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Solitary enchondromas of long bones: pattern of referral and outcome

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Objectives: Enchondromas are benign cartilaginous tumors, often found incidentally and diagnosed by the radiographic appearance. Active growing enchondromas/low grade chondrosarcomas are diagnosed by clinical symptoms and possibly an aggressive appearance on the radiographs. This study aimed to answer the following questions: Who requests a referral? The radiologist reporting a possibility of sarcoma or the referring physician? What is the outcome of these patients?

Methods: We retrospectively reviewed the medical records of 115 patients with final diagnosis of enchondroma over three consecutive years and recorded the radiological diagnosis on report, patients' symptoms, our initial diagnosis, follow-up, and any decision for a biopsy/surgical management, as well as the histological final diagnosis.

Results: Nearly 80% of patients were referred from an orthopedic surgeon. About half of the imaging reports mentioned a malignancy in the differential diagnosis of enchondroma. Very few had the classic signs of an aggressive/growing cartilage tumor. In radiological evaluation, we found scalloping/cortical erosion, lytic areas, cortical breaks, soft tissue extension in only 12 cases of which 8 underwent a biopsy. Of the study patients, 65% were diagnosed with adjacent joint problems.

Conclusion: Enchondromas are mostly diagnosed incidentally. They are frequently associated with adjacent joint or soft tissue pathologies, which are main source of the symptoms. Even small, well-defined lesions are often confused with a sarcoma or other malignancies, which may be due to the lack of education on bone tumors for both the radiologists and general orthopedists.

Key words: Bone; orthopedics; radiology; solitary enchondroma.

Enchondromas involving long bones are usually asymptomatic, and are commonly recognized as an incidental finding identified either on standard radiographs for unrelated trauma or joint disease or on a radionuclide bone scan for the investigation of unrelated musculoskeletal pathologies.[1] Regional pain about an enchondroma is more frequently related to a nearby joint or a local soft-tissue disorder than to the tumor itself.[2] An asymptomatic enchondroma in a long bone does not require treatment beyond follow-up by sequential clinical assessments and radiographic evaluations to rule out progression. The morbidity of curettage or even biopsy usually outweighs any potential benefit for these indolent lesions.[3] Symptomatic enchondromas can be treated by intralesional excision without the risk of local recurrence. Active growing enchondromas/low grade chondrosarcomas are diagnosed by local symptoms

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and possibly an aggressive appearance on the radiographs. The imaging characteristics that should suggest chondrosarcoma are endosteal scalloping or cortical erosion, lytic areas, cortical breaks, periosteal reaction and soft tissue extension. In the absence of these findings, a short period of conservative therapy (rest and non-steroidal anti-inflammatory medication) will often relieve symptoms and allow pain related to chondroid lesions to be distinguished from joint-mediated pain.

In an orthopedic oncology office, a large number of patients with enchondroma are seen, and often the clinical and radiographic findings easily point to the benign nature of the lesion. As such, the purpose of our study was to understand the patterns for referral of these patients, and to present the management at our institution and the outcomes.

Patients and methods

We retrospectively reviewed charts of patients over three consecutive years, noting any mention of an enchondroma in our differential diagnosis; and reviewed the radiology report, patients symptoms, our initial diagnosis, follow-up and any decision for a biopsy/surgical management as well as the histological final diagnosis. The patients with lesions of small tubular bones (enchondromas of hands and feet) and multiple lesions (Ollier's disease or Maffucci's syndrome) were excluded from the study as they can appear more aggressive both radiographically and pathologically and create a confusing presentation. All the patients were managed by an algorithm that we used in our institution (Fig. 1). Clinical evaluation was done including patient's age, gender, site of the lesion, presentation (pain or incidental finding), and any presence of night pain. Imaging studies were assessed for suggestive features of aggressive or malignant lesion including deep endosteal scalloping, lytic changes, cortical break, soft tissue extension, and bone marrow edema. The size of the lesion alone was not considered as a sign of the malignancy since there was no relevant supporting literature. The diagnoses on initial radiographic reports or referral doctors' letters, our final diagnoses, and decision of management were recorded.

Results

A total of 115 patients were identified, with a mean age of 61 years (range 21-73 years). All of the patients were referred by another clinician, of which nearly 80% were from an orthopedic surgeon while 18% were from a primary care doctor (general practitioner). Nearly two-thirds of the lesions were located in the distal femur and proximal humerus, with lesser numbers in the other bones (Fig. 2). Of the

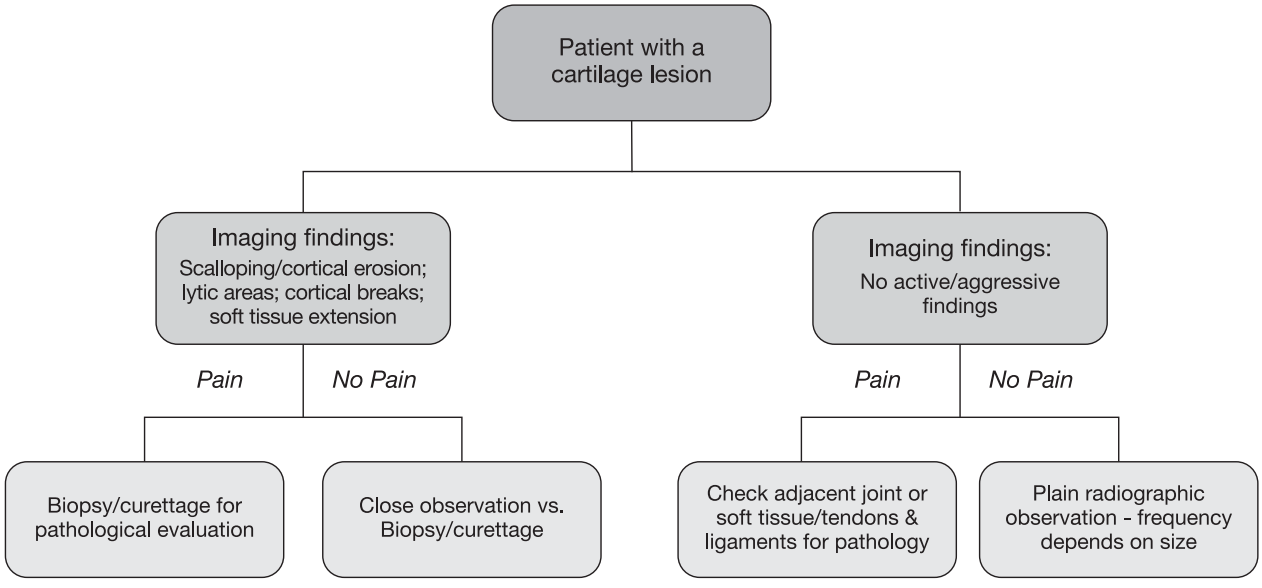

Fig. 1. The algorithm that we used for management of cartilage lesions of long bones.

patients, 76% presented with pain of the affected area and the rest were found incidentally. Only 12 patients (10.4%) complained of night pain as well, although none of these ended up with a biopsy. All of the patients had some type of imaging studies. Of these, 15% did not present with a plain radiograph, but had magnetic resonance imaging (MRI). Overall, 85% had MRI, 46% bone scan and 10% computed tomography (CT) (Table 1).

Approximately 50% of the initial radiology reports included "malignancy, neoplasm, sarcoma" either alone or in combination and sometimes also including the "possibility of an enchondroma". Adjacent arthritis and/or tendinopathy were mentioned on the initial radiology reports in 40%. Our initial radiographic evaluation showed the scalloping/cortical erosion, lytic areas, cortical breaks, soft tissue extension in 12 cases (10.4%) of which only 8 (7% of total) underwent a biopsy (Table 2). Adjacent joint arthritis or tendinopathy was diagnosed in 65% (Fig. 3). Based on the imaging studies, our diagnoses of "benign" and "enchondroma" were made on 87.8% and 95.6% of patients, respectively, with "malignancy" mentioned in 7.7% (Table 3). The final diagnosis was an enchondroma in all of our patients, including the small percentage that underwent a biopsy. Our recommendations were referral back to the primary orthopedic surgeon in nearly 50% of the cases for further care of the arthritis and/or tendinopathy. A recommendation of follow-up with plain radiographs was made for 93% either by our team or by the referring doctor.

Table 1
Characteristics of patients at referral

	Number	%
Referred by		
Orthopedic surgeon	92	80.00
Primary care doctor	21	18.26
Other	2	1.73
Site		
Distal femur	45	39.13
Proximal humerus	42	36.52
Proximal tibia	11	9.56
Proximal femur	9	7.82
Proximal fibula	8	6.95
Presentation		
Pain	88	76.52
Incidental	27	23.48
Night pain		
No	103	89.56
Yes	12	10.44
Radiography		
No	18	15.65
Yes	97	84.34
Bone scan		
No	62	53.91
Yes	53	46.09
Computed tomography		
No	103	89.56
Yes	12	10.44
Magnetic resonance imaging		
No	17	14.78
Yes	98	85.22
Total	115	100.00

Discussion

Enchondromas are benign cartilaginous tumors, often found incidentally and diagnosed by the radiographic appearance. It is the second most common benign bone tumor and represents nearly 20% of all cartilage-forming tumors. It is seen either as a monostotic or polyostotic form (enchondromatosis, Ollier's disease and Maffucci's syndrome). The risk of malignant transformation is likely 25% or higher for enchondromatosis, however, solitary enchondromas very rarely lead to malignant change (less than 1%). Solitary enchondroma is typically first discovered between the second and or fourth decade. It is commonly located centrally within the marrow of the bone, with a predilection for short tubular bones, followed by the proximal humerus and femur. Most enchondromas are stage 1 or 2 (latent or active) lesions.[4]

Unlike the intramedullary chondrosarcomas, the enchondromas are asymptomatic tumors in majority of the cases. They are generally recognized during work-up for musculoskeletal problems unrelated to the tumor, and often are due to pain from the adjacent joint or tendons.[5] Levy et al.[6] showed that in patients with enchondromas of the proximal

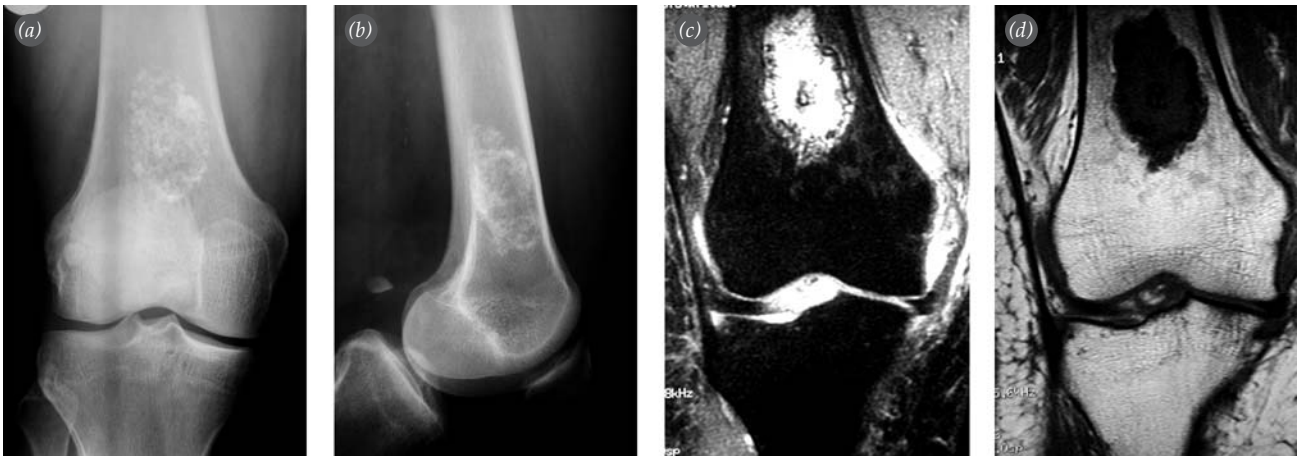

Fig. 2. Imaging studies of a 43-year-old woman with knee pain demonstrate typical characteristics of incidentally diagnosed enchondroma in the distal femur. **(a, b)** Anteroposterior and lateral radiographs of distal femur show a central lesion with calcified matrix. **(c)** Coronal T1-weighted MRI demostrates predominantly low signal intesity lesion with lobulated margins. **(d)** On a coronal T2-weighted image, the lesion displays mostly high signal areas (cartilage tissue) with some dark areas (calcifications).

humerus, MRI demonstrated that shoulder related problem (e.g. impingement, cuff problems, tendinitis) was the source of the pain in 82% of the patients. The most worrisome symptoms, which are suggestive for malignancy, are night pain and pain at rest.[2,5] In our series, nearly two-thirds of the patients presented with pain and only 10% of them had night pain. In almost 65% of our patients, an association with a joint or tendon related disease was noted.

Of our patients, 15% did not present with plain radiographs, but had MRI. Despite the sophisticated imaging methods, plain radiographs remain the initial examination of choice in the diagnosis of cartilaginous tumors in bones. Bone scans, CT and MRI are usually not necessary for the evaluation of well-calcified lesions.[2] Plain radiographs usually demonstrate a small (<5 cm) cartilaginous lesion with intramedullary calcifications without cortical involvement or soft-tissue extension.[2] These mineralizations form small punctate or short ring-like or arc-like densities. The cortex can show the scalloping to focal thinning due to cartilage nodules that border on the endosteal cortex.[1,7] Although both enchondromas and low-grade chondrosarcomas may have endosteal scalloping, the extent and degree of scalloping correlate with the likelihood of the lesion being a chondrosarcoma.[2,5] CT, MRI and bone scan are helpful when there is a suspicion for malignancy. Murphey et al.[5] studied the radiological parameters to allow distinction of appendicular enchondroma and chondrosarcoma. Of the parameters which were investigated, deep endosteal scalloping (greater than two-thirds of cortical thickness), cortical destruction and soft-tissue mass (on CT or MRI), periosteal reaction (on radiograph), and marked uptake of radionuclide (greater

Table 2
Radiographic findings of imaging studies

	Number	%
Scalloping		
No	112	97.39
Yes	3	2.61
Lytic		
No	110	95.65
Yes	5	4.35
Cortical break		
No	112	97.39
Yes	3	2.61
Soft tissue extension		
No	115	100.00
Yes	0	0.00
Edema		
No	114	99.13
Yes	1	0.87

than the anterior iliac crest) on bone scintigraphy reached statistically significance. None of these radiological parameters existed in 90% of our patients. Very few had the classic signs of an aggressive/growing cartilage tumor. However, it was noted that even small and well-defined lesions were often confused for a sarcoma or other malignancies.

Even if there were no clear joint or tendon diseases found in proximity to the "enchondromas", the radiographic findings were benign appearing, and were followed clinically and radiologically, and all remained benign. As such, the initial symptoms may be confusing, but warrant a close follow-up. Again, our results showed that in the end, only a small percentage (7%) truly ended up with a biopsy, and even those ended up as an enchondroma. The others were followed, and the clinical picture improved with stable radiographic findings.

When a patient presents with a cartilage lesion and clinical and radiological features are typical for diagnosis of an enchondroma, a biopsy is not necessary.[8] Only eight cases of our patients underwent biopsy. The final diagnosis was enchondroma in all. A recent study showed that reliability of histopathological and radiological grading cartilaginous lesions in long bone was low even among radiologists and pathologists who are specialized and experienced in musculoskeletal tumors.[2] Thus, clinics of the patients should be considered to avoid diagnostic and therapeutic mistakes.

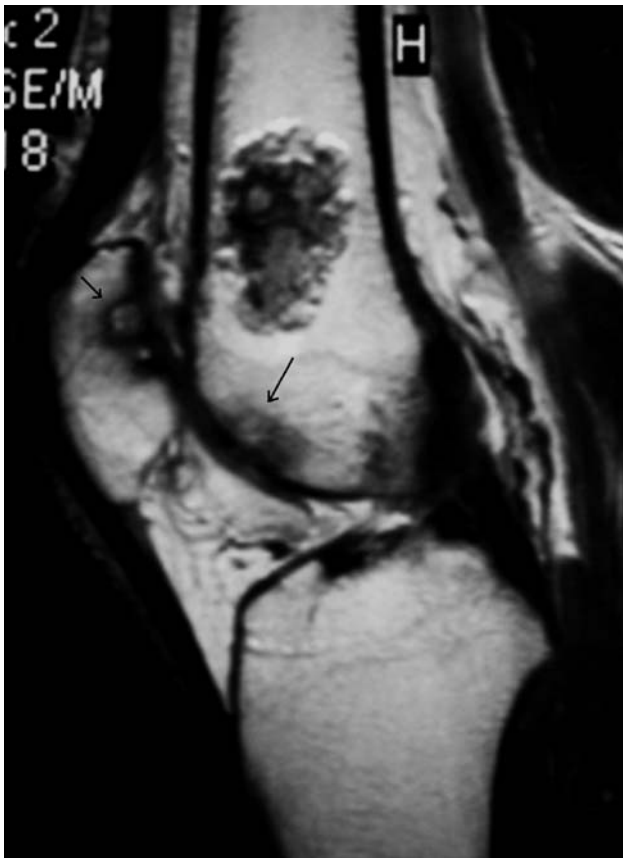

Fig. 3. Enchondroma of metaphysis of distal femur in a 56-year-old man with anterior knee pain. Sagittal T1-weighted MRI shows well-demarcated, hypointense, lobulated lesion without peritumoral edema, cortical destruction or endosteal scalloping. A grade IV chondromalacia appears on both sides of the patellofemoral joint and as the main reason for symptoms (arrows).

Table 3

Diagnoses of initial radiological reports and our review based on imaging studies

	Diagnoses on initial radiological reports		Our diagnoses based on radiological review	
	n	%	n	%
Benign	92	80.00	101	87.82
Enchondroma	102	88.69	111	95.65
Malignant or sarcoma	50	43.47	9	7.73
Joint osteoarthritis	28	24.34	30	26.08
Tendinitis	18	15.65	30	26.08
Infarct	27	23.47	4	3.48
Other	9	7.82	4	3.48

Nearly 80% of our patients were referred by an orthopedic surgeon, while 18% were from a primary medical doctor. About half of the imaging reports mentioned a malignancy in the differential diagnosis of enchondroma, encouraging referral to a specialist. This may be due to the lack of education on bone tumors for both the radiologists and general orthopedists.

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