

Bizarre parosteal osteochondromatous proliferation (Nora's lesion) affecting the distal end of femur: a case report and review of the literature

Nikolaos P. Kotsakis¹, Efstathios Zogakis¹, Antonios Asimakopoulos¹, Ioannis K. Triantafyllopoulos²

¹2nd Department of Orthopaedic Surgery, "G. Gennimatas" General Hospital, Athens

²Laboratory for the Research of Musculoskeletal System

Medical School, National & Kapodistrian University of Athens, Greece

ABSTRACT

Bizarre parosteal osteochondromatous proliferation (BPOP) or else Nora's lesion, as it is commonly known, was first described by a pathologist named Nora in 1983. Typically this rare disease is occurred in the small tubular bones of the hands and feet and especially in the proximal and middle phalanges as well as the metacarpal and the metatarsal bones. Here we present a case of a young female with a distal femur lesion originally thought to be an exostosis. A 19-year old, Caucasian female was referred to our outpatient clinic complaining of pain at her right femur. Plain x-rays revealed a 1.25cm, well – delimited, ossified, rounded mass with edema around it. Computed tomography showed a lesion in contact with the femoral metadiaphysis and surrounding by a slight thickening of the cortex. This lesion was more compatible with the presence of an exostosis. Based on the magnetic resonance imaging, the clinical and radiological findings the lesion seemed to represent BPOP. The isotope bone scan showed nothing pathological in other regions of the skeleton. The patient was taken to the operating room for resection of her bone lesion. The diagnosis of BPOP was confirmed by pathologic examination.

KEY WORDS: Bizarre parosteal osteochondromatous proliferation (BPOP); Nora's lesion; Femur

Introduction

Bizarre parosteal osteochondromatous proliferation (BPOP) or else Nora's lesion, as it is commonly known, was first described by a pathologist named Nora in 1983 [1]. His paper about this unique le-

sion was first published in The American Journal of Surgical Pathology and reported 35 cases of BPOP of the hands and feet [1]. Since then, over 200 cases of Nora's lesion have been presented in the literature, very few of which have affected the femur as

CORRESPONDING
AUTHOR,
GUARANTOR

Nikolaos P. Kotsakis,

2nd Department of Orthopaedic Surgery, "G. Gennimatas" General Hospital,
Mesogeion 154 Avenue, Athens, 156 69, Athens, Greece

e-mail: nikolaoskotsakis@gmail.com



Figure 1: Initial anteroposterior and lateral femur radiographs demonstrating a 1.25cm, well – delimited, ossified, rounded mass that was arising from the medial cortical aspect of the distal third of femoral diaphysis, above the medial femoral condyle

in the case of our patient.

Typically this rare disease is occurred in the small tubular bones of the hands and feet and especially in the proximal and middle phalanges as well as the metacarpal and the metatarsal bones [1]. But other more uncommon sites have been described. In 1993, almost 10 years after Nora, Meneses reported 65 cases of bizarre periosteal osteochondromatous proliferation [2]. Seventeen out of these cases involved the long bones. Six lesions involved the ulna, three the radius and the femur and two the tibia and the fibula. Even more rare sites that have been reported in the literature are the mandible, maxilla, sks skull and the sesamoid bones [2, 3, 4, 5].

Nora's lesion is occurred to a wide range of age.

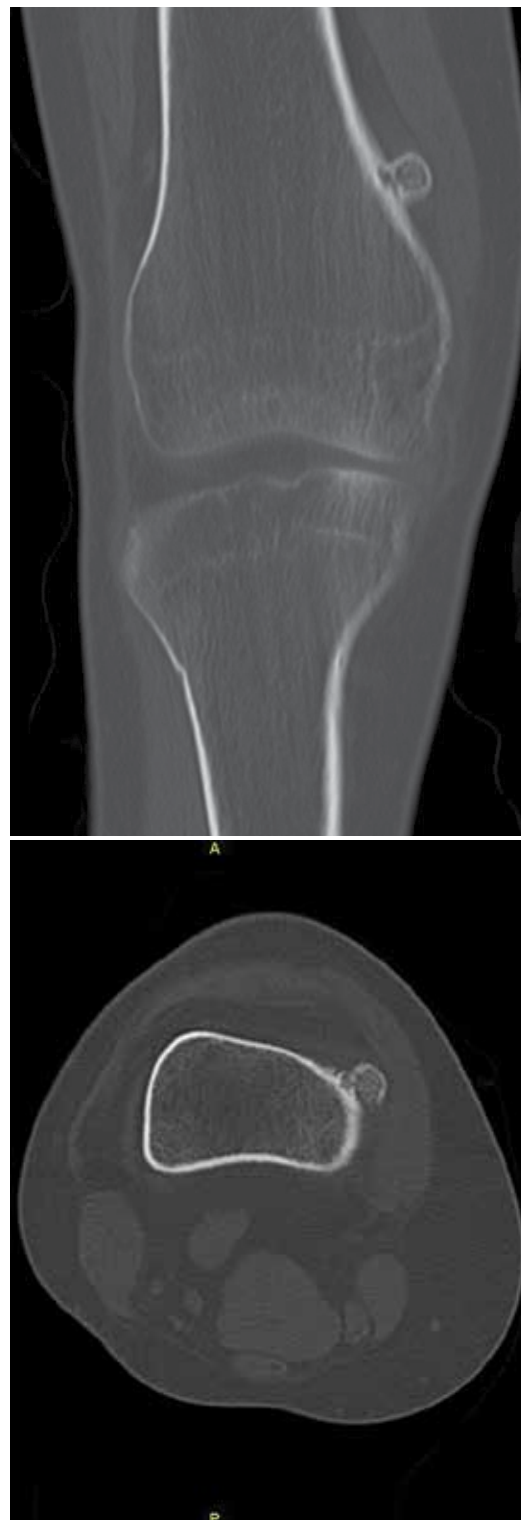


Figure 2: CT findings. Coronal and axial images show a lesion in contact with the medial aspect of the femoral meta-diaphysis, surrounded by cortex. Around the lesion was recognized a slight thickening of the cortex

Luigia Abramovici reported a range from 12 to 63 years with an average age of 30.3 years [6]. Meneses reported cases from 8 to 73 years old with an average of 33, 9. [2]. All authors agree that it is more usual during the third and fourth decade of life. Our patient was 19 years old when she came to our outpatient clinic. She experienced pain for the last six months to the medial aspect of her distal femur. Before this period she had not felt any pain or made any test to her femur so we cannot be sure when this lesion started growing or the rate which it had been growing

Case presentation

A 19-year-old woman presented to our outpatient clinic complaining of pain at her right femur. She could localize it particularly to the medial aspect of her distal femur. The pain was present almost every night, with no apparent trigger the last year. The patient referred that the last six months this could easily be reproduced with palpation. She denied any injury at her femur the last years and she had no history of any prior surgical intervention.

Clinical examination revealed a non-tender and non-mobile mass at the medial aspect of her distal femur. It was painful at palpation and her knee had a full range of active and passive motion.

Standard radiographs showed a 1.25cm, well-delimited, ossified, rounded mass that was arising from the medial cortical aspect of the distal third of femoral diaphysis, above the medial femoral condyle (Fig. 1). An impression of an edema of the soft tissue in the area was given.

Computed tomography revealed a clearly visible lesion in contact with the medial aspect of the femoral metadiaphysis, surrounded by cortex and exhibiting a maximum cranial-caudal diameter of approximately 8mm. Around the lesion was recognized a slight thickening of the cortex (Fig. 2). This lesion was more compatible with the presence of an exostosis and it was located medially of the vastus medialis.

On magnetic resonance imaging (MRI), the margins of the lesion had a low signal intensity on T1-weighted images and a high signal intensity on T2-

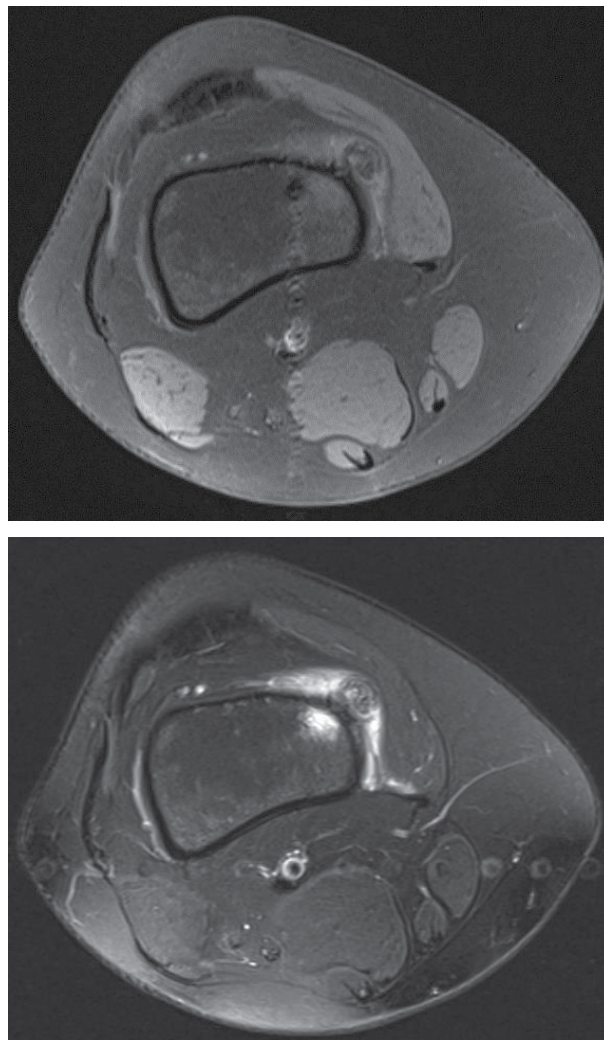


Figure 3: MRI findings. An axial T1-weighted MRI image that shows a low signal intensity at the margins of the lesion. An axial T2-weighted MRI image that shows a high signal intensity at the margins of the lesion.

weighted image (Fig.3). Based on the clinical and radiological findings, the lesion was felt to represent BPOP.

During the first phase of the isotope bone scans noted no pathologic concentration of the radiopharmaceutical in the examined area. In the second and third phases there was an increased and well delimited concentration - fixation of the radiopharmaceutical in the medial area of the femoral metadiaphysis. From the remaining skeleton nothing pathological was observed (Fig. 4).

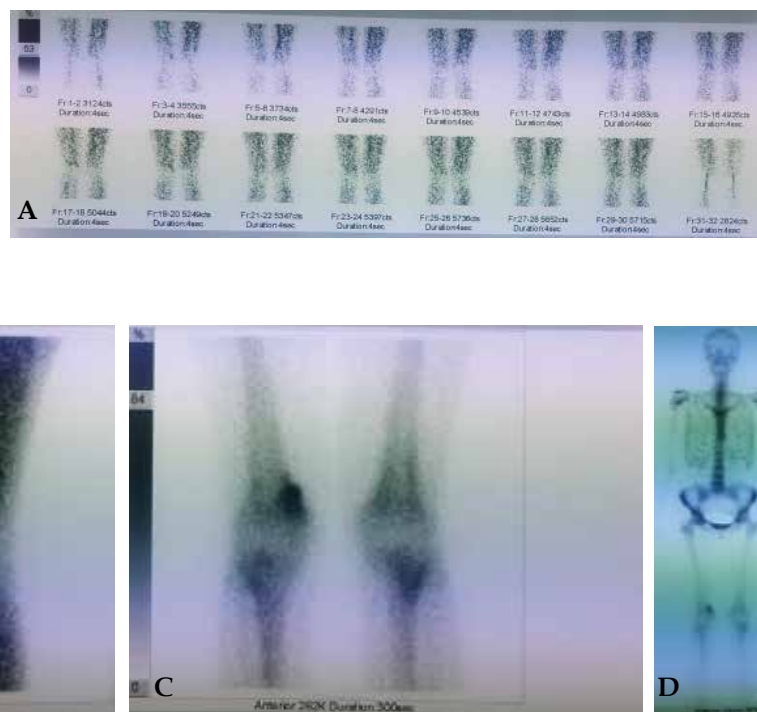


Figure 4: A First phase, B intermediate phase, C delayed phase and D whole-body flash views of the isotope bone scan. No pathologic concentration of the radiopharmaceutical during the first phase. In the second and third phases, there was an increased and well – delimited concentration – fixation of the radiopharmaceutical in the medial area of the femoral metadiaphysis. From the remaining skeleton nothing pathological was observed.

The patient underwent an excision of the femoral mass twelve months after the initial symptoms. The mass was identified through a medial approach under the medial vastus. The surface of the

the lesion was found to be covered by a cartilage cap and its interior to be composed of osteoid tissue in continuity with the cortical bone. The mass was excised with an osteotome and the abnormal area of underlying bone was decorticated (**Fig. 5**).

The gross and the microscopic examination revealed a reactive type of lesion consisting of bone and a cartilaginous hood on the surface of the bone. The cartilaginous plaque was rich of cells, the chondrocytes appeared to be enlarged and some of them binucleated. On the base of the plaque there was an endochondral ossification with blue calcification (stained with hematoxylin and eosin) at the junction point. Osteoid had a prominent osteoblastic edge, while the inter-

mediate spaces contained capillaries and loose layer. Moreover, it was noticed a spindle cell proliferation between bony trabeculae without atypia. These findings confirmed the diagnosis of BPOP.

Discussion

Bizarre parosteal osteochondromatous proliferation affects both sexes the same. In all the literature, we have found no difference between males and females in any research [2].

Data on symptoms are scarce. Most of the cases are asymptomatic or there is a pain during palpation of the lesion [1, 6]. Sometimes the authors correlate the tenderness with a quicker growth of the lesion [1]. More symptoms are reported and they depend on the site and the size of the tumor and the anatomical structures that are near. Varun K. Bhalla reported a case of a young child with a Nora's le-



Figure 5: Postoperative anteroposterior and lateral femur radiographs demonstrating resection of the mass.

sion on his distal femur that caused a popliteal artery pseudoaneurysm [7]. Beverlie L. Ting reported a case report of BPOP lesion of the ulna that caused an erosion of the adjacent radius [8]. Due to the fact that it is usually asymptomatic, most times it is discovered incidentally. A percentage of patients report prior trauma on the site of the growth but it is confirmed as a triggering factor by any research [9].

The radiologic findings of BPOP is useful to differentiate them from osteochondromas [1]. This benign tumor arises directly from the cortical surface of the bone and has no communication with the medulla. As a result, it does not disturb the architecture of the bone [6]. The lesions are usually calcified and they resemble as a soft-tissue tumor like mass that is attached to the bony cortex [10, 2]. Nora's lesion can be observed in plain radiographs but because of its soft tissue nature it can be very useful to perform


an MRI test on the patient.

Despite the clinical and radiographic characteristics of this disease, the diagnosis can only be obtained through a biopsy and a histological examination. The size is usually from 0.3 to 3cm [1].

The histologic findings are firstly an irregular maturation of cartilage into bone that produces chondro-osteoid that has a characteristic blue quality and it is often called blue bone. Secondly it contains large bizarre binucleated chondrocyte that often matures into bone. Lastly, it can be noticed a spindle cell proliferation between bony trabeculae without atypia [1, 2, 6].

The common treatment of this disease requires surgery and it is a complete excision of the lesion. It has high recurrence rates ranging from weeks postoperatively to years [2, 6]. Despite this fact and its sometimes-aggressive outgrowth, it is a benign tu-

mor. There are no reports of malignant conversion, metastases or correlated diseases or deaths because of this disease in the literature [11]. In our patient the whole mass was excised and the underlying bone was decorticated. We preferred this method so that we can reduce the possibility of recurrence. Follow up was up to two years in our outpatient clinic. No pain was reported during this period. The patient had no complications because of the surgery. Two years after the operation, the patient was contacted by telephone and no symptoms were mentioned.

In summary, bizarre parosteal osteochondromatous proliferation is a benign tumor-like lesion that has a characteristic radiographic and histologic appearance. It is usually asymptomatic but it needs surgical removal so it can be differentiated from other malignant tumors. It has great rates of recurrence so it is very important to follow up the patient for a period of time in order to diagnose it. 

Conflict of interest:

The authors declared no conflicts of interest.

REFERENCES

1. Nora FE, Dahlin DC, Beabout JW et al. , Bizarre parosteal osteochondromatous proliferations of the hands and feet, *Am J SurgPathol*, 1983.
2. Meneses MF, Unni KK, Swee RG et al., Bizarre parosteal osteochondromatous proliferation of bone (Nora's lesion), *Am J SurgPathol*. 1993.
3. Dashti HM, Reith JD, Schlott BJ et al., Bizarre parosteal osteochondromatous proliferation (Nora's Lesion) of the mandible. a rare bony lesion, *Head Neck Pathol* , 2012.
4. Nobusawa A, Sano T, Neqishi A et al., Bizarre parosteal osteochondromatous proliferation of the maxilla: a case report, *Oral Surg Oral Med Oral Pathol Oral Radiol* , 2012.
5. Berber O, Dawson-Bowling S, Jalqaonkar A et al., Bizarre parosteal osteochondromatous proliferation of bone: clinical management of a series of 22 cases, *J Bone Surg Br*, 2011.
6. Abramovici L, Steiner GC, Bizarre parosteal osteochondromatous proliferation (Nora's lesion): a retrospective study of 12 cases, 2 arising in long bones, *Hum Pathol* , 2002.
7. Bhalla VK, Coulson H, Parker w et al., Popliteal pseudoaneurysm caused by Nora's lesion of the femur in a young child: a rare presentation and first report, *J PediatrSurg* , 2012.
8. Tinq BL, Jupiter JB, Recurrent bizarre parosteal osteochondromatous proliferation of the ulna with erosion of the adjacent radius: case report, *J Hand Surg Am*, 2013.
9. Kershen LM, Schucany WG, Gilbert WG, Nora's lesion: bizarre parosteal osteochondromatous proliferation of the tibia, *Proc (BaylUniv Med Cent)*, 2012.
10. Rybal LD, Abramovici L, Kenan S et al., Cortico-medullary continuity in bizarre parosteal osteochondromatous proliferation mimicking osteochondroma on imaging, *Skeletal Radiol*, 2007.
11. Gruber G, Giessauf G, Leithner A, Bizarre parosteal osteochondromatous proliferation (Nora lesion): a report of 3 cases and a review of the literature, *Can J Surg*, 2008.

READY - MADE
CITATION

Kotsakis NP, Zogakis E, Asimakopoulos A, Triantafyllopoulos IK. Bizarre parosteal osteochondromatous proliferation (Nora's lesion) affecting the distal end of femur: a case report and review of the literature. *Acta Orthop Trauma Hell* 2019; 70(2): 65-70.