 20-24 age group, 3 patients are in the 25-29 age group, 4 patients in the 30-34 age group, 7 patients in the 35-39 age group, 4 patients in the 40-44 age group, 4 patients in the 45-49 age group, 3 patients in the 50-54 age group, 3 patients in the 55-59 age group, 2 patients In the 60-64 age group, 4 patients were in the 65-69 age group, 2 patients were in the 70-74 age group, 1 patient was in the 75-79 age group, and 2 patients were in the 80-84 age group. As stated, there was no aggregation in any age group.

When tumor localizations were examined, the tumor was found in 8 patients in the region containing the upper extremity long bones, scapula and associated joints, in 1 patient in the region containing the upper extremity short bones and related joints. In 50% of 52 patients, the region containing the lower extremity long bones and associated joints, in the area of the lower extremity short bones and associated joints in 1 patient, in the vertebral column in 1 patient, in the costa, sternum, clavicle and associated joints in 8 patients, and in the pelvic area in 6 patients bones were in the region containing sacrum, coccyx and related joints. In one patient, the tumor was located on the bone, but its localization was not reported.

When the histological grades of the tumors were examined, 23 patients (44.2%) had grade I (good differentiated) level, 19 patients had grade II level (moderately differentiated), 5 patients had grade III level (poorly differentiated) and 2 patients had grade IV level (un- differentiated, anaplastic) tumor. Grade information of 3 patients was not available.

When evaluated in terms of tumor direction, in 24 patients (46.1%) the tumor was located on the left side of the body, 20 tumors were located on the right side. Eight patients did not have direction information.

Surgical resection in 24 (46.1%) patients, local excision in 13 patients, partial resection in 5 patients, hemipelvectomy in one patient was performed. One patient refused surgical treatment, one patient died before surgery, the information about the surgery performed in seven patients was not available in the data.

The mean tumor size of 26 patients was 82.4 mm (between 27 mm and 220 mm).

The mean follow-up period of the patients was 125.5 months (4-358 months). Forty-six patients were still alive and had an average follow-up time of 130 months, while eight patients (15.3%) died in an average of 98 months.

Finally, 24 of the patients were single (19 were never married, five were divorced), 24 were married (46.1%), and there was no information about the marital status of 4 patients.

Discussion

The aim of this study was to identify patients diagnosed with juxtacortical chondrosarcoma, a very rare variant of chondrosarcoma, using the SEER database of the United States, and to determine their demographic characteristics, surgeries, and survival rates. The database we have mentioned provides us with a high number of patients about very rare tumors such as juxtacortical chondrosarcoma

Table 1. Demographic Data of Patients Included

Data	n=52
Gender, n (%)	
Male	36 (69.0)
Woman	16 (31.0)
Age, years	
Mean ± SD Median (min-max)	41.3 ± 20.0 38 (8.0-84.0)
Race, n (%)	
White	44 (84.6)
Black Other	4 (7.7) 4 (7.7)
Marital Status (n = 48), n (%)	
Single	24 (50.0)
The married	24 (50.0)
Direction (n = 44), n (%)	
Left Right	24 (46.1) 20 (53.9)
Tumor Localization, n (%)	
Long extremity bones, scapula Short limbs	8 (15.3)
of the upper limb	1 (1.9)
Lower extremity long bones, joints Lower	26 (50)
extremity short bones, joints Vertebral	1 (1.9)
column	1 (1.9)
Costa, sternum, clavicle	8 (15.3)
Pelvis, sacrum, coccyx, joints	6 (11.5)
Tumor Degree n (%)	
Grade I	23 (44.2)
Grade II	19 (36.5)
Grade III	5 (41.6)
Grade IV	2 (3.8)
Follow-up Time, months	125.5
Average	176 (4-358.0)
Median (min-max)	
Follow-up Results, n (%)	
Live	44 (84.7)
Exitus	8 (15.3)

and enables us to obtain detailed information about these diseases [8].

When the data of our country related to the musculoskeletal system tumors are examined, it is seen that it is quite similar to the world literature. According to studies conducted in our country, chondrosarcomas are the third most common malignant bone tumor following osteosarcoma and Ewing's sarcoma [10].

Not much data is available in the literature about the rare variant of chondrosarcoma JKK. Since it is a very rare tumor, there are usually case reports or case series [4,11,12,13]. In this study, a total of 52 patient data were obtained using SEER data between

1983-2016. To our knowledge, we offer the largest series in the literature.

In the literature, it has been reported that the most common age of periosteal chondrosarcoma is between the 2nd and 4th decades, and periosteal chondrosarcoma tendsto occur more frequently in men [2,4,5]. In our studies, it was found that it is more common in men. In terms of age, the average age of the patients was 41 in our study, and it was found that the age of the patients was distributed over a wide age range, with no frequency observed in any age range.

Classical chondrosarcomas of ten tend to settle in the pelvic bones and proximal appendicular skeleton [10]. When the archive data were examined in this study, the most common locations were the lower limb long bones and associated joints.

On the radiographs of the chondrosarcoma, a smoothly bounded soft tissue mass is located on the bone surface, and specific findings can be detected for cartilage tumors. Periosteal bridge formation is frequently found at the borders of the tumor due to chronic periosteal reaction. In radiographic examination of periosteal chondrosarcoma, MRI is superior to other imaging modalities, as with many musculoskeletal tumors, because it is very useful in studying intramedullary space and soft tissue extension with superior contrast resolution [2,3,6].

In the differential diagnosis of periosteal chondrosarcoma, especially the benign tumor that should be kept in mind is the periosteal chondroma and the malignant tumor is the periosteal osteosarcoma. Although it is known that osteosarcoma is seen at an earlier age (often 1st and 2nd decade) compared to chondrosarcoma, juxtacortical chondrosarcoma can be seen in a wide age range including these ages. The presence of extensive cortex destruction and periosteal reaction on radiographs should raise suspicion in terms of osteosarcoma [5,6,14]. Periosteal chondroma also tends to occur at an earlier age and is a benign tumor, often without symptoms. Apart from these tumors, it is important to make differential diagnosis in terms of other benign and malignant tumors and tumor-like lesions [1,10].

Treatment of periosteal chondrosarcoma is resection of the tumor with wide surgical margins. The response of chondrosarcoma to chemotherapy and radiotherapy is low [11]. Recurrence of the disease is most often associated with the inability to remove the tumor with wide limits. [2,4,6,11-13]. Since the chondrosarcoma is a tumor of the periosteal location, central site location is very rare, but patients should be checked for intramedullary placement in pre- operative evaluation [2,3,6,15]. Cases with intramedullary localization should be treated like centrally located chondrosarcoma.

Histological ratings of chondrosarcoma are associated with the clinical behavior of the disease and prognosis. Grade I tumors have better prognosis, while the prognosis worsens as grade level increases. Grade III tumors tend to metastasize 70% [3]. In general, the prognosis of JK is much better than central chondrosarcoma. Metastases are very rare and occur very late in parallel with the slow course of the disease [2,3,6]. In our study, more than 40% of patients were grade I tumors, and approximately 85% of patients on average 125-month follow-up were still alive and underfollow-up.

This study has some limitations. Firstly, the data were analyzed retrospectively and the number of patients was relatively low as it is a rare disease. Because this study was a study analyzing a database data and detailed data could not be found in the database, some patients could not be included in statistical studies. Some of the demographic data and all symptoms, treatments and similar data were not mentioned. For this reason, there is a need for large multicentre studies with a higher number of patients in the future.

Conclusion

Periosteal chondrosarcoma is a malignant tumor that can be seen in a wide age range and tends to occur more frequently in men. Its prognosis is quite good compared to central chondrosarcoma after appropriate surgical treatments.

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