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Management of tumors and tumor-like lesions of the hand: a review of 191 patients

El tümörleri ve tümör benzeri lezyonların tedavisi: 191 hastanın değerlendirilmesi

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Objectives: In this study, we aimed to investigate the incidence of benign and malignant hand tumors, localization and management approaches.

Patients and methods: Between March 1986 and October 2008, 191 consecutive patients (86 males, 105 females; mean age 35.06 years; range 1 to 96 years) who were diagnosed with tumors or tumor-like lesions of the hand in our clinic were retrospectively analyzed. Patients with only intraosseous ganglion cysts were included in the study.

Results: The most common lesion was enchondroma (n=59, 30.89%). A giant-cell tumor of the tendon sheath was the most common benign soft tissue tumor (n=30, 15.70%). Alveolar rhabdomyosarcoma was the most frequent malignant soft tissue lesion (n=5, 2.61%). The most common non-metastatic malignant lesions of the bone were chondrosarcomas (n=6, 3.14%). Of the patients with chondrosarcoma, two were diagnosed with primary lesions, and four were diagnosed with chondrosarcoma when benign cartilaginous lesions underwent malignant transformation during follow-up. Metastases to the hand were seen in six patients.

Conclusion: The incidence of benign tumors of the hand is higher than malignant tumors. Since hand tumors may affect several parts of the body, diagnosis and treatment are best provided through a multidisciplinary approach.

Key words: Bone tumor; demography; hand tumor; hand; radiography; surgery.

Amaç: Bu çalışmada iyi ve kötü huylu el tümörlerinin görülme sıklığı, yerleşim yeri ve tedavi yaklaşımları araştırıldı.

Hastalar ve yöntemler: Mart 1986 - Ekim 2008 tarihleri arasında kliniğimizde el tümörü veya tümör benzeri lezyonu tanısı konulan 191 ardışık hasta (86 erkek, 105 kadın; ort. yaş 35.06 yıl; dağılım 1-96 yıl) retrospektif olarak değerlendirildi. Gangliyon kistleri yalnızca kemik içerisinde olan hastalar çalışmaya dahil edildi.

Bulgular: En sık görülen lezyon enkondrom (n=59, 30.89%) idi. Tendon kılıfının dev hücreli tümörü en sık görülen iyi huylu yumuşak doku tümörüydü (n=30, %15.70). Yumuşak dokunun kötü huylu tümörlerinden en sık görüleni, alveoler rabdomyosarkomdu (n=5, %2.61). Kemiğin en sık görülen metastatik olmayan kötü huylu tümörü kondrosarkomdu (n=6, %3.14). Kondrosarkomlu hastaların ikisi primer olarak tanılanırken, dört hastada iyi huylu kıkırdak lezyonlarının takipleri sırasında kötü huylu tümöre değişim göstermesi sonucu tespit edildi. Altı hastada ele metastaz görüldü.

Sonuç: Elin iyi huylu tümörleri, kötü huylu tümörlere kıyasla, daha sık görülmektedir. El tümörleri vücudun çeşitli bölgelerini etkileyebileceği için, tanı ve tedavide en iyi sonuç multidispliner yaklaşım ile elde edilir.

Anahtar sözcükler: Kemik tümörü; demografi; el tümörü; el; radyografi; cerrahi.

Tumors of the hand are uncommon, and malignant tumors of the hand are particularly rare. Most of the tumor types encountered in bone and soft tissue in other body regions also occur in the hand, but the frequency distributions are different. For example, malignant vascular tumors and osteosarcoma are very rarely seen in the hand.[1-3] This can make accurate diagnosis difficult. Benign and malignant

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tumors of the hand may present with similar clinical findings, e.g. pain and swelling, but because they can require very different methods of treatment, correct diagnosis is important.[3] Functional outcome is also closely related to the nature of the disease and the type of treatment used.

The major references on bone and soft-tissue tumors provide only limited information on the epidemiology of tumors in the hand.[2,5,6] The objective of this study was to retrospectively review 191 consecutive patients who had tumors or tumor-like lesions in the hand.

PATIENTS AND METHODS

We retrospectively examined the records of all patients who were diagnosed with tumors or tumor-like lesions in the hand in our institution during the period March 1986 - September 2008. Tumor-like lesions were defined as those lesions which resemble tumors but are not true neoplasms, e.g. simple bone cyst, aneurysmal bone cyst. All tumor-like lesions were benign. Patients with ganglion cyst were included only if the lesion was intraosseous. In all, the records of 191 patients (86 males, 105 females; mean age 35.06 years; range 1 to 96 years) were included for analysis.

Data recorded for each patient included demographic information, clinical presentation, radiologic findings [plain radiographs, with computed tomography (CT) and magnetic resonance imaging (MRI) when available]. For most of the patients who were treated with surgery, preoperative diagnosis was based on clinical presentation and radiologic appearance, and the diagnosis was confirmed via histopathologic examination of surgical specimens.

RESULTS

Of the 191 patients in the study, 169 (88.48%) had benign tumors or tumor-like lesions, and 22 (11.51%) had malignant tumors, including metastases to the hand. Primary malignant tumors were found in 16 patients (8.37%). Primary malignant bone tumors in the hand were the rarest among these general categories, and were seen in only three patients (1.57%). Metastatic tumors were found in six patients (3.14%). Demographics, tumor types and clinical courses are summarized in Tables 1-3.

Benign tumors and tumor-like lesions

Most of these patients presented clinically with pain and/or mass (n=150, 88.75%). Most were treated with surgery (n=146, 86.4%, Table 1). The most common benign tumor in our study, as well as being the most common tumor overall, was enchondroma. Of the patients with enchondroma, nine had multiple enchondromatosis (Ollier's disease), two had Maffucci's syndrome (enchondromatosis plus soft-tissue hemangioma), and 12 presented with pathological fractures. Of the nine patients with Ollier's disease, two developed low-grade chondrosarcoma. One of these patients underwent amputation of the distal phalanx, and the other was treated with ray amputation. One of the patients with Maffucci's syndrome developed low-grade chondrosarcoma and underwent ray amputation of the fourth and fifth fingers.

Osteoid osteoma was encountered in nine patients, all presenting with pain; in seven of these, aspirin markedly relieved the pain. Of the eight patients with glomus tumors, six presented with purple lesions under the nail. One patient with chondromyxoid fibroma later developed chondrosarcoma in the same location. Of four patients with intraosseous ganglion cyst, the lesions in two were incidental findings, noticed on radiographs taken for other reasons.

Primary malignant tumors

At initial diagnosis, 16 patients had primary malignant tumors, comprising 8.37% of all patients in this study (Table 2). Of the patients with primary malignant tumors, most were male (9/16) and the mean age of patients in this group was 49.31 (range, 17-96). All presented clinically with pain and/or mass. All were treated with surgery. In Table 2, the four patients whose benign disease transformed into malignancy are not included.

Metastatic tumors

Of the patients with metastases to the hand (n=6, 3.15% of all patients, Table 3), most were male (4/6), and the overall mean age of patients in this group was 61.7 (range, 49-82). All presented clinically with pain and/or mass. Surgical treatment was possible for only one patient in this group (Table 3).

Surgical technique

For most benign bone lesions, intralesional curettage with filling of the defect was used. In patients treated with en bloc resection, the most commonly performed tissue transfer technique was reverse radial forearm flap (n=3), with the next most common being groin flap (n=1).

DISCUSSION

Tumors are less commonly found in the hand than in other body regions. In Campanacci's series of 19,673

TABLE I

Benign tumors and tumor-like lesions of the hand

	Tumor	Number of patients	Age at diagnosis Mean	Age at diagnosis Range	Side (R/L)	Gender (M/F)	Location	Number of lesions	Surgical therapy	n	Follow-up (months) Mean	Follow-up (months) Range	Complications	n
Bone tumors														
	Enchondroma (Including Ollier and Maffucci syndrome)	55	28.6	14-66	33/26	25/30	Hamate	2	Yes	49	41.6	6-251	Recurrence	3
							Metacarpal	12					Ankylosis	1
							Phalanx	46					Refracture	1
							Trapezium	1					Metacarpal shortening	1
													Secondary chondrosarcoma	3
													Local recurrence	2
	Exostosis (Including solitary and multiple form)	20	31.2	6-73	13/11	15/5	Metacarpal	8	Yes	5	86	6-169	Sudeck atrophy	1
							Phalanx	19					Myositis ossificans	1
													MP joint stiffnes	1
	Osteoid osteoma	9	22	13-31	5/4	3/6	Phalanx		Yes		41.4	5-87	Joint stiffness	3
													Recurrence	1
	Aneurysmal bone cyst	6	24.3	4-54	4/2	3/3	Metacarpal		Yes		41.8	8-188	Recurrence	2
													Pseudoarthrosis	2
	Intraosseous ganglion cyst	4	43	32-58	0/4	2/2	Phalanx	1	Yes	1	36.5	14-48	Wrist motion stiffnes	1
							Lunatum	2						
							Trapezoideum	1						
	Giant cell tumor	3	35.7	26-49	3/1	0/3	Metacarpal	2	Yes		29.3	14-39	MP degeneration	2
							Phalanx	1						
	Brown tumor	3	56	53-64	1/3	1/2	Metacarpal	2	Yes		44.5	22-72		
							Phalanx	3						
	Simple bone cyst	2	26	8-44	1/1	1/1	Metacarpal		Yes		127	26-228	Recurrence	1
							Scaphoid							
	Chondromyxoid fibroma	2	33	23-43	2/0	0/2	Metacarpal		Yes		49	38-60	Secondary chondrosarcoma	1
							Phalanx							
	Intraosseous neurofibroma	1	23		0/1	1/0	Phalanx		Yes		14			
Soft tissue tumors														
	Giant cell tumor of the tendon sheath	30	37.9	12-64	10/20	8/22	Flexor tendons	15	Yes	28	26	11-140	Recurrence	3
							Extensor tendons	15					Joint stiffness	3
	Glomus tumor	8	37	20-58	3/5	2/6	Palm	1	Yes		28	7-144	Joint stiffness	1
							Finger	7						
	Hemangioma	5	17.6	1-38		1/4	Palm	1	Yes	3	14.5	9-20		
							Finger	4						
	Enchondroma	4	43	25-60	0/4	0/4	Finger	3	Yes		24	12-48	Recurrence	3
							Palm	1						
	Pigmentous villonodular synovitis	3	57	54-60	2/0	2/0	Wrist joint	Yes			13	10-16		
	Benign fibrous histiocytoma	2	33		0/2	1/2	Finger	Yes		30			Joint stiffness	1
	Fibroma	2	47.5	45-50	0/2	0/2	Finger	Yes			34	13-55		
	Lipoma	2	43.5	43-44	0/2	1/1	Thenar region	Yes			18	7-29		
							MP joint							
	Angiolipoma	1	32		0/1	0/1	Finger	Yes		19				
	Angiofibroma	1	38		0/1	1/0	Finger	Yes		27				
	Angiofibrolipoma	1	15		0/1	1/0	Finger	Yes		6				
	Aggressive fibromatosis	1	37		0/1	0/1	Dorsum of the hand	Yes		60			Recurrence	
	Schwannoma	1	37		1/0	1/0	Finger	Yes		12			Recurrence	
	Hemangiomatosis	1	48		0/1	1/0	2, 3, 4 fingers			96				
	Atypical fibroxanthoma	1	38		0/1	1/0	Finger	Yes		12				
	Syringocystadenoma papilliferum	1	42		1/0	0/1	Finger	Yes		14				

R/L: Right / left; MP: Metacarpophalangeal.

TABLE II

Primary malignant tumors of the hand

	Tumor	Number of patients	Age at diagnosis Mean	Age at diagnosis Range	Side (R/L)	Gender (M/F)	Location	Number of lesions	Surgical therapy	n	Follow-up (months) Mean	Follow-up (months) Range	Complications	n
Bone tumors														
	Chondrosarcoma	2	68.5	49-80	0/2	2/0	Metacarpal	2	Surgery		33.5	9-58		
	Epithelioid hemangioendothelioma	1	42		1/0	0/1	Metacarpal		Surgery		30			
Soft tissue tumors														
	Alveolar rhabdomyo-sarcoma	5	31.6	17-45	1/4	2/3	Wrist	1	Adjuvant Cht	3	53.25	29-108	Death	2
							Palm	1	RT	2			Alive with disease	1
							Finger	2	Neoadjuvant Cht	1				
							Dorsum of the hand	1	Surgery	4				
	Squamous cell carcinoma	4	74.5	52-96	2/2	3/1	Finger	4	Surgery	4	6.47	6-8	Death	1
									Adjuvant Cht	1			Death due to unrelated disease	1
									Neoadjuvant Cht	1				
	Basal cell carcinoma	1	64		1/0	1/0	Finger		Surgery	112				
	Chondrosarcoma	1	32		1/0	0/1	Finger		Neoadjuvant Cht	42				
							Surgery							
	Epithelioid sarcoma	1	34		0/1	0/1	Palm		Adjuvant Cht	48				
									Radiotherapy					
									Surgery					
	Synovial sarcoma	1	32		1/0	1/0	Palm			31			Recurrence	

Cht: Chemotherapy.

TABLE III

Metastatic tumors of the hand

	Number of patients	Age at diagnosis Mean	Age at diagnosis Range	Side (R/L)	Gender (M/F)	Location	Therapy	n	Follow-up (months) Mean	Follow-up (months) Range	Complications	n
Lung	3	52	45-62	3/0	3/0	Metacarpal phalanx	Surgery	1	6	1-15	Death	3
						Carpal bones	Cht alone	1				
Breast	2	67	63-69	0/2	0/2	Metacarpal	Cht alone	1	1.5	1-2	Death	2
						Carpal bones						
Prostate	1	82	–	1/0	1/0	Phalanx	Orchiectomy	1	6	–	Death	1

Cht: Chemotherapy.

patients with tumors in bone or soft tissue, only 4.7% of the patients had tumors or tumor-like lesions in the hand.[2] Of these patients with hand disease, benign or tumor-like lesions accounted for 89.8%, and malignant disease for 10.2%. For the patients in our study who were part of a larger series of 4,300 patients diagnosed with bone or soft tissue tumors, the proportions for hand versus other body regions, and for benign or tumor-like versus malignant, were remarkably similar to those in Campanacci's series. Among our 4,300 patients, those with tumors or tumor-like lesions in the hand accounted for 4.44% of the total, and of these, patients with malignant tumors accounted for 11.51%. Data from epidemiologic studies of bone tumors are consistent with these figures. In Dahlin's series of 11,087 patients, 194 (1.7%) had tumors in the hand.[6] These hand tumors were benign in 86.6% of the patients. Of 2,069 patients reported in the Leeds Regional Bone Tumour Registry, 80 (3.9%) were found to have tumors in the hand or wrist.[7]

In Campanacci's series of tumors in all body regions, approximately half of the patients were found to have malignant disease, and the other half benign or tumor-like conditions.[2] It is interesting that the malignant versus benign ratio was very different among patients having tumors in the hand, with approximately 90% having benign disease. We found this same ratio among our patients with tumors of the hand.

Benign and tumor-like lesions of soft tissue

Ganglion cyst, also known as mucous cyst or hygroma, is the most common non-traumatic soft-tissue lesion in the hand.[1] However, in this study we included patients with ganglion cysts only when the lesion was located within the bone; ganglion cysts elsewhere were not always noted in the patients' data forms, due to the fact that some surgeons do not consider it to be a tumor. In its shape, ganglion cyst resembles a tumor, but histologically it consists of mucoid material.[1,2] The second most common benign tumor of soft tissue in the hand is giant cell tumor of the tendon sheath, also known as pigmented villonodular tenosynovitis.[8] Biopsy is usually necessary for diagnosis of this lesion because clinically it is similar to epitheloid and synovial carcinomas.[2] Our findings are consistent with the first and second rankings of ganglion cyst and giant cell tumor of the tendon sheath; since patients with extraosseous ganglion cyst were not included in our study, it would be expected that giant cell tumor of the tendon sheath would be the most common soft tissue tumor, and this was the case.[9]

Benign and tumor-like lesions of bone

Enchondroma is the most common primary bone tumor arising in the hand, and approximately one-third of all enchondromas are encountered in the hand.[1] In the present study, 59 patients had enchondroma of the hand (four within soft tissue), out of a total of 210 patients with enchondroma in our larger series (28.09%). Given the possibility of transformation to malignancy, as occurred in four patients in our study, cartilaginous tumors should be followed closely, particularly when they are multiple. Histologically, enchondromas can be difficult to distinguish from low-grade chondrosarcomas. Clinical and radiologic features are therefore important. Signs suggestive of malignancy include changes in pain characteristics, enlargement of lesions, thinning of the cortex, destruction of previous calcification, and expansion into soft tissue.[2]

Primary malignant tumors

From a whole-body perspective, osteosarcoma is the second most common primary tumor of bone after multiple myeloma, but in the hand osteosarcoma is rare.[2,10] In Campanacci's series only one patient with osteosarcoma of the hand was encountered, and in our series of hand tumors there were no patients with osteosarcoma. Chondrosarcoma likewise shows a difference in whole-body versus hand distribution. Among primary malignant tumors of bone, chondrosarcoma is fourth in frequency overall, but is first in the hand.[1,2,11,12] Ewing's sarcoma in the hand is rare, and was not encountered in any of our patients, but should be kept in mind in the differential diagnosis of hand lesions because in its clinical presentation Ewing's sarcoma can closely resemble infection.[1] Among malignant soft-tissue tumors in the hand, the most common types are epithelioid sarcoma and synovial sarcoma.[1,13] However, in our study the most common primary malignancy in soft tissue in the hand was alveolar rhabdomyosarcoma.

Metastases

Although metastases are the most frequent malignancy of the skeleton overall, metastases to the hand are rare.[2] In Campanacci's series, patients with metastases to hand bones comprised 0.5% of patients with metastases to bones overall.[2] Among our patients, the corresponding figure was 1.4%. Metastases to the hand are similar to metastases to bones in other body regions in terms of the site of origin, the most common sites being breast, lung, prostate and kidney.[2,14,15]

Metastases from primary tumors in the hand to other body regions were not detected in any of our patients at the time of diagnosis. As with primary musculoskeletal tumors in other locations, however, metastases can occur. Bone tumors, when they metastasize, generally do so via the blood vessels, and for this reason a common destination of metastasis is lung. Soft tissue tumors, especially rhabdomyosarcoma, epithelioid sarcoma, clear cell sarcoma, and angiosarcoma, can metastasize to regional lymph nodes.[1,2]

Diagnosis and evaluation

Independent of tumor type, most of our patients with hand tumors presented clinically with pain and/or mass. Clinical findings can likewise be nonspecific in metastatic disease, and can include the general signs of inflammation (pain, swelling and erythema).[1] Metastases to the hands or feet can also resemble rheumatoid arthritis clinically.[16] Pathologic fractures can occur with benign lesions such as enchondroma, and with malignancies such as chondrosarcoma. For a few tumor types, clinical presentation can be helpful in diagnosis. Pain due to osteoid osteoma is characteristically relieved by aspirin.[17] Lesions located on tendons suggest tumors of tendinous origin, such as giant cell tumor of the tendon sheath. A purple lesion under the nail accompanied by pain with no history of trauma is suggestive of glomus tumor. Some nail abnormalities can be nonspecific, however; an appearance that suggests onychomycosis can also be a manifestation of squamous cell carcinoma of the nail apparatus.[18]

Imaging generally provides a preliminary basis for deciding whether a lesion is benign or malignant. With bone tumors in the hand, as in other body regions, sclerotic margins suggest a benign lesion, while a lack of margins or a breakdown of cortex suggests malignancy. Bony changes are more apparent on CT, and in particular any intramedullary involvement can be evaluated for preoperative planning of the extent of resection that will be needed. Soft tissue lesions can be shown in greater detail with MRI. On imaging studies, however, differences in tumor type can be subtle. For example, enchondroma generally appears as a well-demarcated lytic lesion with stippled calcification, whereas chondrosarcoma more commonly shows cortical breakdown and expansion into the soft tissue but these are only general tendencies.[1,15] Given the difference in treatment and prognosis for these two tumors, imaging studies should be complemented with other methods. Needle or surgical biopsy may be necessary in providing a precise diagnosis for treatment planning. After incisional biopsy, hemostasis is important in preventing the spread of cells beyond the biopsy site.

Treatment

For patients with benign lesions in bone, the main indication for surgery is pain. Benign lesions in soft tissue may require surgical treatment if they are locally aggressive or if they interfere with hand function. For aggressive benign bone tumors such as giant cell tumor, the use of adjuvants with curettage has been recommended.[19] However, for giant cell tumors in the finger, amputation may be necessary.[20] For benign bone lesions in general, the curettage site should be checked intraoperatively with radiographs to rule out any residual tumor. During surgery for enchondroma, frozen section may be needed intraoperatively if there is uncertainty about whether the tumor is low-grade chondrosarcoma. Initial clinical misdiagnoses of chondrosarcomas as other lesions have been described, as well as chondrosarcomas presenting with long durations of symptoms, in some cases several years.[12]

For lesions that are suspected of being malignant, histopathologic diagnosis is needed before surgery. If the lesion is found to be malignant preoperatively, then during surgery, the biopsy tract should be removed en bloc with the tumor mass, along with a margin of normal tissue surrounding the tumor. Obtaining a wide surgical margin may require amputation. For example, with tumors of the finger, when extracompartmental extension or pathologic fractures are present, removal of the finger or ray resection is indicated.[21] In metastatic disease, definitive treatment depends on finding the primary malignancy and treating the patient accordingly. Before tourniquet inflation for biopsy or surgery, limb exsanguination is not recommended due to the risk of sending tumor cells to other parts of the body.

It may be emphasized that for patients with tumors in the hand, a multidisciplinary approach is crucial for diagnosis and treatment design. Due to the hand's complex structures being located in a small space, the risk of inadequate surgery that preserves structure and function must be balanced against the risk of unnecessarily extensive surgery.

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