Volume 21 Number 2
February 2015

Photo vignette

Digitial endochondroma

CA Wilson[1], RH El-Khayet[1], J Somerville[2], K McKenna[1]

Dermatology Online Journal 21 (2): 15

[1]Departments of Dermatology, Belfast City Hospital, Belfast, County Antrim, UK.

[2]Departments of Pathology, Belfast City Hospital, Belfast, County Antrim, UK.

Correspondence:

Dr Rami El-Khayat Department of Dermatology
Belfast City Hospital
51 Lisburn Rd
Belfast, County Antrim BT9 7AB, UK
E-mail: ramihkh@yahoo.com

Abstract

A patient with Ollier disease presenting with onycholysis and nail dystrophy related to a subungual enchondroma is presented.

Case synopsis

We report a 27-year-old man with a known history of Ollier disease who was referred for a dermatology consultation because of a four year history of a gradually enlarging swelling below the left middle fingernail. Apart from multiple fractures during childhood, he had no other past medical history of note.

On examination, the patient had a 1.3cm by 1cm subungual nodule resulting in significant onycholysis and nail dystrophy (Figure. 1a). Furthermore, multiple firm subcutaneous nodules were noted on his third and fourth digits (Figure.1b). The differential diagnoses included bony, cartilanginous and soft tissue tumors and subungual melanoma. Given the background of Ollier disease, an X-ray of the hand was obtained. This demonstrated multiple lucencies within the proximal and middle phalanges of the left middle and ring fingers in keeping with enchondromas. A further enchondroma was seen in the head of the third metacarpal (Figure.2). A referral was subsequently made to the orthopedic team. The nodule located on the distal phalynx of the middle finger was excised, with nail bed reconstruction. Histological examination showed lobules of moderately cellular cartilaginous tissue in keeping with chondroma (Figure.3).

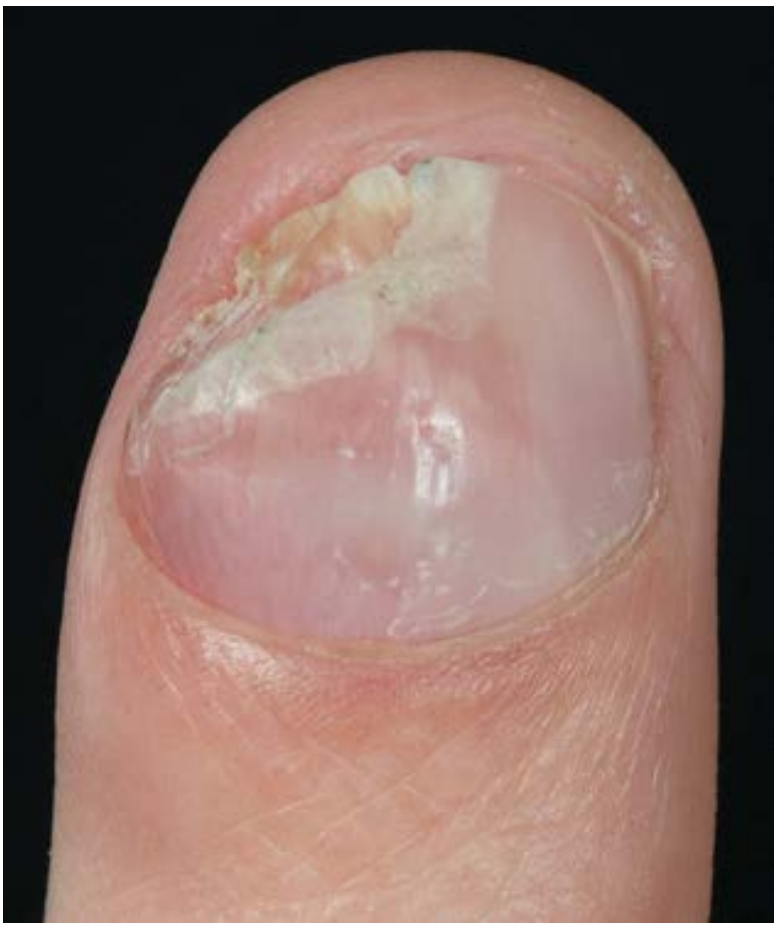
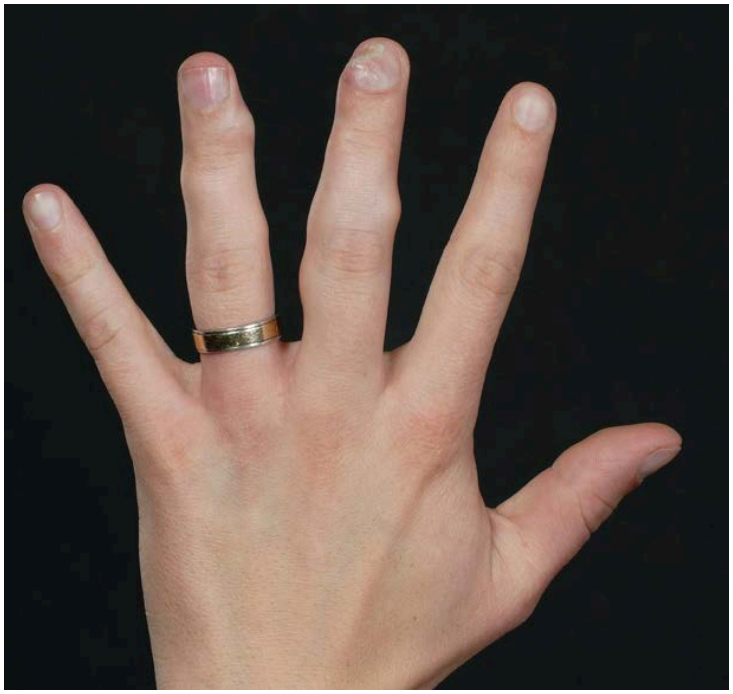


Figure 1A. Subungual lesion with significant onycholysis and nail dystrophy.



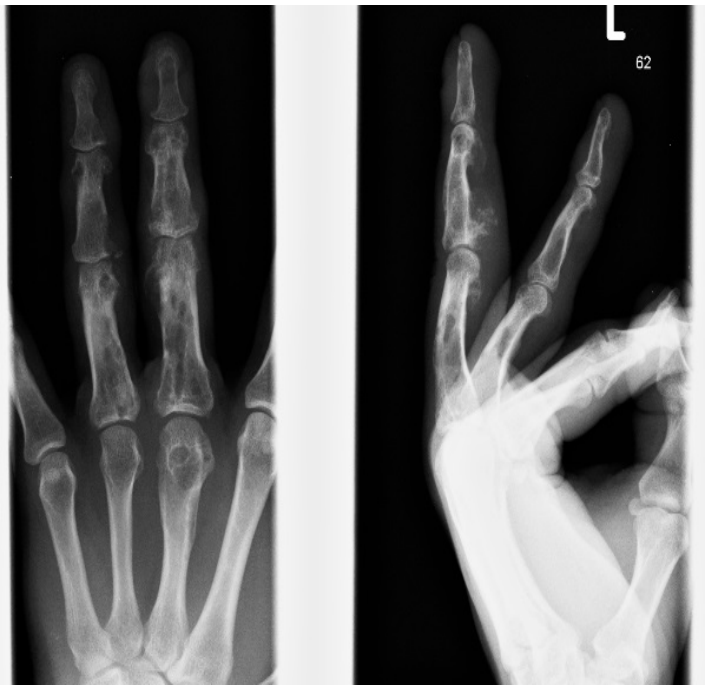


Figure 1B. Multiple firm subcutaneous nodules were noted on his third and fourth digits. **Figure 2.** Multiple lucencies within the proximal and middle phalanges of the left middle and ring fingers. On the lateral view, a further lucency was noted in the head of the third metacarpal.

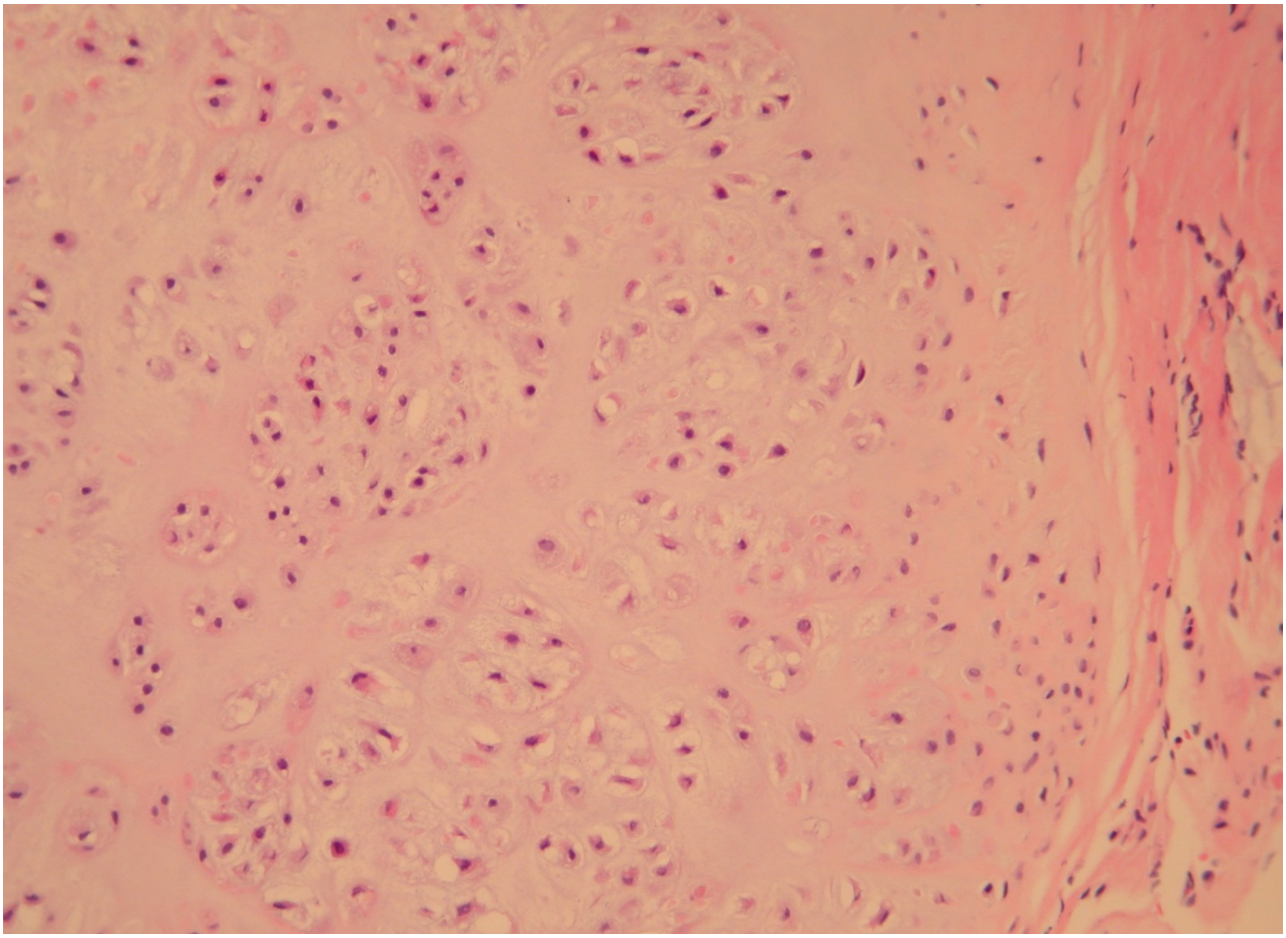


Figure 3. Lobules of moderately cellular cartilaginous tissue (hematoxylin-eosin, original magnification ×100)

In addition, the lateral view of the left middle finger demonstrated enchondromas arising from the volar aspect of the middle phalynx. The patient is currently under regular follow-up by the orthopedic team regarding this.

Discussion

Ollier disease is a rare non-familial condition resulting in multiple enchondromas (benign, intramedullary neoplasms of hyaline cartilage) and dysplastic cartilage, which usually develop in childhood [1]. The condition has an estimated prevalence of 1 in 100,000 [2]. This condition usually affects the small bones of the hands and feet, the long tubular bones, and flat bones

such as the pelvis [1,2]. The cause of Ollier disease is as yet unknown [3]. It is believed that this condition usually stops spontaneously with skeletal maturity. Therefore, any lesion showing activity after termination of the growth period requires thorough examination to evaluate for malignant change [4].

Classical manifestations of Ollier disease include pathological fractures, leading to the discovery of enchondromas on radiological imaging, as well as deforming subcutaneous nodules of the digits [5]. Our patient presented to our department with onycholysis and nail dystrophy associated with a subungual mass resulting from his underlying Ollier disease. This is a manifestation that has been rarely reported.

Ollier's disease has distinct similarities to Maffuci syndrome without the cutaneous vascular malformations seen in the latter. The risk of sarcomatous transformation in Ollier disease compared to Maffuci syndrome is much smaller, 25-30% compared to 100%, at long-term follow-up. [1,6]. Given this, a prompt referral should be considered in cases of known Ollier's disease presenting with an enlarging subcutaneous nodule.[5]

Conclusion

Ollier disease is a rare condition, characterized by the development of enchondromas, predominantly in the hands and feet. When enchondromas develop subungually, they can cause onycholysis, as in our case. Malignant transformation should be considered in any rapidly growing subcutaneous nodule in patients with Ollier disease.

Key Learning Points Ollier disease is a rare non-familial condition resulting in multiple enchondromas, usually affecting the hands, feet, long tubular bones, and flat bones such as the pelvis.

Like its counterpart Maffuci Syndrome, it has the potential for malignant transformation. Therefore, any new changes in an affected individual should prompt further investigation.

Patients with Ollier disease typically present with subcutaneous nodules and pathological fractures. If these nodules develop subungually, they can cause onycholysis.

References

1. D'Angelo L, Massimi L, Narducci A, DiRocco C. Ollier disease. Childs Nerv Syst 2009; 25:647-653. [PMID: 19326125]
2. Pannier S, Legeai-Mallet L. Hereditary multiple exostoses and enchondromatosis. Best Pract Res Clin Rheumatol 2008; 22:45-54. [PMID: 18328980]
3. Pansuriya TC, Kroon HM, Bovee JVMG. Enchondromatosis: insights on the different subtypes. Int J Clin Ex Path 2010; 3(6): 557-569. [PMID: 20661403]
4. Schwartz HS, Zimmerman NB, Simon MA et al. The malignant potential of enchondromatosis. J Bone Joint Surgery 1987; 69:269. [PMID: 3805090]
5. Casal D, Mavioso C, Mendes MM, Mouzinho MM. Hand involvement in Ollier Disease and Maffuci Syndrome: a case series. Acta Reumatol Port 2010; 35(3): 375-378. [PMID: 20975644]
6. Zwennecke Flach H, Ginai AZ, Wolter Oosterhuis J. Best cases from the AFIP. Maffuci syndrome: radiologic and pathologic findings. Armed Forces Institutes of Pathology. Radiographics 2001; 21:1311-1316. [PMID: 11553836]