

## **Malignant transformation of Ollier's disease in the hand.**

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### *Background*

Enchondroma is the most common primary benign skeletal neoplasm of the hand, predominantly locates in the phalanges. Multiple enchondromatosis, so called Ollier's disease, is rare condition. It was defined by the presence of multiple enchondromas and characterized by an asymmetric distribution of cartilage lesions that can be extremely variable. Multiple enchondromatosis can sometimes transform into chondrosarcoma, but this event is extremely rare in the hand. We present here two cases with chondrosarcomas of fingers associated with Ollier's disease. The duration from first detection of the disease to newly growing tumor was 60 years. Our cases had the longest disease progression and tumor size was surprisingly huge.

### *Case 1*

Case 1 is a 78-year-old man. He noticed a small mass at the finger and underwent tumor resection at the age of 18. In the last 1 years, the tumor acutely enlarged in size and seriously deformed appearance with ring and little fingers. Plain radiographs showed an extra-osseous tumor shadow with multiple calcifications. The little finger had a recalcitrant skin ulcer at the tip and infected with *pseudomonas aeruginosa*. There was neither spontaneous pain nor tenderness that was the reason why he delayed consultation to the clinic. We performed preoperative open biopsy, strongly suspecting chondrosarcoma grade 1-2. Wide resection with ray amputation of both fingers was performed. In the final pathology report, the tumor showed focally benign enchondroma, suggestive of malignant transformation from Ollier's disease to chondrosarcoma. Local recurrence or metastasis weren't found at 3 year follow-up.

### *Case 2*

Case 2 is a 71-year-old woman. She had noticed elevated lesion of her index and middle fingers from a child, and was performed operation at the age of 18. Tumor size

gradually increased in the last few years and recently those had a recalcitrant skin ulcer with infection. She has multiple bone neoplasms with calcification in the bones of the left side of the body, and those are probably enchondromas. No distant metastasis was found in the visceral organ. The index and middle ray amputation with reconstruction of local flap is planned in the near future.

### *Discussion*

The prevalence of Ollier's disease is estimated at 1/100,000. The rate of malignant transformation of Ollier's lesion has been estimated at 20-50%. The most frequent transformation is to chondrosarcoma and this occurs in about 25% of patients by the age of 40 years. Chondrosarcoma of the hand secondary to multiple enchondromatosis appears to be extremely rare. (Table 1) The signs of malignant transformation of a benign enchondroma are increase in size of the tumor, onset of pain or tenderness, and cortical destruction associated with soft tissue invasion. However, our case showed no pain and tenderness, although the lesion was enlarged. The diagnosis of chondrosarcoma was delayed for this reason. Further, those patients who lives in the countryside, they has been living in hiding their hands. Because chondrosarcoma is resistant to the radiotherapy and chemotherapy, surgical resection is the only treatment. Poor outcome for chondrosarcoma of the hand has been associated with either inadequate resection followed by local relapse, or delayed surgical treatment. Fortunately, our case had grade 1 chondrosarcoma and distant metastasis had not occurred up to the final follow-up of 3 year. Most secondary chondrosarcoma that develop in patients with Ollier's disease represent low-grade malignancies with a favorable prognosis following adequate wide resection.

**Figure 1. Macroscopic findings of case 1 (left) and 2 (right)**



**Table 1. Reported cases of chondrosarcoma of the hand secondary to enchondromatosis**

Authors	Sex/ age	Site	Disease progression	Pain	Grade	Treatment	FU
Block	25/ M	Ring&little	7y	No	Low	Ray A.	7m NR
Palmieri	73/M	Thumb	Not shown	Yes	Unknown	Excision	8y NR
	81/F	Index	Not shown	Yes	Unknown	Ray A.	2y NR
	62/M	Ring	Not shown	No	Unknown	Ray A.	4y NR
	59/M	Index	Not shown	Yes	Unknown	Rat A.	3y NR
Goto	27/ M	Little F	10y	No	II	Ray A.	6m NR
	76/ M	Ring F	2y	No	I	Amputation	1y NR
Altay	24/ M	unknown	10y	unknown	unknown	Ray A.	22 NR
Martinez	53/ F	Ring&little	28y	No	unknown	Ray A.	12m NR
Our cases	78/M	Ring&little	60y	No	I	Ray A.	1y NR
	71/F	Index&Mid	60y	No	I		

Ray A.: Ray amputation, NR: No recurrence