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Case report

BIZARRE FINGER PAROSTEAL OSTEOCHONDROMATOUS PROLIFERATION (NORA'S LESION): A CASE REPORT.

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ABSTRACT

Nora's lesion is a rare benign lesion that occurs as a locally aggressive parosteal mass localized in the short bones of the hands and feet. Its clinical presentation and radiological features are non-specific, so there is the risk that it may easily be misdiagnosed and treated inappropriately. The first-line treatment for Nora's lesion is surgical excision. Here we present a case of a 40-year-old woman affected by Nora's lesion of the distal phalanx of the second finger of the right hand, diagnosed by X-ray, Computed Tomography and Magnetic Resonance Imaging, and which required aggressive surgical management by amputation.

1. Introduction

Nora's lesion, also known as bizarre parosteal osteochondromatous proliferation (BPOP), is a rare benign lesion that occurs as a locally aggressive parosteal mass localized in the short bones of the hands and feet. Its clinical presentation and radiological features are non-specific, so there is the risk that it may easily be misdiagnosed and treated inappropriately. The first-line treatment for Nora's lesion is surgical excision. We here present a case of a 40-year-old woman affected by BPOP of the distal phalanx of the second finger of the right hand, diagnosed by X-ray, computed tomography (CT) and magnetic resonance imaging (MRI), and which required aggressive surgical management by amputation.

2. Case presentation

Here we report the case of a 40-year-old woman who presented at the Emergency Department due to a painful mass of the right hand, specifically the second finger distal phalanx, with accompanying functional weakness.

The patient had noticed the lesion, which had markedly increased in size in the last week, a few months before. It had now become painful. She reported neither trauma to her right hand in the months prior to admission and she denied having any family history of bone tumors.

Physical examination showed a 5 mm mass located in the palmar side of distal phalanx of the right hand second finger, fixed and elastic. There was a progressive limitation of flexion range of movement, not associated to swelling, redness, ecchymosis or skin ulceration.

Laboratory tests did not show any abnormalities.

The patient underwent a right-hand X-ray that revealed a cystic-like exostosis on the palmar side of the distal phalanx with bone cortical interruption without periosteal reaction (Figure 1).

In the same day, the patient underwent a wrist and hand CT that confirmed the X-ray findings, showing a 5 x 4 x 4 mm solid mass with bone cortical interruption, but without periosteal reaction or marrow invasion (Figure 2).

After some days, the patient underwent a contrast-enhanced right-hand MRI for a better mass characterization. MRI revealed a hyperintense mass in PD-SPAIR and T2 weighed sequences and hypointense in T1 weighed sequences, with a thin calcific ring. This lesion showed high contrast enhancement, cortical bone infiltration and flexor tendon compression (Figure 3).

Based on these findings, the lesion was biopsied. Histopathology report revealed proliferative and randomly arranged spindle cells and chondrocytes, with both new and calcified bone, strongly suggestive of BPOP. Therefore, aggressive surgical management was required and middle and distal phalanges of the right hand second finger amputation was performed.

The post-operative course was normal and without complications.

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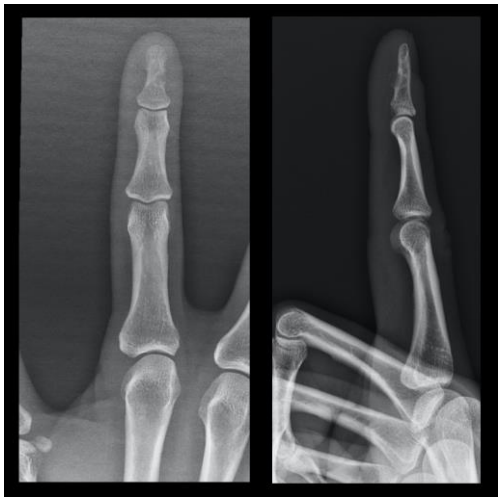

Figure 1. X-ray of the second finger of the right-hand, 2 projections: cystic-like exostosis at the palmar side of distal phalanx with bone cortical interruption without periosteal reaction.

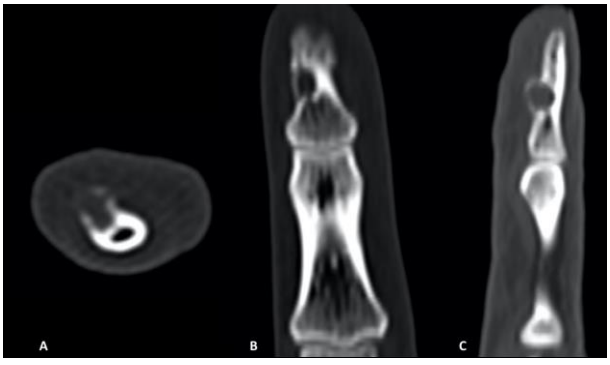

Figure 2. CT of the second finger of the right hand: axial (A), coronal (B) and sagittal (C) reconstructions. The finding is a 5 x 4 x 4 mm solid mass with bone cortical interruption, without periosteal reaction and marrow invasion.

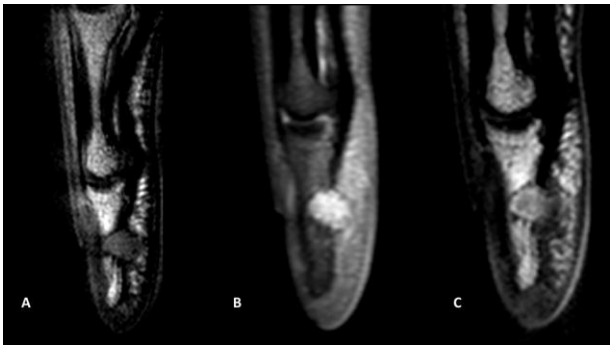

Figure 3. MRI of the second finger of the right hand: sagittal reconstructions in T1-w sequence (A), PD-SPAIR-w sequence (B), contrast-enhanced T1 (C).The finding is a mass hypointense in T1 weighed sequences and hyperintense in PD-SPAIR, with a thin calcific ring (A-B) and cortical bone infiltration. The lesion shows high contrast enhancement (C).

3. Discussion

Nora's lesion, also known as BPOP, is a rare benign and locally aggressive tumor arising from the bony cortex generally in the short bones [1-6]. BPOP may be located in different bones, but it is most commonly located in the short bones of the hands and feet [6] and more rarely in the long bones, skull or mandible [1; 7]. Notably 56% of cases involve the hand and 20% the foot [8].

It is a very rare condition, and only 160 cases have appeared in the literature [9] since it was described by Nora in 1983 [5]. It affects people at any age, most commonly in their third to fourth decades, without gender preference [10].

It is a mushroom-shaped parosteal locally aggressive mass that grows slowly over months or years [2-3] and protrudes from the superficial cortical bone, creating a pedunculated mass that can interrupt the cortical bone itself but does not invade the normal bone marrow.

The disease is caused by chromosomal abnormalities such as t(1;17) (q32;q21) [12] and t(1;17) (q42;q23) [4]. The exophytic mass may be composed of three heterogenous histological tissues: bone, cap cartilage and fibrous tissue [3-4].

Its clinical presentation is non-specific, it may present with pain and functional weakness or may be asymptomatic and discovered incidentally, for example after an accidental trauma at the site of occurrence [6].

Laboratory tests also are non-specific and not useful to diagnosis.

The imaging features of BPOP are variable and nonspecific. On X-ray, BPOP appears as a well-defined dense exophytic mass arising from cortical bone with or without osteolysis, projecting into the soft tissues [11]: these X-ray features are non-specific and alone they cannot reliably diagnose the lesion.

The main differential radiographic diagnoses are calcinosis, osteochondroma, periosteal chondroma, parosteal osteosarcoma, periosteal osteosarcoma and turrets exostosis [12-13].

On CT scan, BPOP appears as pedunculated intensely calcified and ossified masses with well-demarcated margins arising from the cortical bone with or without cortical interruption, but always without the involvement of the marrow cavity [2-4; 14]. The main differential CT diagnoses include osteochondroma, periosteal chondroma, parosteal osteosarcoma, periosteal osteosarcoma, periosteal chondrosarcoma, myositis ossificans and florid reactive periostitis ossificans [2-4; 15-16].

MRI can reveal more specific features of Nora's lesion, showing a bone cortex involvement without marrow invasion, periosteal reaction or soft tissue infiltration [17]. On MRI images, the lesion appears with hypointense signal in T1-weighted sequence, hyperintense signal in T2-weighted sequences [2] and homogeneous enhancement after gadolinium administration on T1 weighted sequences [3;5;18-20]. The main differential MRI and CT diagnoses are the same.

Although preoperative radiological studies should be considered essential to reach the correct diagnosis, the definitive diagnosis is made as a result of histopathological examination. No treatment is required if the patient is asymptomatic, while in symptomatic cases with pain and/or functional impotence the definitive treatment is surgical resection with wide margins or surgical amputation. After surgical treatment high rates of Nora's lesion recurrence have been reported at (20–55 %) [3;21] at a time range of between 10 and 120 months from surgical resection [22].

4. Conclusions

Nora's lesion is a rare benign exostotic osteochondromatous tumor arising from the bony cortex that occurs most commonly in the short bones of the hands and feet. Its clinical presentation is non-specific and it is sometimes asymptomatic.

We have here showed a case of BPOP in a hand finger, showing its diagnostic features both with X-ray, CT and MRI techniques.

Radiological presentation is variable and non-specific and it is difficult to reach a definitive diagnosis. Although preoperative radiological studies should be considered essential for correct diagnosis, it is important to progress a diagnostic suspicion of BPOP in a symptomatic patient to eventual surgery and therefore to the definitive diagnosis through histopathological examination.

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