



Bizzarre parosteal Osteochondromatous Proliferation

Case Report

Ann. Ital. Chir.

Published online (EP) 15 April 2013

pii: S2239253X13020999

www.annitalchir.com

Maria Giuseppina Onesti*, Sara Carella*, Vittoria Amorosi*, Doria Pedace**,
Domenico Campagna***, Pasquale Fino*, Claudio Latini****

University of Rome "Sapienza", Policlinico Umberto I, Rome, Italy

*Department of Plastic, Reconstructive and Aesthetic Surgery

**Specialist in Dermatology and Venereology

***Department of Pathology, San Giovanni-Addolorata Hospital, Rome, Italy

****Department of Plastic and maxillofacial surgery, San Giovanni-Addolorata Hospital, Rome, Italy

Bizzarre parosteal osteochondroma proliferation. Case report

Nora's tumor, also known as bizzarre parosteal osteochondromatous proliferation (POPB), is an exophytic outgrowth arising from the cortical surface of the bone that consists of a mixture of bone, cartilage and fibrous tissue. It is a benign lesion with atypical microscopic features and a tendency to recur. It must be distinguishable from parosteal osteogenic sarcoma, parosteal chondrosarcoma, osteochondroma, florid reactive periostitis, turret exostosis, subungueal exostosis, myositis ossificans. The treatment is surgical, but a high rate of local relapse is described. The diagnosis is histological. We report a case of a patient with POPB involving the foot, underwent surgical excision and with no evidence of recurrence at one year.

KEY WORDS: Bizarre parosteal osteochondromatous proliferation; Nora's lesion.

Introduction

Nora's lesion, first described by Nora in 1983 ¹, is a rare benign lesion with an atypical microscopic aspect and a high local recurrence rate ^{1,2}. Over 160 cases of BPOP have been presented in the literature to date. The more frequent anatomical sites are the bones of the hand and

foot (70% of cases), other places reported are the long bones (humerus, radius, ulna, tibia, fibula, and femur) and also the skull-facial bones ²⁻⁴. The lesion occurs in patients of all ages (between 8 and 74 years of age), with a higher rate in the 4th and 5th decade of life, with no particular difference between sexes ⁴. Clinically it appears as a round type mass, adherent to the underlying bone surface, with a thick consistency and tends to increase in volume, rarely painful. Radiographs show calcified and osseous mass adjacent to the affected bone. The underlying bone had no cortical flaring or structural alteration. The computed tomography scans show intensely calcified and ossified masses with well defined margins. There is no continuity with the medullary canal of the bones from which the masses originated; it can extend to the nearby soft tissues without however infiltrating them ⁵⁻⁷. Bone scintigraphy with Tc99 demonstrates an abnormal uptake in the mass while the uptake in other parts of the body is normal ⁸. Radiographically

Pervenuto in Redazione Novembre 2012. Accettato per la pubblicazione Gennaio 2013

Correspondence to: Pasquale Fino M.D., Department of Plastic, Reconstructive and Aesthetic Surgery, University of Rome "La Sapienza", Policlinico Umberto I, Viale Pantelleria, 35, Scala B, Interno 1/A, 00141, Rome, Italy (e-mail: pasquale.fino@gmail.com, pasquale.fino@uniroma1.it)

BPOP can resemble malign growths or osteomyelitis when in presence of an intramarrow extension of the inflammation. Preservation of cortical bone under the osteocartilaginous mass on T1-weighted magnetic resonance imaging and homogenous intramedullary enhancement with gadolinium diethylenetriamine pentaacetic acid can be helpful for distinguishing BPOP from malignant lesions⁹. Diagnosis is confirmed by histological examination that reveals a fibrose and osteocartilaginous proliferation. In particular the lesion is composed of fibrocartilaginous tissue where the chondrocytes have increased in number, with a bizarre aspect (enlarged) and binucleate^{2,6-8,10}. With regards to the fibrocartilaginous tissue there are areas of bone metaplasia with osteoid tissue irregularly calcified^{2,4,10}. The basal area of the lesion, which corresponds to the osteocartilaginous interface, is irregular and the bone trabeculae stained mostly deep blue with hematoxylin and eosin^{2,5,8,11}. The therapy is surgical and consists of the excision of the pseudocapsule, a part of the adjacent periosteal tissue and the cortical of underlying bone^{2,11-13}.

This report presents one case of BPOP of the left foot, its clinical and histological features as well as the different etiologic theories. The differential diagnosis is broad including both benign and malignant processes. Despite its rarity, it is important to know and consider Nora's lesion in all cases of growths of the parosteal bone surface.

Case Report

A 57-year-old man presented with a nodular mass of hard consistency on the plant of the left foot in March 2010. Anamnesticly, the lesion appeared 2 years before and no injury at the site of the neoformation was described. During the time the lesion increased reaching the dimension of 5 cm. The patient lamented sensibility impairment and inability to flex and extend the 3th and 4th finger of the foot. The clinical exam showed in the region of the 3th and 4th metatarsus at the base of the left foot multi-lobulated swelling, which was bluish white in appearance, and measuring 5 x 4 x 3,2 cm. The skin above was without inflammation. On touch the lesion appeared hard and adhering to the bone surface below. There was no evidence of lymphadenopathy and the general clinical exam was normal. In April 2010 the patient underwent surgical excision of the lesion at our institute, which in surgery appeared pseudo encapsulated (it did not infiltrate into the soft surrounding tissues), white grey in colour with a wide base in the region of the 4th metatarsus lying between the flexor tendons of the foot which it in part compressed. The lesion was osteotomized at its base. Cut section of the mass showed glistening, traslucent appearance with grayish white areas at the periphery and granular bony tissue below it, with areas of chalky white appearance in between (Fig. 1). The



Fig. 1: Cut sections of the lesion on the the plant of the left foot.

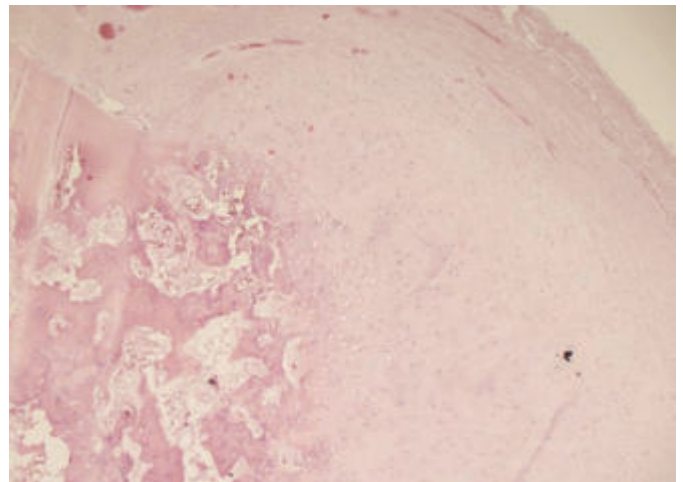


Fig. 2: Histologically, a zonal architecture is apparent with central or basally located new bone surrounded by a peripheral cap of cartilage. (Hematoxylin and eosin stain, low magnification).

histopathological exam showed a hypercellular osteocartilaginous proliferation made up of enlarged chondrocytes with a bizarre aspect and binucleate. There were areas of bone metaplasia coloured blue with hematoxylin and eosin staining (Figs. 2, 3, 4). On the basis of the clinical and histopathological data a diagnosis of Nora's lesion was emitted. After 2 years from surgery there was no evidence of recurrence and the patient no longer reports symptoms of pain during movements and extensions of the foot.

Discussion

Nora's lesion or Bizarre Parosteal Osteochondromatous Proliferation (POPB) is a rare benign mesenchymal min-

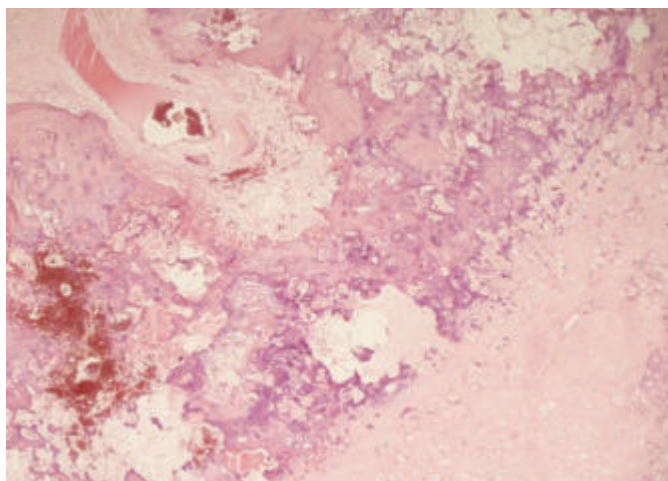


Fig. 3: Light micrograph of the lesion showing a osteoblast-rimmed osteoid material and a surrounding chondroid zone. (Hemotoxylin and eosin stain, low magnification).

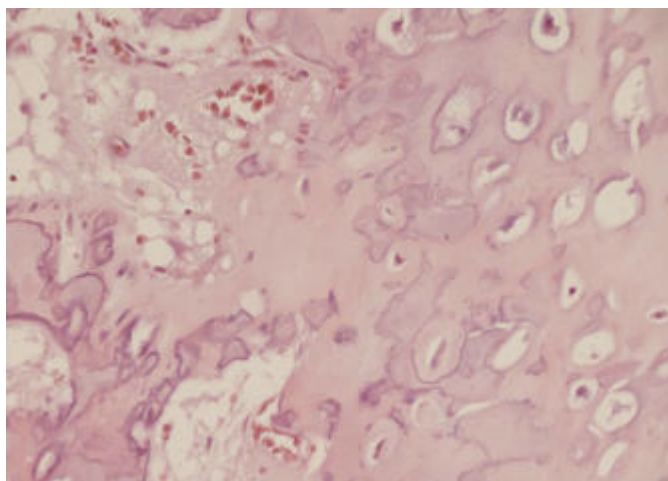


Fig. 4: Light micrograph of Nora's lesion with trabeculae of osteoid. A mixture of cartilage, fibrous tissue and bone. The chondrocytes were bizarre and irregularly arranged, with occasional bi-nucleated cells. (Hemotoxylin and eosin stain, high magnification).

eralized proliferative lesion that typically interests the surface of the bones of the hand and foot, usually the proximal phalanges and metacarpal and metatarsal bones^{2,4,14}. These lesions present a high rate of local recurrence after surgical removal. The rates of recurrence vary between 29% and 55% in an interval of 2 years. There have also been cases of a second recurrence^{2,4}. Nora et al. presented 35 cases of BPOP with 18 (51%) local recurrences¹. Meneses found a rate of 55% in 65 patients treated surgically^{2,15}. Dhont reported a rate of recurrence of 29% in 24 patients treated^{2,16}. The high rate of recurrence after surgery seems to depend on an uncomplete asportation during surgery, which requires also the removal of the adjacent periosteal tissue and

the decortication of the bone involved^{11,12,17}. PQPB must be differentiated from other benign or malign proliferative lesions with parosteal localizations.

Firstly it must be differentiated from the parosteal osteosarcoma which rarely appears on the bones of the hand and foot and which is made up of mesenchymal anaplastic tissue with osteoid production. The nearby cortical becomes thinner because of the tumour invasion and the neoplastic tissue infiltrates the peripheral soft tissues^{2,4,18}. The condrosarcoma of which a type with parosteal localization is known, is the most frequent malign bone tumour of the hand and is different from Nora's lesion because of the cytological anaplasia and the local infiltration^{2,4,18,19}. It must also differentiate Nora's lesion from the osteochondroma, an osteocartilaginous exostosis with parosteal localization which appears in the early years of life.

Osteochondromas are extremely uncommon in the small bones of the distal extremities. They show the typical continuity with the medullary canal and the cartilage does not show any signs of atypia. It seems as if the cortical of the adjacent bone extended outwards to become the cortical of the exostosis whose spongiosis continues with that of the metaphysis of the bone from which it originates. Histopathologic examination is the best method to identify this lesion and should be performed for definitive diagnosis. In the osteochondroma it shows a cartilaginous cap of variable thickness with endochondral ossification in the proximity of the central part of the neoformation^{2,4,19,20}. The differential diagnosis between BPOP and periosteal chondroma is fundamental, as highlighted by Lichtenstein in 1952²¹. Clinical anamnesis, objective observation and radiological exams are insufficient to discern between the two neoformations. Even though BPOP is more common in the fourth decade of life and periosteal chondroma in the third one and fourth decade, both may be also found in a wide interval of time. Furthermore many cases of BPOP seem to have a traumatic etiology but in most cases of BPOP and periosteal chondroma this correlation has not been described. Pain is not a discriminator either, as it may or may not be present in either lesion. Histology is the best discriminatory parameter between the two lesions²².

BPOP must be differentiated from some periosteal reactive processes which develop after injury such as florid reactive periostitis, turret exostosis, myositis ossificans and subungual exostosis^{2,4,13,23,24}. Florid reactive periostitis usually concerns the bones of the hand and is histologically characterized by a subperiosteal proliferation of bone trabeculae within a fibrous matrix. It is different from POPB because of the absence of a cartilaginous matrix and of the typical bizarre and binucleate chondrocytes.

Turret exostosis also defined "acquired osteochondroma" is more frequently localized on the fingers of the foot and is characterized by a central bone covered with a

cartilagineo envelope. Subungual exostosis represents the localization of subungual turret exostosis. Myositis ossificans is an heterotopical intramuscular ossifying phenomenon and a consequence of injury, which histologically translates into the presence of mature osteoid tissue on the borders and indifferentiated mesenchymal tissues in the central part. There is no indication of continuity with the nearby bone cortical as it delimited by muscle tissue.

Dorfman et al.²⁵ suggest that Nora's lesion, florid reactive periostitis and turret exostosis are different evolutive morphological expressions of the same reactive process to an injury where POPB could represent an intermediate evolutive stage^{2,4,26}. The studies of Horiguchi in 2001 suggest that POPB is the expression of a periosteal process secondary to injury²⁷. Meneses et al. noted a correlation between antecedent trauma and development of a BPOP in 9 of their 65 patients²⁸. According to Yuen et al. the initial stimulus, like an injury, causes a subperiosteal hemorrhagic proliferation which then further develops^{6,13}. In contrast with the above stated theories, Zambrano et al., through a study on the chromosomic rearrangement of POPB lesions, suggest the neoplastic nature of the lesion²⁹. The cytogenetic study carried out by Nilsson et al. on 5 cases of POPB shows a recurrent balanced chromosomic translocation t(1;17)(q32; q21). To investigate the specificity of this reciprocal translocation, they screened the karyotypes of more than 43000 neoplasms and found no identical translocation. It seems to be a recurrent and pathogenetically significant aberration in BPOP. The occurrence of translocation supports the assumption that a neoplastic process may be the etiologic agent³⁰. Cases of malign transformation, local and distant metastasis or death associated with BPOP have never been found⁶. However further studies are necessary to establish a clear etiology of this condition.

Riassunto

Il tumore di Nora, conosciuto anche come proliferazione osteocondromatosa parostale bizzarra (POPB), è una neoformazione esofitica a partenza dalla superficie corticale dell'osso, costituita da tessuto osseo, cartilagineo e fibroso. È una lesione benigna, con aspetti microscopici atipici e tendenza alla recidiva locale. Deve essere differenziato dal sarcoma osteogenico parostale, dal condrosarcoma parostale, dall'osteochondroma, dalla periostite reattiva florida, dall'esostosi turrita, dall'esostosi sub ungueale, dalla miosite ossificante. Il trattamento è chirurgico, basato sull'escissione completa, ma c'è un'alta tendenza di recidiva locale. La diagnosi può essere posta mediante l'esame istologico. Gli Autori riportano un caso localizzato al piede.

References

1. Nora FE, Dahlin DC, Beabout JW: *Bizarre parosteal osteochondromatous proliferations of the hands and feet*. Am J Surg Pathol, 1983; 71:245-50.
2. Gruber G, Giessauf C, et al.: *Bizarre parosteal osteochondromatous proliferation (Nora's lesion): A report of 3 cases and a review of the literature*. Can J Surg, 2008; 51(6):486-89.
3. Torreggiani WC, Munk PL, et al.: *MR imaging features of bizarre parosteal osteochondromatous proliferation of bone*. Eur J Radiol, 2001; 40:224-31.
4. Cigna E, Tarallo M, Scuderi N, et al.: *Nora's lesion of the thumb and a differential diagnosis: Case report*. Eur J Plast Surg, DOI 10.1007/s00238-010-0456-y
5. Le Bellec Y, Asfazadourian H: *Prolifération ostéocartilagineuse parostéale bizarre ou tumeur de Nora. A' propos de deux cas*. Chir Main, 2005; 24:265-69.
6. Bush JB., Reith JD, Meyer MS: *Bizarre parosteal osteochondromatous proliferation of the proximal humerus: Case report*. Skeletal Radiol, 2007; 36:1535-40.
7. Rybak LD, Abramovici L, Kenan S. et al.: *Cortico-medullary continuity in bizarre parosteal osteochondromatous proliferation mimicking osteochondroma on imaging*. Skeletal Radiol, 2007; 36:829-34.
8. Garcia-Alvarez F, Laclériga AF, et al.: *Bizarre parosteal osteochondromatous proliferation. Difficulty in diagnosis*. Chir Organi Mov, 1999; 84(2):179-82.
9. Orui H, Ishikawa A, Tsuchiya T, et al.: *Magnetic resonance imaging characteristics of bizarre parosteal osteochondromatous proliferation of the hand: A case report*. J Hand Surgery, 2002; 27A:1104-108.
10. Moretti B, Di Giovanni A, Martino F et al.: *Nora's lesion. Clinical and therapeutic considerations*. Chir Organi Mov, 2008; 92(1): 45-49.
11. Mohammad A, Kilcoyne A, Blake S, et al.: *Second toe swelling: Nora's lesion or glomus tumour, case report and literature review*. Ir. J Med Sci, DOI10.1007/s11845-009-0435-0
12. Campanacci D A, Guarracino R, Franchi A, et al.: *Bizarre parosteal osteochondromatous proliferation (Nora's lesion). Description of six cases and a review of the literature*. Chir Organi Mov, 1999; 84:65-71.
13. Yuen M, Friedmann L, Orr W, et al.: *Proliferative periosteal processes of phalanges; A unitary hypothesis*. Skeletal Radiol, 1992; 21: 301-03.
14. Harty JA, Kelly P, Niall D, et al.: *Bizarre parosteal osteochondromatous proliferation of the sesamoid (Nora's lesion): A case report*. Foot Ankle, Int, 2000; 21:408-12.
15. Meneses MF, Unni KK, Sweet RG: *Bizarre parosteal osteochondromatous proliferation of the bone (Nora's lesion)*. Am J Surg Pathol, 1993; 17:691-97.
16. Dhondt E, Oudenhoven L, Khan S: *Nora's lesion, a distinct radiological entity?* Skeletal Radiol, 2006; 35:497-502.
17. 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; 33(12):1205-210.

18. Bandiera S, Bacchini P, Bertoni F: *Bizarre parosteal osteochondromatous proliferation of bone*. Skeletal Radiol, 1998; 27:154-56.
19. Singh R., Jain M., et al: *Unusual presentation of bizarre parosteal osteochondromatous lesion of the second toe (Nora's lesion)*. Foot Ankle Spec, 2010; 3(6):347-51.
20. Gursel E, Jarrahejad P, et al.: *Nora's lesion: Case report and literature review of a bizarre parosteal osteochondromatous proliferation of a small finger*. Can J Plast Surg, 2008; 16(4):232-35.
21. Lichtenstein L, Hall JE: *Periosteal chondroma; A distinctive benign cartilage tumor*. J Bone Joint Surg Am, 1952; 24A: 691-97.
22. Flint JH, McKay PL: *Bizarre parosteal osteochondromatous proliferation and periosteal chondroma: A comparative report and review of the literature*. The Journal of Hand Surgery, 2007; 32A:893-98.
23. Rampoldi M., Mariano P. Casareale P: *Bizarre parosteal osteochondromatous proliferation (Nora's lesion): report of two cases*. J Orthopaed Traumatol, 2005; 6:101-04.
24. Endo M, Hasegawa T, Tashiro T et al.: *Bizarre parosteal osteochondromatous proliferation with a t(1;17)Translocation*. Virchows Arch, 2005; 447:99-102.
25. Dorfman HD, Czerniak B: *Bone tumors*. St. Louis (MO): Mosby; 1998.
26. Sundaram M, Wang L, Rotman M, et al.: *Florid reactive periostitis and bizarre parosteal osteochondromatous proliferation: prebiopsy imaging evolution, treatment and outcome*. Skelet Radiol, 2001; 30: 192-98.
27. Horiguchi H, Sakane M, et al.: *Bizarre parosteal osteochondromatous proliferation (Nora's lesion) of the foot*. Pathol Int, 2001; 51:816-23.
28. Meneses MF, Unni KK, Sweet RG: *Bizarre parosteal osteochondromatous proliferation of bone (Nora's lesion)*. Am J Surg Pathol, 1993; 17:691-97.
29. Zambrano E., Nose V, Perez-Atayde AR et al.: *Distinct chromosomal rearrangements in subungual exostosis and bizarre parosteal osteochondromatous proliferation (Nora's lesion)*. Am J Surg Pathol, 2004; 28:1033-39.
30. Nilsson M, Domanski HA, Mertens F, et al.: *Molecular cytogenetic characterization of recurrent translocation breakpoints in bizarre parosteal osteochondromatous proliferation (Nora's lesion)*. Hum Pathol, 2004; 35(9):1063-19.

