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Chondrosarcomas of the phalanges of the hand

El falankslarının kondrosarkomları

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ABSTRACT

Objectives: This study aims to evaluate the diagnosis and treatment approaches of the rarely seen chondrosarcomas of the phalanges of the hand.

Patients and methods: Fifty-two patients (27 males, 25 females; mean age 41.2 years; range 12 to 70 years) with chondroid lesions localized in hand phalanges who were performed surgical treatment between December 2012 and September 2016 were retrospectively reviewed. The study included 62 phalangeal chondroid lesions. Patients' mean follow-up duration was 60.6 months (range 13 to 165 months). Incisional biopsy was performed for the diagnosis. One patient with bilateral and multiple involvement was performed tru-cut biopsy. Phalangeal chondrosarcoma was diagnosed in five patients (9.6%).

Results: Of the chondroid lesions, 37 were localized in proximal phalanges (59.6%), 16 in midphalanges (25.8%), and nine in distal phalanges (14.6%). Chondrosarcoma was detected in 15 phalanges of five patients. Of the two patients with Ollier disease, localization was detected in nine phalanges (four proximal, two mid, three distal phalanges) of one patient and in three phalanges (one proximal, two midphalanges) of the other patient. None of the patients had distant metastasis on diagnosis. Ray amputation was performed in two patients under general anesthesia and amputation was performed in one patient. One patient did not give consent for operation. The other patient with Ollier disease gave consent for amputation of only one finger. No local recurrence was seen.

Conclusion: The hand localization of chondrosarcomas is rare with scarce information in the literature. Their metastasis potential is low but local recurrence rates are high after insufficient surgery. Amputation or ray amputation is the applicable treatment.

Keywords: Chondroid lesion; chondrosarcoma; hand phalanges.

ÖZ

Amaç: Bu çalışmada nadir görülen el falankslarının kondrosarkomlarının tanı ve tedavi yaklaşımları değerlendirildi.

Hastalar ve yöntemler: Aralık 2012-Eylül 2016 tarihleri arasında el falankslarında yerleşimli kondroid lezyonu olan ve cerrahi tedavi uygulanan 52 hasta (27 erkek, 25 kadın; ort. yaş 41.2 yıl; dağılım 12-70 yıl) retrospektif olarak gözden geçirildi. Çalışmaya 62 falangeal kondroid lezyonu alındı. Hastaların ortalama takip süresi 60.6 aydı (dağılım 13-165 ay). Tanı için insizyonel biyopsi uygulandı. İki taraflı ve multipl tutulumlu bir hastaya tru-cut biyopsi yapıldı. Beş hastada falangeal kondrosarkom tanısı konuldu (%9.6).

Bulgular: Kondroid lezyonların 37'si proksimal falanks (%59.6), 16'sı midfalanks (%25.8) ve dokuzu distal falanks (%14.6) yerleşimliydi. Beş hastanın 15 falanksında kondrosarkom saptandı. Ollier hastalığı olan iki hastanın birinin dokuz falanksında (dört proksimal, iki mid, üç distal falanks), diğerinin üç falanksında (bir proksimal, iki mid falanks) yerleşim saptandı. Tanı sırasında hastaların hiçbirinde uzak metastaz yoktu. İki hastaya genel anestezi altında ray ampütasyon ve bir hastaya amputasyon uygulandı. Bir hasta ameliyat için onay vermedi. Ollier hastalığı olan diğer hasta sadece tek parmak ray amputasyonunu kabul etti. Lokal nüks görülmedi.

Sonuç: Kondrosarkomların el yerleşimi nadir olup konu hakkında literatürde az bilgi vardır. Metastaz potansiyelleri düşük olmakla beraber yetersiz cerrahi sonrası lokal nüks oranları yüksektir. Geçerli tedavi yöntemi amputasyon ya da ray amputasyondur.

Anahtar sözcükler: Kondroid lezyon; kondrosarkom; el falanksları.

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Chondrosarcoma is a rare malignant bone tumor which stems from proliferated cartilage tissue. It is occasionally seen in pelvis, proximal femur, and proximal humerus.[1,2] Chondrosarcomas generally arise as painful, slowly growing masses and fifth and sixth decades are the most frequent ages.[1] The most common cartilaginous tumors of the hand and small bones of the hands are enchondromas that rarely transform into chondrosarcomas. Even if malignant transformation of multiple enchondromatosis is common, hand involvement of enchondromatosis is not common.[1] Less than 0.5% of all chondrosarcomas are localized into hand.[3] The differentiation of phalangeal chondrosarcomas from enchondromas is difficult both for pathologists and the clinicians. If there is any doubt in the radiographic and clinical evaluation, magnetic resonance imaging (MRI) and histopathologic evaluation must be performed. Limited and marginal interventions lead to a rise in relapses. The proper and definitive treatment is amputation.[1,4] In this study, we aimed to evaluate the diagnosis and treatment approaches of the rarely seen chondrosarcomas of the phalanges of the hand.[5]

Figure 1. Ollier disease (bilateral and multiple involvement).

PATIENTS AND METHODS

Fifty-two patients (27 males, 25 females; mean age 41.2 years; range 12 to 70 years) with chondroid lesions localized in hand phalanges who were performed surgical treatment at Ankara Oncology Training and Research Hospital between December 2012 and September 2016 were retrospectively reviewed. Patients applied with masses with or without pain and pathologic fracture. The mean follow-up period was 60.6 months (range 13 to 165 months). There were chondroid lesions in 62 phalanges of the 52 patients. Two patients had Ollier disease. Unilateral and multiple involvements were present in one and bilateral and multiple

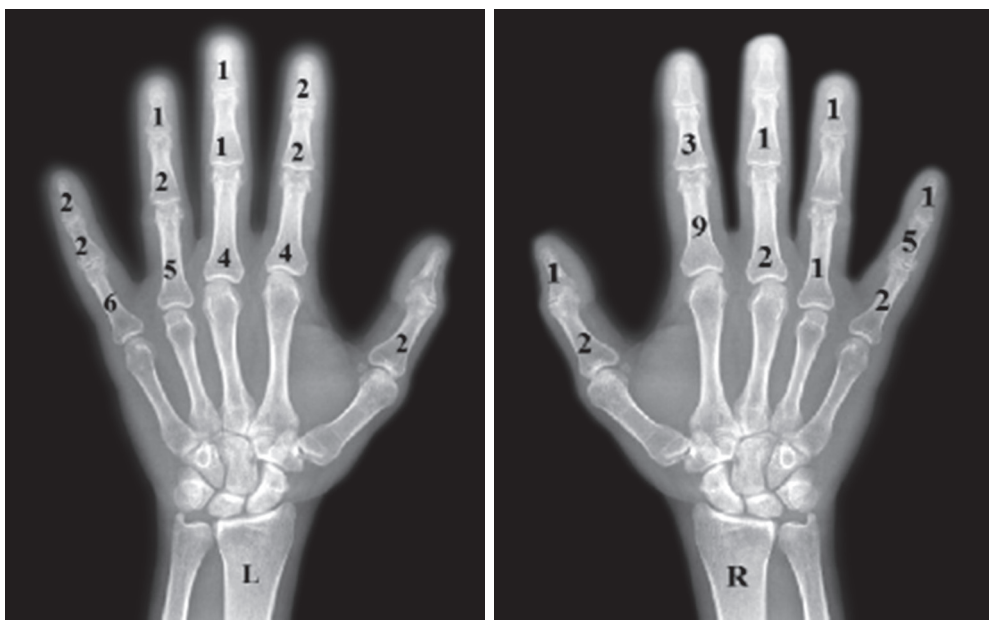

Figure 2. Distribution of chondroid lesions localized to phalanges.

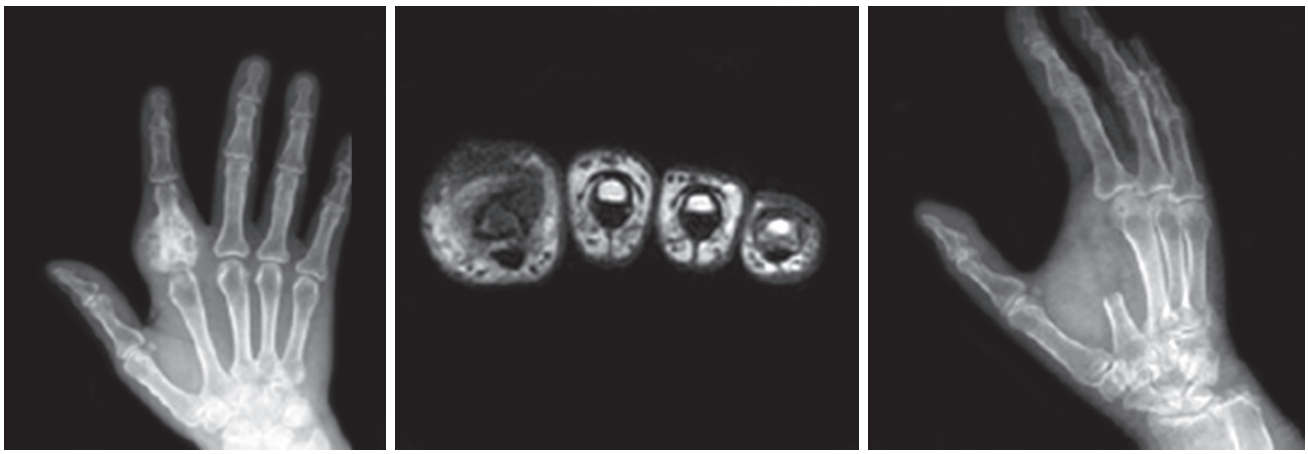

Figure 3. Chondrosarcoma case of proximal phalanx of second finger without soft tissue involvement and treatment with amputation.

involvements were present in the other patient (Figure 1). The study protocol was approved by the Ankara Oncology Training and Research Hospital Ethics Committee. A written informed consent was obtained from each patient. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Of the chondroid lesions, 37 were localized in proximal phalanges (59.6%), 16 in midphalanges (25.8%), and nine in distal phalanges (14.6%). Right hand involvement was present in 28 patients (45.1%), and left hand involvement was present in 34 patients (54.8%) (Figure 2). All patients were evaluated with conventional direct graphies. Patients who were suspected to have chondrosarcoma were evaluated with MRI. Tru-cut biopsy was performed for a patient with Ollier disease due to bilateral and multiple involvements. Except this patient, all patients underwent incisional biopsy. Ray amputation was performed for three patients and amputation was performed for one patient (Figure 3).

RESULTS

Chondrosarcoma diagnosis was defined for totally 15 phalanges of five of 52 patients (9.6%) with chondroid lesion in hand phalanges after histopathologic evaluation. The ages of patients were 23, 32, 47, 68, and 70. Three phalanges (one proximal, two midphalanges) of one patient and nine phalanges (four proximal, two mid, three distal phalanges) of the other patient with Ollier disease were involved. None of the patients had distant metastasis on diagnosis. One patient did not approve operation. One patient approved ray amputation only for one finger. The mean follow-up period of chondrosarcomas' patients were 53.4 months (range, 13 to 132 months) (Table I).

DISCUSSION

In this study, we evaluated the diagnosis and treatment approaches and localization features of chondrosarcoma cases of hand phalanges, which are rare and may lead to difficulties in both diagnosis and treatment. The hand involvement rate of

TABLE I

Clinical features of chondrosarcomas

No	Age/Gender	Localization	Symptom	Treatment	Follow-up period (months)
1	70/F	Left 4 PF	Pain and mass	Ray amputation	132
2	32/M	Left 5 PF	Mass and pathologic fracture	Ray amputation	72
3	68/F	Right 2 PF	Pain and mass	Amputation	14
4	47/M	Right 5 MF 3 MF Right 2 PF	Mass and pathologic fracture	Ray amputation (Right 2)	36
5	23/M	Left 4-5 PF/MF/DF Right 1-3 PF/1 DF	Mass and pathologic fracture	-	13

PF: Proximal phalanx; MF: Medial phalanx; DF: Distal phalanx.

Figure 4. Pop-corn-like lesion and intralesional calcification.

chondrosarcoma changes between 0.5% and 3.2%.[2,5] Nigrisoli et al. revealed four patients (<1%) with hand involvement of chondrosarcoma in a 506-patient-series, while Unni et al. revealed 1.5% of chondrosarcoma diagnosis in a 635-patient-series.[6] To our knowledge, there is no study on the rate of malignancy of chondroid lesions with sole hand involvement in the literature. Moreover, there is no percentage reported for the localization frequencies in hand. In this study, five of 52 patients' phalangeal chondroid lesions were diagnosed as chondrosarcoma (9.6%) that is prominently high with respect to even only the rate of hand chondrosarcomas. In the diagnosis of chondrosarcomas localized in phalanges, cortical destruction, soft tissue involvement, periosteal reaction and intralesional calcification (pop-corn like lesion) are signs of potential malignancy in direct radiography (Figure 4).[2,6] Particularly, the differentiation of low grade chondrosarcomas and enchondromas is challenging for both clinicians and pathologists. Clinical and radiologic features must be carefully followed-up. The changes in the character of pain, cortical thinning, soft tissue invasion of the lesion, and destruction in bone are warnings for malignancy. Magnetic resonance imaging is the basic radiologic evaluation method for the definition of pathologic and normal bone margins, intramedullary, extraosseous and soft tissue invasion.[7] The definitive diagnosis of the patients is established with clinical examination, conventional radiography followed by MRI and biopsy and histopathologic evaluation. In this study, five of six cases followed with suspicion of chondrosarcoma and possible chondrosarcoma diagnosis with MRI were diagnosed as chondrosarcoma histopathologically; however, the diagnosis of the sixth case did not support chondrosarcoma. The most common localization of the chondroid tumors was proximal phalanx. This result is similar to that of other studies.[6,7] Of all chondroid tumors in our study, 59.6% were on proximal phalanx; in a similar way, chondrosarcoma cases were mostly localized in proximal phalanx (eight of 15 phalanges, 53.3%). To our knowledge,

Figure 5. A case with ray amputation of second finger.

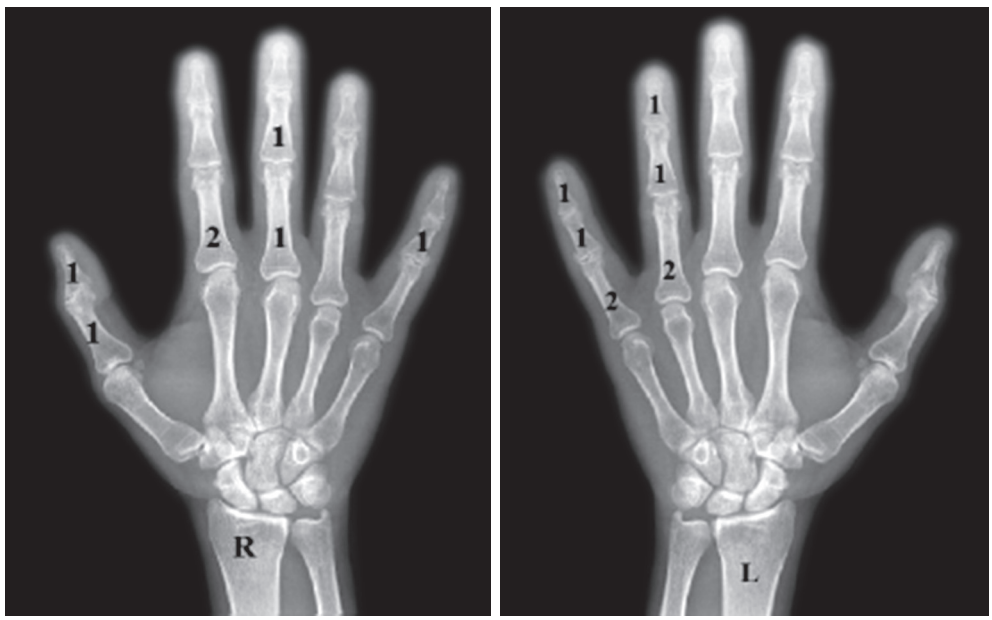

Figure 6. Distribution of chondrosarcomas localized to phalanges.

there is no study in the literature regarding which hand is mostly involved with chondrosarcomas. In our study, eight were localized at the left hand and seven were localized at the right hand. According to these results, there is no difference between right or left hand involvement.

Two cases in our study were diagnosed as Ollier disease. Pathogenesis of Ollier disease is not known; it is characterized with three enchondromas and its major complication is malignant transformation (20-50%), with a prevalence of 1/100,000. It is mostly localized in pelvis, proximal humerus, and distal femur.[8] The patient with bilateral hand involvement in our study did not approve the treatment. The other patient was proposed a ray amputation for second and fifth phalanges. However, the patient gave consent for ray amputation for only the second phalanx (Figure 5). It is reported in the literature that phalangeal chondrosarcomas are seen most commonly in the fifth phalanx and least commonly in the fourth phalanx.[9] Of the 15 phalangeal chondrosarcomas in our study, five of the chondrosarcomas were on the fifth finger, four were on the fourth finger, two were on the third finger, two were on the second finger, and two were on the first finger. Fifth finger was the most common localization in harmony with the literature (33.3%). Fourth finger was found to be the second most common localization in this study (26.6%) (Figure 6).

Chondrosarcomas are resistant to chemotherapy and radiotherapy. Chemotherapy has no place in up-to-date treatment. Metastasis of the hand chondrosarcomas are rare.[10] In their series, Bovée et al.[10] found no metastasis in 28 of 35 hand and foot chondrosarcoma patients. They reviewed the literature until 1999 and detected only two of 84 patients that distant metastasis occurred and they concluded that phalangeal chondrosarcomas were locally aggressive and had very low metastatic potential. Furthermore, Mankin[11] reported that hand chondrosarcomas had a more benign course than that of other involvements in skeletal system. Patil et al.[6] with their 23-patient-series of metacarpal and phalangeal chondrosarcomas reported no distant metastasis similar to Mittermayer et al.[12] with their 13-patient-series of hand chondrosarcomas. In our study, there was no distant metastasis during the follow-up period. Intralesional limited surgical interventions such as curettage have been increasing the local recurrence rates.[1,2,6,13] There are 11%-50% local recurrence rates reported in the literature.[6,14] While Bovée et al.[10] did not observe any local recurrence after amputation or ray amputation for the 28 of the 35 patients in their series; they reported local recurrence in 10 patients who underwent marginal resection. In addition, Demireli et al.[13] reported that they performed amputation for patients followed-up with the prediagnosis of enchondromas localized in second finger of the right hand due to the fact that the patients had come up with recurrence in a destructive manner after seven months of follow-up. Radical resection of the chondrosarcomas localized

in the hand phalanges is the suggested treatment. Amputation of the affected finger or metacarpal is the correct treatment to avoid local recurrence or distant metastasis.[1,2,4,6,10,12,15] Phalangeal amputation was performed in our study in order to prevent high recurrence rates and potential distant metastasis for the treatment of chondrosarcomas localized to phalanges. There was a diagnosis of chondrosarcoma of proximal phalanx of a patient with Ollier disease and two patients who underwent ray amputation. Ray amputation was performed for these cases in order to prevent local or distant metastasis due to soft tissue involvement through hand region shown clinically and by MRI.

In conclusion, in this study, we accounted for 9.6% of chondroid tumors localized to phalanges and probably the rate is higher than estimated. During diagnosis; clinical examination findings and conventional radiologic studies must be evaluated well, and MRI studies must be performed to evaluate soft tissue involvement and support the diagnosis for cases that are suspected as chondrosarcoma. Nowadays, the current treatment is amputation that necessitates radical resection. Ray amputation must be performed particularly for cases with proximal phalanx or soft tissue involvement through the hand shown by MRI.

Declaration of conflicting interests

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