Secondary chondrosarcoma arising from a solitary enchondroma at the index finger of the right hand: A case report

Sağ el işaret parmak yerleşimli soliter enkondromdan gelişen sekonder kondrosarkom: Olgu sunumu

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ABSTRACT

Enchondromas are benign cartilaginous tumors and tend to present in small bones of hands and feet. Malignant transformation, though rare, is seen more often in multiple rather than solitary enchondromatous. A 70year-old woman presented with swelling and pain lasting for years at the index finger of the right hand. Excisional curettage material of the mass revealed to be entirely an enchondroma. Seven months later, the lesion recurred at the same localization and amputation of the finger was performed. This time the tumor was found to be transformed into an intermediate grade (grade II) which demonstrated chondrosarcoma intermingled with classical enchondromatous areas. Enchondromas arising at the hands and feet very rarely transform into chondrosarcomas. Since enchondromas at this site often show histological and clinical features suggestive of malignancy, it is often is difficult to make a histological distinction between benign cartilaginous tumors and chondrosarcomas. Detection of radiological as well as pathological findings are essential for differential diagnosis. This case is remarkable for she has a solitary enchondroma located at the index finger of the right hand that transformed into chondrosarcoma in seven months.

Key words: Enchondroma, chondrosarcoma, hand, phalanx

ÖZET

Benign kıkırdak tümörleri olan enkondromlar, el ve ayakların küçük kemiklerinde yerleşme eğilimindedirler. Soliter lezyonlarda çok ender olan malign transformasyon, multiple endokondromatozislerde soliter lezyonlara göre daha sık görülmektedir.

Sağ el işaret parmağında yıllardır süren şişlik ve ağrı yakınması olan 70 yaşında kadın hastada kitlenin eksizyonel küretaj materyali tümüyle enkondrom olarak değerlendirilmiştir. Yedi ay sonra aynı yerde nüks eden lezyon nedeniyle uygulanan parmak amputasyonunda tümörün klasik enkondrom alanları da barındıran intermediate grade (Derece II) kondrosarkoma transforme olduğu saptanmıştır.

El ve ayaklarda yer alan enkondromlar nadiren kondrosarkoma ilerlerler. Bu yerleşim yerlerindeki enkondromlar maligniteyi düşündürecek histolojik ve klinik özellikler taşıdıkları için, benign kıkırdak tümörleriyle kondrosarkomlar arasında histolojik ayrım zordur. Bu nedenle patolojik bulgular kadar radyolojik özellikler de ayırıcı tanıda önemlidir. Bu olgu, yedi ay içinde kondrosarkoma ilerleyen sağ el işaret parmak yerleşimli enkondrom nedeniyle dikkat çekicidir.

Anahtar sözeükler: Enkondrom, kondrosarkom, el, falanks

INTRODUCTION

Enchondromas are the most common pri-

mary benign bone tumors of the hand and rarely transform into chondrosarcomas. This transformation is seen more often in multiple enchondromatosis than in solitary lesions, but even this event rarely occurs in the hands (1-13). Since enchondromas at this site often show histologi-

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Figure 1. Lesion showing punctate amorphous calcifications with apparently absent soft tissue mass or cortical destruction at the index finger of the right hand.

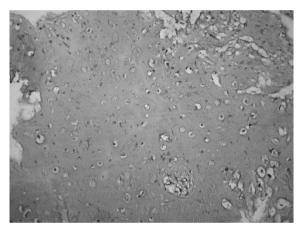


Figure 2. Enchondroma in excisional curettage material (HE x100).



Figure 3. Lesion showing bone expansion, cortical destruction, punctate amorphous calcifications and soft tissue extension at the index finger of the right hand.

cal and clinical features suggestive of malignancy, it is often difficult to make a histological distinction between benign cartilaginous tumors and chondrosarcomas. Radiological as well as pathological findings are essential for differential diagnosis (2,4,6,11-13). This distinction is very important because chondrosarcomas of the hand require wide excision to avoid local recurrence or metastasis (3,5,12-15).

We report a case of such transformation involving index finger of the right hand recurred within seven months.

CASE REPORT

A 70-year-old woman presented with swelling and pain lasting for years at the index finger of the right hand. Radiographic features and histological findings of the mass revealed to be entirely an enchondroma (Figure 1,2). Seven months later, the lesion recurred at the same localization. Radiologically the lesion showed expansion of bone, cortical destruction, punctate amorphous calcifications and soft-tissue extension (Figure 3). The finger was amputated with a clear surgical margin. Grossly, a gray-white, cartilaginous lesion measuring 4x4,5 cm was seen on the cut surface with haemorrhagic and necrotic areas, as well as bone, joint destruction and soft tissue invasion (Figure 4). Microscopic findings revealed a lobulated, destructive, cellular cartilaginous tumor with large myxoid areas, necrotic foci and invasion of cortical bone and soft tissue (Figure 5). Permeation with entrap-



Figure 4. Cut surface of amputation material showing a graywhite, cartilaginous, destructive tumor.



Figure 5. Invasion of cortical bone and soft tissue (HE x100).

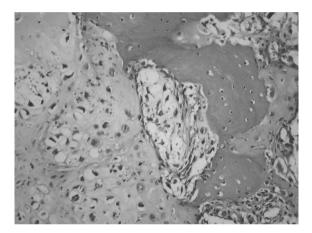


Figure 6. Permeation areas within the tumor (HE x200).

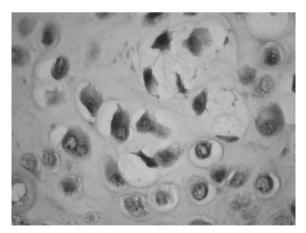


Figure 7. Atypical tumoral cells (HE x400).

ment of bony trabeculae was seen (Figure 6). In a single area, a classical enchondromatous focus was noticed. There is no histologic finding suggesting a pathological fracture. Tumor cells were hyperchromatic, pleomorphic and occasionally binucleated (Figure 7). Mitotic index was 3/10 HPA. After examination of amputation material, slides of excisional curettage material and radiological images of the primary mass were re-evaluated and compared with those of the secondary lesion. With these findings, the diagnosis of intermediate grade (grade II) chondrosarcoma secondary to solitary enchondroma was made.

No other kind of therapy was applied and at five months after surgery the patient is still free from recurrence or metastasis.

DISCUSSION

Enchondromas are benign cartilaginous tumors and tend to present in small bones of the hands and feet (3-5). Our patient had a painful swelling for years at the index finger of the right hand. Histological examination of the excisional curettage material of the mass revealed an obvious enchondroma without any malignant neoplastic features.

Enchondromas rarely transform into chondrosarcomas (1,3,4,6-13). This transformation is seen more often in multiple than solitary enchondromatous lesions but even this event rarely occurs in the hands (9,10,12-13). This case is remarkable because it was a solitary enchondroma located at the index finger of the right hand. Seven months later, amputation material was diagnosed as intermediate grade (grade II) chondrosarcoma, intermingled with a single classical enchondromatous area identical to the areas in the former curettage material. This finding suggested that this chondrosarcoma is secondary to the former solitary enchondroma.

Distinction of enchondroma and chondrosarcoma affecting the appendicular skeleton is a frequent diagnostic dilemma. Similar clinical findings are seen, particularly painful sensations related to the lesion. But radiological findings can help to make differential diagnosis between enchondroma and chondrosarcoma.

Radiological features such as cortical destruction, periosteal reaction and soft tissue mass strongly suggest the diagnosis of chondrosarcoma (2,4,6,11-13).

Histologically, enchondromas of small bones of hands and feet may be hypercellular, with double-nucleated cells and myxoid changes in the matrix, but permeation with entrapment of bony trabeculae is the most important sign of malignancy. Destruction of cortex and invasion of soft tissue are seen in chondrosarcomas (1,12,13). These malignant neoplastic features were seen in our patient's tumor.

Proximal phalanx is the most common site for chondrosarcoma of the short tubular bones of hands and feet. The majority of chondrosarcomas are of high histologic grade (grade 2-3) with extensive myxoid areas (12-15). The less aggressive behaviour and low risk of metastases despite high histologic grade indicates that chondrosarcomas of the hand behave differently from chondrosarcomas found elsewhere (12-15). However, chondrosarcoma of the hand requires a prompt and more radical treatment than enchondroma. Wide excision is recommended to avoid local recurrence or metastasis (3,5,12-15). Five months after finger amputation, our patient has still no signs of recurrence or metastasis.

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