Bizarre parosteal osteochondromatous proliferation (Nora’s lesion) of the talus: a case report

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INTRODUCTION

Bizarre Parosteal Osteochondromatous Proliferation (BPOP), or Nora’s Lesion, is a very rare condition, with less than 200 documented cases that exist in the literature.1 It’s first described by Nora in 1983, and therefore termed “Nora’s Lesion.”1-5 It commonly occurs on the proximal and middle phalanges, metacarpal, and metatarsal bones, with a predominance of 4:1 in the hand.1 It occurs mainly in adults and its occurrence in children and adolescents is rarely described in the literature. Patients are most commonly in their third to fourth decades, and both sexes are affected equally.3,4 Some authors hypothesized that traumatic events could be on the origin of it.1 Less than five cases of BPOP of the talus that have been reported in the literature. BPOP / Nora’s Lesion of the talus has not been reported in any literature in Indonesia. Therefore, this study aims to present a unique and rare case of BPOP (Nora’s Lesion) of the talus.

CASE PRESENTATION

An eighteen-year-old male patient admitted to hospital with a complaint of palpable lump at his front right ankle region since seven years ago with the size of a marble. On physical examination, there was a swelling of the anteromedial aspect of the ankle, hard in consistency, well-defined, immobile, and without neurovascular impairment. There was pain exacerbated by sports activities. There was a history of trauma at the located lump; he was fell while running on the ground a few years before the lump first arose with no fracture, wound, or swelling at that area. The range of motion (ROM) of the right ankle was less than the left ankle. The operation was performed under general anesthesia. After an incision was made, the mass was exposed and we did an osteotomy and a complete surgical excision.

Case:

A lump was seen at the anteromedial side of the right ankle region. The ankle joint X-Ray was performed. It showed a well-defined exostosis arising from the anteromedial aspect of the talus bone cortex, but not in continuity with it. Histopathological examination consistent with BPOP. We suggest a surgical “En-bloc” (complete surgical excision) as a recommended treatment for BPOP.
CASE REPORT

marginated of cartilage, lamellar bone, and fibrous tissues forming haphazard (irregular) formation. The H&E (Hematoxylin & Eosin) stain was used in this examination. Some parts of the cartilage component seemed hypercellular and contained the area of blue bone (“blue” tinctorial appearance), as well as the endochondral ossification. The fibrous tissues consisted of loosely-formed spindle cells between trabecular bone and there’s marrowbone between them (Fig. 4). The pathology finding was consistent with BPOP.

No sign of malignancy was seen in any of the resected specimens. The post-operative ankle joint X-Ray was performed (Fig. 5). The ROM of his right ankle was normal without pain a week after the operation.

Another follow-up was done three months after the operation. The ROM of the right ankle was great. No more pain was felt. No more lump is seen at the anteromedial side of the right ankle (Fig. 6).

DISCUSSION

BPOP, as known as Nora’s Lesion, was first described in 1983, when Nora et al. reported 35 examples of a proliferative lesion involving the small bones of the hands and, less often, the feet. The next was Meneses et al. in 1993, who presented 65 cases of this condition, which were consisted of 34 females and 31 males, ages ranged from 8 to 73 years (average, 3.9 years).

BPOP is a very rare condition, with less than 200 documented cases that exist in the literature. There are some doubts regarding its etiology and evolution. Some say that it is uncommonly related to trauma. It is described as a distinct entity containing atypical and bizarre cartilage that often undergoes a characteristic irregular ossification. It remains a challenging diagnosis due to the extremely rare presentation and its clinical and radiological similarities to more common osteogenic and chondrogenic tumors.

BPOP has no gender preference. It affects patients of any age, although most are in their 20s and 30s. Rottler et al. mentioned that the BPOP concerns in

The operation was performed under general anesthesia. After an incision was made, the mass was exposed and we did an osteotomy and a complete surgical excision. An irregular surfaced, well-defined, nearly 4 cm x 4 cm mass consisted of a pale-white bony structure surrounded by hard capsule showed up clearly (Fig. 3). Histopathological examination revealed tumor tissue slices with a well-
Resonance Imaging (MRI), can give more.

Tomography (CT) scan or Magnetic
is characteristic of BPOP.

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osteolysis, cortical flaring, or periosteal
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resemble osteochondromas, in that they
report by Rottler et al. in 2019.

most cases the metacarpal and tarsal bones
(76%), the long tubular bones (27%), and
the hand (56%) more than the foot (20%); hence the clinical suspicion in the talus
is low. The BPOP of the talus was first
reported by Rottler et al. in 2019.

Macroscopically the lesions do
resemble osteochondromas, in that they
consist of bone covered by cartilage. Radiographically, the lesion arises
from the cortical bone with or without
osteolysis, cortical flaring, or periosteal
reaction. A lack of medullary involvement
is characteristic of BPOP.

Other examinations, such as Computed
Tomography (CT) scan or Magnetic
Resonance Imaging (MRI), can give more
detailed information to plan a correct
therapeutic approach. On MRI, BPOP
presents with bone cortex involvement
without medullary involvement, periosteal
reaction, or soft tissue swelling. It can be
differentiated from osteogenic tumors
by the absence of cortical flaring of the
affected bone, and from chondromatous
tumors by the lack of communication
with the underlying medullary canal. However, the definitive diagnosis is given
by the histopathological examination.

Macroscopically, BPOP has three
components in varying amounts: cartilage,
bone, and fibrous tissue. The cartilage
usually forms a cap; less frequently, it
is arranged in lobules separated by
dense fibrous tissue with irregular
maturation into bone (endochondral
ossification). The bone that is produced
has a characteristic dark blue tinctorial
quality, especially at the interface with
the cartilage. The inter trabecular spaces
contain proliferating spindle cells that
lack cytological atypia. The cartilaginous
component is hypercellular and contains
irregular groups of binucleated and bizarre
chondrocytes. Several differential diagnoses
must be differentiated with BPOP.
The first are benign tumors, such as
osteochondroma, turret exostosis, and
florid reactive periostitis (FRP).

Osteochondroma, unlike BPOP,
radiologically presents medullary
continuity between the lesion and
the native bone, flaring cortex, and
histopathologically presents an organized
cap with hyaline cartilage. It commonly
occurs in the metaphysis of long bones,
such as the distal femur, proximal tibia, and
proximal humerus. Turret exostosis and
FRP should also be considered if there’s a
history of trauma. The most common
site of turret exostosis is index and little
fingers, yet there is also some literature
mentioning another location, such as
head, hallux, metacarpal, and talus. Histopathologically, turret exostosis shows
organized bone production, mature bony
architecture with thin cartilaginous. For
FRP, radiologically the underlying bone is
abnormal which shows medullary invasion,
reactive sclerosis, and periostitis. Usually,
the cartilaginous. Histopathologically,
FRP shows proliferating fibrous or spindle
cells with minimal osteocartilaginous
differentiation.

The second are malignant bone tumors, such as parosteal
or periosteal chondrosarcoma and
osteosarcoma. Nora’s Lesion can be
differentiated from malignant tumors by
its lack invasion into the adjacent muscle
and there is no neoplastic production of
the ostoid tissue.

A surgical “En-bloc” resection
(complete surgical excision) is the
recommended treatment for BPOP. Resection of its capsule and decortication
of the underlying cortical bone is reported
as fundamental to reduce recurrence
rates. This is important as the literature
reveals an index of recurrence of 20–55%
after surgical resection. Gursel et al.
mentioned that the recurrence was seen
in more than one-half of the cases and
generally occurred between two months
and two years postoperatively. Matsui et
al. had a different opinion, which was the
recurrence to occur from 10 to 120 months
(49 months on average) after surgery.

A limitation of our report is lack of CT-
scan and MRI evaluation, which might
have shown the discontinuity of the lesion
with the medullary cavity clearly.

CONCLUSION
BPOP / Nora’s Lesion of the talus is a
very rare case since there’re less than

Figure 5. Complete removal of Nora’s Lesion of the talus.

Figure 6. No more lump was seen at the anteromedial side of the right ankle.

5 cases that have been reported in the literature until 2020. BPOP of the talus has not been reported in any literature in Indonesia. Even though rare, it should be considered in the differential diagnosis of any osteogenic or chondromatous growth found in the foot. Histopathological examination is the definitive diagnosis of BPOP. In this case, the radiographic imaging showed well-defined exostosis arising from the anteromedial aspect of the talus bone cortex, but not in continuity with it, and the exostosis lacked invasion into the adjacent muscle. Histopathological examination revealed tumor tissue slices with a well-marginated cartilage, lamellar bone, and fibrous tissues forming haphazard (irregular) formation, which were consistent with BPOP. A surgical “En-bloc” (complete surgical excision) is the recommended treatment for BPOP. A further follow-up to the patient is important to ensure no recurrence arises at the excised bone.

ETHICAL STATEMENT

This study protocol has been approved by the Ethics Commission of Faculty of Medicine, Universitas Nusa Cendana.

CONFLICT OF INTEREST

None.

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AUTHOR’S CONTRIBUTION

All of authors contributed equally in this article.

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