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Fortuitous Discovery of a Multifocal Enchondromatosis in a Case at the "Luxembourg" Hospital Center in Bamako

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Abstract

The aim of this study was to clarify the interest of standard radiography in the management of Ollier's disease. **Observation**: Enchondromas are benign lesions that may present on imaging with nonspecific features in children, and they are relatively large lesions, with frequent endosteal erosion and rare matrix mineralization. We report a case of predominantly right-sided multifocal enchondromatosis in a 5-year-old girl with no known medical history, diagnosed fortuitously on standard radiography during a trauma assessment and confirmed by histology. No sign of pain was noted after a 6-month follow-up. But the radiographic control noted signs of diffuse osteoarthritic remodeling in the surgical areas and early fusion of the growth cartilages. **Conclusion**: Ollier's disease is rare, you have to know how to think about it in the face of fortuitous discoveries, especially at an early age. Enchondromas are benign lesions that may present on imaging with nonspecific features in children. In all cases, standard radiography is essential in the diagnosis and follow-up of Ollier's disease.

Keywords

Enchondromatosis, Fortuitous, Girl, Standard Radiography

1. Introduction

Ollier's disease or enchondromatosis is a rare bone disorder characterized by the

presence of multiple enchondromas, generally of asymmetrical distribution and confined to the peri-metaphyseal skeleton, sparing the skull [1].

This disease is one of the rare diseases, its prevalence is estimated at 1/100,000 in France [2].

It develops intra-osseously near the growth cartilage, it mainly affects the extremities, in particular the phalanges and the metacarpus, but can affect several sites. These can be of different number, size and location, explaining the extreme clinical variability between carriers of this disease.

The damage is usually predominant on one side. This disease is not transmissible and is not a hereditary disease: it would seem that mosaic somatic mutations are the origin [3].

Enchondromas are benign lesions that may present on imaging with nonspecific features in children, and they are relatively large lesions, with frequent endosteal erosion and rare matrix mineralization [2].

In countries south of the Sahara, to our knowledge very few cases are reported in the literature, hence the interest of this study.

We report the case of a 5-year-old girl with multifocal Ollier disease discovered fortuitously during a hand X-ray for sports trauma.

The objective of this study was to clarify the interest of standard radiography in the management of Ollier's disease.

2. Observation

5-year-old patient, with no known medical history on the side of her biological parents. The onset of the disease would be brutal, the children were playing ball at school when suddenly after a stroller from one of her classmates fell on her hands that she felt a sharp pain in them. She consulted the pediatric surgery department of the "Luxembourg" hospital center for treatment.

On clinical examination, we noted a girl in good general condition, the conjunctivae and the integuments well colored, afebrile. On inspection and palpation: there was no skin scar, there was painful swelling of the phalanges of both hands, predominantly on the right, with no other associated clinical signs (**Figure 1**).

The biological assessment, particularly inflammatory and phosphocalcic, was normal.

The standard X-ray objectified with regard to the metacarpals, the metatarsals, the proximal and middle phalanges of the two hands, the two feet, the distal end of the radius and the right tibia multiple lacunar images centromedullary metaphysoepiphyseal with well-defined clear contours limited with cortical notches and calcifications in places (**Figures 2-4**). The pelvic and skull bones were normal in appearance.

Following this examination, the diagnostic hypothesis of enchondromatosis of the proximal and middle phalanges of both hands, both feet and the distal end of the right tibia was retained.

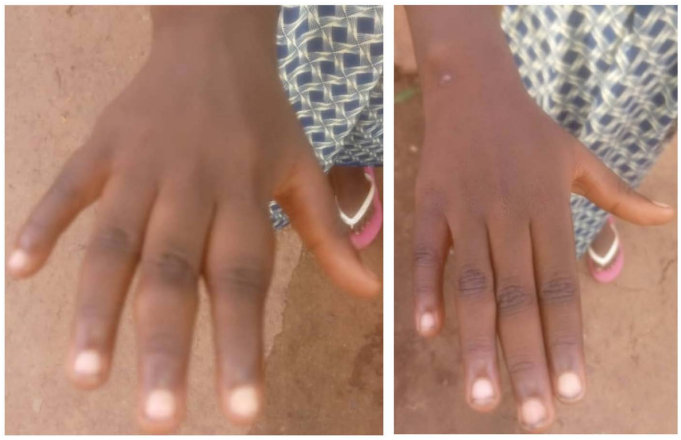

Figure 1. Photo of the hands of the 5-year-old girl after the trauma.

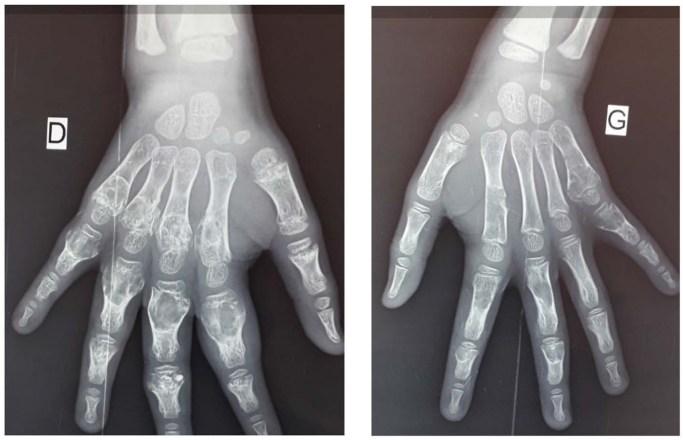

Figure 2. AP X-ray image of both hands showing, opposite the metacarpals, the proximal and middle phalanges of both hands, both feet, multiple centromedullary lacunar images with clear content, well with cortical notches and microcalcifications in places.

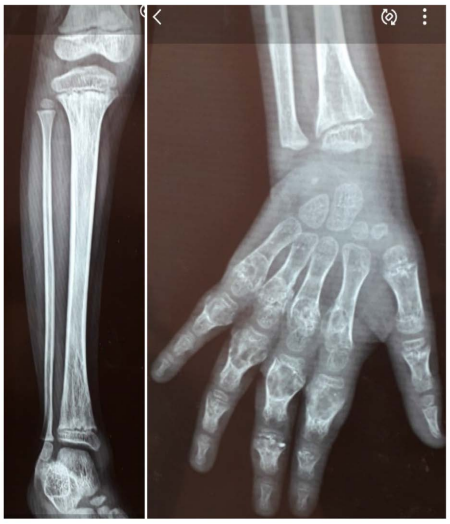

Figure 3. Frontal X-ray image of the right leg and right hand showing at the level of the distal end of the tibia a fibrillar and clear appearance of the tibia and centromedullary lacunar image with well-limited clear content of the base of the metacarpals and phalanges.

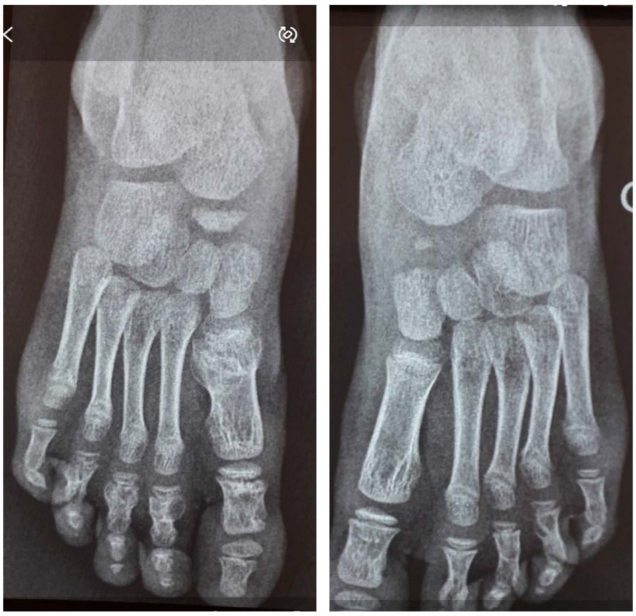

Figure 4. AP X-ray image of both feet showing, opposite the metatarsals, proximal and middle phalanges of both feet, multiple centromedullary lytic images with clear content, well limited and predominantly on the right.

Faced with the functional impairment, a surgical treatment was proposed for the right hand with the type of biopsy and curettage of the bone lesions of the first phalanges of the 3rd and 4th finger of the right hand with washing of the cavities and suturing plane by plane. No sarcomatous transformation was noted.

The anatomopathological analysis of the excision specimen confirmed the diagnosis of enchondromatosis already suspected on the radiological picture and revealed the absence of sarcomatous transformation.

No specific treatment was carried out, the patient will be subjected to treatment based on Aspégic 250 mg to calm the pain and post-operative inflammation.

Post-operative follow-up was good with first-line wound healing and disappearance of pain (**Figure 5**).

No sign of pain was noted after a 6-month follow-up. But the radiographic control noted signs of diffuse osteoarthritic remodeling in the surgical areas and early fusion of the growth cartilages (**Figure 6**).

3. Discussion

Enchondromatosis is one of the rare diseases, its prevalence is estimated at 1/100,000 in France [2]. Over 5 years of experience in radiology, this is the only case that we have been able to detect so far in the service.

This disease is not transmissible and is not a hereditary disease: it would seem that mosaic somatic mutations are the origin [3] and these attacks are generally predominant on one side. This agrees with our case, which presented multifocal lesions but predominantly on the right, especially in the right hand, and no member of his family was a carrier.

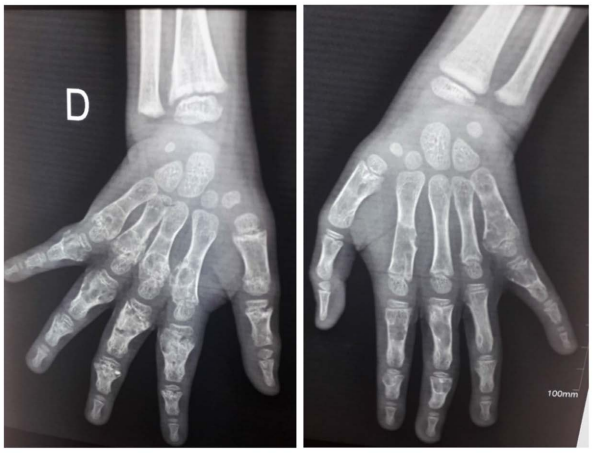

Figure 5. AP radiographic image of the hands after surgery showing a remodeled appearance of the metacarpals, phalanges and phalanges of both hands with clear centromedullary areas, cortical notches and calcification on the phalanx of the right 2nd ray.

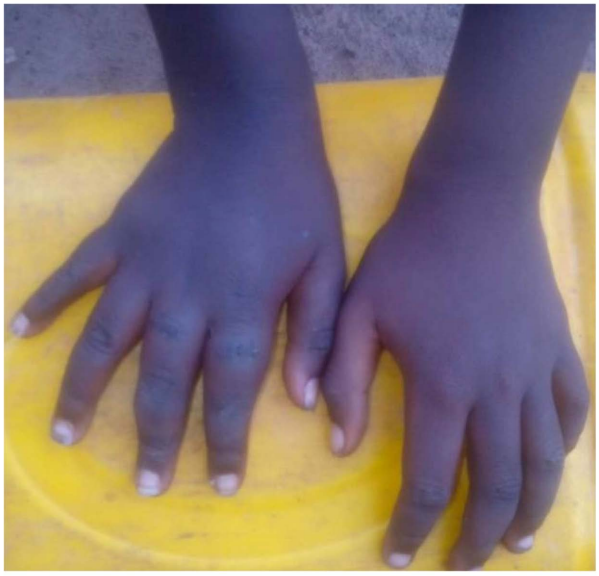

Figure 6. Photo of the hands after surgery showing scars on the phalanges of the 2nd and 3rd ray of the right hand.

These tumors are most often asymptomatic and painless, the discovery of which is often due to more or less significant deformities [4]. In our case, the discovery was fortuitous following the sports trauma.

Enchondromas are benign lesions that may present on imaging with nonspecific features in children, and they are relatively large lesions, with frequent endosteal erosion and rare matrix mineralization [2].

Most chondromas are located in the center of the bony part and are thus referred to by the term enchondroma. However, some are developed on the surface of the bone and constitute enchondromas [1].

Diagnosis is based on clinical and radiological arguments; histology is rarely used, except when a malignant transformation is suspected. The radiological as-

sessment is of capital importance, because the confirmation of the diagnosis can be made on a simple standard X-ray; the typical appearance of lesions in Ollier's disease is that of metaphyseal gaps of variable shape and size, with diaphyseal extension. These lacunae are surrounded by a thin line of osteosclerosis with fairly often intra-lesional calcifications [3]. X-rays also make it possible to highlight any deformations and shortenings of the affected bones [1]. Magnetic resonance imaging and possibly bone scintigraphy [5] are only requested if there is suspicion of sarcomatous transformation of chondromas. Several clinical forms of multiple enchondromatosis have been identified, including Maffucci syndrome [6] [7] which combines multiple chondromas and angiomatosis (especially subcutaneous). The risk of degeneration is high in this form [8] [9]. The frequency of degeneration is estimated at about 30% of cases. It should be mentioned when the chondromas become painful in the event of a rapid increase in their volume, as was the case for our patient. The location of these chondromas is also a risk factor for sarcomatous transformation. Indeed, unlike chondromas of the extremities, which are most often benign, the proximal location of tumors increases their risk of malignant degeneration. In our patient, the standard X-ray objectified, next to the proximal and middle phalanges of both hands, both feet, the distal end of the radius and the right tibia, multiple centromedullary lacunar images with clear content and clear contours with in places cortical notches and calcifications (**Figures 2-4**). We used histology in our case to confirm the disease because the mode of discovery was unfamiliar. It should be noted that no immature nature of the bone gaps was noted in our patient. The management of Ollier's disease is not codified. There is no medical treatment for this pathology. Therapeutic abstention is the only therapeutic option and surgery is reserved for disabling situations [10]. Our patient benefited from treatments based on diagnostic surgery-resection and oral Aspégic to calm the pain and inflammation.

Radiological monitoring is necessary in the management of Ollier's disease [11]. Today in our patient, the bone lesions remain stable without pain, however there are osteoarthritic changes in the hand (**Figure 6**). Finally, some authors have reported the indication of palliative radiotherapy in cases of inoperable chondrosarcomas, with satisfactory results [12]. Our patient did not present malignant lesions so she did not benefit from other treatments.

4. Conclusions

Ollier's disease is rare, you have to know how to think about it in the face of fortuitous discoveries, especially at an early age. Enchondromas are benign lesions that may present on imaging with nonspecific features in children.

In all cases, standard radiography is essential in the diagnosis and follow-up of Ollier's disease.

Conflicts of Interest

The authors declare no conflicts of interest.

Author Contributions

All authors contributed to data acquisition, data analysis and interpretation, and writing of the article.

Study Ethics

Informed consent from the patient's parents was obtained before the start of the study.

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