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Case report

Dieterich's disease: a case report of a very rare disease☆

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ABSTRACT

Dieterichś disease, or avascular necrosis of the metacarpal head, is a very rare disease, with just over 50 cases reported in the literature. Of unknown aetiology, it can manifest clinically in a variable way, from asymptomatic to obvious inflammation and painful functional limitation of the affected metacarpophalangeal joint. The case is presented of an 82-year-old patient who presented with pain at the level of the metacarpophalangeal joint of the third finger of the right hand of 1 year of duration without apparent cause. The physical examination showed no functional limitation or pain. Furthermore, no erythema, swelling, or mass effect was observed. A radiological study was carried out, leading to a diagnosis of advanced Dieterichś disease. Conservative treatment was started with nonsteroidal anti-inflammatory drugs, with a significant clinical improvement

Enfermedad de Dieterich: reporte de un caso de una enfermedad muy poco frecuente

Palabras clave:
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RESUMEN

La enfermedad de Dieterich o necrosis avascular de la cabeza de los metacarpianos es una enfermedad muy poco frecuente, con poco más de 50 casos reportados en la literatura. De etiología desconocida, clínicamente se puede manifestar de forma variable, desde asintomática hasta con evidente inflamación y limitación funcional dolorosa de la articulación metacarpofalángica afectada. Presentamos el caso de un paciente de 82 años que presentaba dolor a nivel de la articulación metacarpofalángica del tercer dedo de la mano derecha, de un año de evolución, sin causa aparente. La exploración física no evidenciaba

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limitación funcional, ni dolor, tampoco se objetivó eritema, tumefacción o efecto masa. Se realizó un estudio radiológico con diagnóstico de enfermedad de Dieterich avanzada, estableciendo tratamiento conservador con antiinflamatorios no esteroideos con mejoría clínica significativa.

Introduction

Dieterich's disease, or avascular necrosis of the head of the metacarpals, is an extremely rare condition, with just over 50 cases reported in the literature worldwide.[1] The most frequently affected metacarpals are the second and third, but any of them may be affected, or even several simultaneously.

The etiology of the disease is unknown and it may present spontaneously,[2] though it has also been associated with trauma,[3] steroid therapy,[4] avascular necrosis of the head of the head of the metatarsals[5] or kidney transplant.[6] The clinical manifestations are quite variable, ranging from absence of symptoms, to overt inflammation and painful functional limitation of the metacarpophalangeal joint affected. This article discusses a case involving this condition, of which little is known.

Clinical case

This is an 82-year old male patient who lives in a rural area and has been experiencing pain in the metacarpophalangeal joint of the third digit of the right hand for one year. The only relevant history was hypertension and high cholesterol, both receiving medical treatment. Initially, the patient visited his primary care physician, who wrote a detailed medical history expressing that the patient experienced mild generalized pain of all the interphalangeal and metacarpophalangeal joints in both hands, but pain was more severe in the metacarpophalangeal joint of the third digit of the right hand. The patient failed to recall any traumatic event on this joint to account for the existing pain. During the examination, the primary care physician did not identify any significant functional limitation, or edema, erythema, or hypertrophy on the soft tissues of the metacarpophalangeal joint of the third digit. Following this first visit, the patient received symptomatic therapy with paracetamol 1 g every 8 h. Approximately ten months later, the patient consulted plastic surgery because of the lack of improvement of the metacarpophalangeal joint of the third digit, though he experienced almost total pain remission in the other fingers. A plain posteroanterior and oblique x-ray was ordered, and the radiologist reported a morphological alteration of the head of the third metacarpal, mostly distal, with areas of osteolysis and loss of the metacarpal joint surface, and to a lesser degree, or the proximal phalanx. There were no radiological signs of aggressive disease. Moreover, the radiologist identified signs of arthrosis of the interphalangeal and metacarpophalangeal joints, of the second to the fifth fingers (Figs. 1 and 2). Based on the clinic and the radiological tests, and after ruling out infectious or tumor pathologies, the patient was diagnosed with Dieterich's disease. Based on this diagnosis and with the patient's agreement, conservative therapy was initiated with ibuprofen 600 mg every 8 h, resulting in a significant improvement of pain. The follow-up by the GP evidenced sustained clinical improvement, with no functional deterioration. At the request of the patient, no control imaging tests were performed.

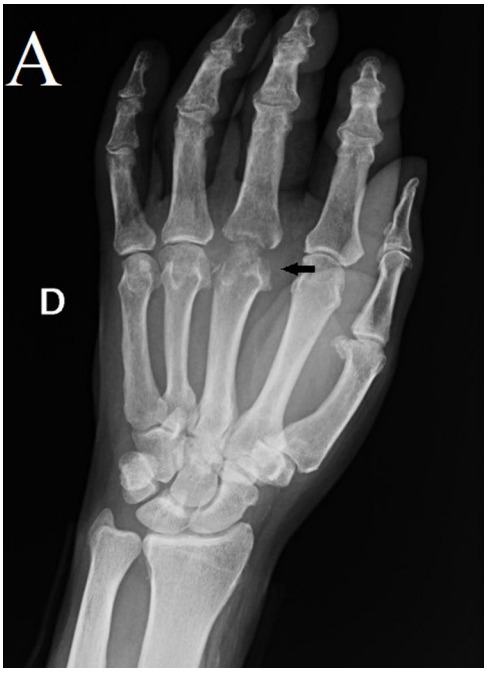

Fig. 1 – Plain PA x-ray of Dieterich's disease; third right hand metacarpal.

Discussion

Dieterich's disease is an extremely rare condition and little is known about it; therefore, its diagnosis is difficult and not standardized. Usually the diagnosis is delayed, which may lead to chronic functional consequences. In this particular case, the patient consulted one year after the onset of symptoms, since he believed his condition was age-related. After the assessment by the primary care physician, the initial suspected diagnosis was arthrosis, but the diagnosis was not confirmed since no imaging tests were conducted. Finally, and after almost two years since the onset of symptoms, the patient was diagnosed with advanced Dieterich's disease, which limited the therapeutic options, due in part to the delayed diagnosis.

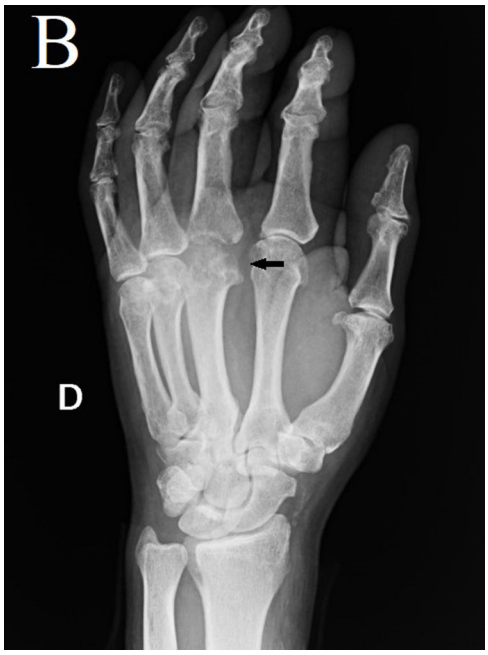

Fig. 2 – Plain oblique X-ray of Dieterich's disease; third right hand metacarpal.

The origin of this condition – such as it was case with this patient – may be unknown,[7] but some cases have been described associated with trauma, steroid therapy and systemic diseases.

The clinical manifestations are quite variable; it may be a casual finding in an otherwise asymptomatic patient, or may present with inflammation and progressively increasing pain of the affected metacarpophalangeal joint. These patients may also present with limitations in the range of movement of the joint, in addition to significant loss of strength.[8,9] The avascular necrosis of the head of the metacarpals affects more often the third metacarpal, followed by the second, and only exceptionally, the fifth,[7] though there are cases involving multiple, and even bilateral joints.[8] More than 50% of the cases develop in young adults under 20 years old, but it may present at any age. In the case of our patient, the affected metacarpal was the third, as is usually the case according to the literature; the clinic developed in an unusual manner, and hence the pathology was overlooked for a long time.

Plain x-rays are mandatory to diagnose Dieterich's disease. The x-rays show flattening of the joint surface of the metacarpal head, bone destruction, stenosis of the joint space, and even destruction and collapse of the joint,[9] as it was the case discussed in which the head of the metacarpal was significantly altered. Moreover, a plain x-ray is the tool of choice to monitor disease progression. Another diagnostic test which has proven to be useful in recent years is MRI, which shows the area of avascular necrosis as a low intensity area in T1, and a double line sign in T2.[10] No MRI was conducted in the patient herein discussed, at the request of the patient himself, who refused to undergo this test or the radiographic control.

The treatment for Dieterich's disease is ill defined, and there are several proposals in the literature with good functional results. Initially, conservative therapy with non-steroidal anti-inflammatory drugs is the most popular, but no consensus has yet been reached in terms of the treatment regimen or adequate treatment duration.[11] In the patient discussed, conservative therapy was successful and lasting, since up to this date, the patient continues without symptomatic or functional progression. When conservative treatment fails, various surgical approaches have been used, with good results, including curettage and autologous cancellous bone graft,[12] osteochondral mosaicplasty,[10] flexion osteotomy of the metacarpal head,[13] and hemiarthroplasty of the metacarpal head.[14]

Conclusion

Avascular necrosis of the head of the metacarpals is an extremely rare clinical condition and honestly, little knowledge exists about it. In many cases, it is an unexpected diagnosis in a patient in which it was not even considered as a differential diagnosis. This lack of knowledge results in delayed diagnosis, which may lead to severe functional sequalae. Due to the small number of cases, it has not been possible to establish a standardized therapeutic approach, though the consensus seems to be to start with conservative treatment, and only when it fails, to consider surgery.

Conflict of interests

None.

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