Management of Nora’s Lesion: Case Series

Nora Lezyonunun Yönetimi: Vaka Serisi

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ÖZET


TARTIŞMA ve SONUÇ: Bizarre parosteal osteokondromatöz proliferasyon, vücudun birçok farklı lokalizasyonunda yerleşebilen, benign karaktere sahip ancak sık rekürrens ile karşımıza çıkmakta olan, nadir bir kemik lezyonudur. İyi bir preoperatif planlama ile yapılan dikkatli bir marginal eksizyon, nüksün önlenmesinde en önemlili anahtardır.

Anahtar Kelimeler: Bizarre parosteal osteochondromatous proliferation, Nora’s lesion, radius, ulna, poplitea

ABSTRACT

INTRODUCTION and OBJECTIVE: Also known as Nora lesion, bizarre parosteal osteochondromatous proliferation (BPOP) is a benign exophytic lesion originated from the bone. In this study, we aimed to evaluate four patients with Nora lesion who were evaluated between 2010 and 2018, and to review current information through a brief literature screening.

MATERIAL and METHODS: A total of four patients with a mean age of 32 ± 24 years (26-37 years) including 2 female and 2 male patients were included in the study. Patients’ time to diagnosis and symptoms, clinical findings, mass size, MRI findings, surgical treatments, treatment outcomes and complications were investigated.

RESULTS: Marginal excision was made to all the patients. The most common mass localization was the long bones of the forearm. Recurrence was developed in one patient at postoperative 24th month, and underwent relapse resection. The remaining three patients did not develop relapse or complications at follow-up.

DISCUSSION and CONCLUSION: Bizarre parosteal osteochondromatous proliferation is a rare bone lesion, which may be localized in many different part of the body, has a benign character, but may be encountered with frequent recurrence. A careful marginal excision to performed with a good preoperative planning is the most crucial key in prevention of relapses.

Keywords: Bizarre parosteal osteochondromatous proliferation, Nora’s lesion, radius, ulna, poplitea
INTRODUCTION

Also known as Nora’s lesion or periostitis ossificans, bizarre parosteal osteochondromatous proliferation (BPOP) is a rare lesion which was described for the first time by Nora et al. in 1983 [1-3]. Although so far BPOP has been reported in the literature many times as case reports, there are less than 10 case series in the literature on this issue [4-13].

BPOP is defined as a lesion containing bone, cartilage or fibrous tissue, and showing exophytic growth from the bone surface [5].

The most commonly involved regions are reported as phalanges, metacarpi and metatarsi, although rarely the lesions may also be seen in the long bones, skull, maxilla [6, 7]. In this study, we analyzed management of four BPOP cases with three localized in the long bones of the forearm, and one in the popliteal region, including the diagnosis, treatment and follow-up in the light of the current literature.

MATERIAL and METHODS

A screening which included only ‘Nora’ term was made on the Medline database from 2010 through 2018. Various search terms (‘nora’, ‘periostitis ossificans’, ‘bizarre parosteal osteochondromatous proliferation (BPOP)’) were used, including the combination of index and free text terms as recommended by Cochrane Handbook for Systematic Reviews of Interventions. The abstracts were screened and the relevant full text articles were further examined in details. References sections of the publications identified were screened for further literature review. All evidence levels were included.

Case Reports

Case 1:
A 29-year-old male patient presented to the outpatient clinic with the complaint of pain in the left forearm and palpable swelling. He stated that the swelling has existed for about one year and increasingly grown. The patient had no history of trauma.

Direct radiograph ordered showed an irregular mass lesion originated from the radius (Figure I). Contrast enhanced forearm MRI revealed a regular and lobulated contoured mass lesion of 27x25x15 mm in the central region of the left forearm, which was in vicinity of the radius central diaphyseal cortex, showed exophytic growth to the soft tissue, and was seen as isointense on T1A sections, and hyperintense on T2A sections and enhanced after gadolinium. (Figure II).

Marginal excision of the mass was performed. Histopathologic examination was compatible with periostitis ossificans. No recurrence or complication was seen at the follow-up for 36 months.

Case 2:
A 37-year-old female patient with occasional pain in the knee, stated that she had developed restrictions in her knee movements and her pain had increased in the last 8 months. She had no history of trauma.

Her radiological investigations revealed a lobulated contoured mass lesion with heterogeneous inner structure, which filled the popliteal fossa, reached to a size of of 47x73x62 mm at the largest part, was seen as isointense on T1A sections, and iso-hyperintense on T2A sections, accompanied by signal free millimetric areas in the central region, and heterogeneously enhanced after the injection of intravenous contrast agent (Figure III).

Figure 1: Case 1, radius originated, irregular contoured mass lesion is seen on left forearm antero-postero-lateral direct radiography.
Case 2: A 27-year-old female patient was referred to the orthopedics outpatient clinic with the complaints of swelling and pain in the right forearm. Radiological investigations of the patients revealed a lesion in the central of the right forearm, which was originated from the ulna, showed exophytic growth to the soft tissue, was seen as isointense on T1A sections, and hyperintense enhanced on T2A sections (Figure IV).

Atypical condrocytes were observed in examination of the diagnostic biopsy material. The patient underwent wide tumor resection. Histopathological diagnosis was reported as bizarre parosteal osteochondromatous proliferation. The patient developed recurrence at the 24th month of the follow-up in the postoperative period. Recurrence resection was performed upon recurrent mass was detected in the investigations of the patient who presented with painful swelling localized in the incision line.

Case 3: A 27-year-old female patient was referred to the orthopedics outpatient clinic with the complaints of swelling and pain in the right forearm. Radiological investigations of the patients revealed a lesion in the central of the right forearm, which was originated from the ulna, showed exophytic growth to the soft tissue, was seen as isointense on T1A sections, and hyperintense enhanced on T2A sections (Figure IV).

The patient underwent marginal excision. Pathologic outcome was compatible with Nora’s lesion. No recurrence was found at the last follow-up of the patient carried out at the 24th month. The patient was lost from the control in the next periods.

Case 4: A 40-year-old male patient presented to the outpatient clinic with the complaint of swelling in the volar side of the left wrist. Physical examination revealed a mass lesion of approximately 3 cm2 in the volar side of the wrist. Neurovascular examination was normal. Direct radiography and MRI revealed a mass lesion of about 33x23 mm in the palmar side of the wrist in the middle line intersosseous area in distal vicinity of the radius and ulna, which
was seen as isointense enhanced on T1A sections and hyperintense on T2A sections.

Tru-cut biopsy was performed, and the diagnosis was reported as chondroid lesion. Then, marginal excision of the mass was carried out. Pathologic outcome was compatible with Nora’s lesion. No recurrence was found at the 6th month follow-up of the patients who are currently under our control.

**DISCUSSION**

There are at least 100 cases that were reported in the literature and one of them extensively evaluated these lesions regarding etiology, cytogenetic aberrations, radiographic interpretation and treatment [5]. Less than 10 of these studies have been presented as case series.

Bizarre parosteal osteochondromatous proliferation (BPOP) is a rare, reactive, mineralized mesenchymal benign lesion, affecting bone surfaces [8]. In the present study, we analyzed case reports and case series published on Medline in 2010 and later. This analysis included total 22 articles and 60 cases.

Although BPOP may be seen in any age, the most common age range has been reported as 20-30 years [9]. In our literature screening, age range was between newborn and 65 years old, and the mean age was found as 34.6 years unlike the literature. In the present study, mean age of 4 patients was 33.2 years.

Studies have reported similar rates of BPOP in among the sexes [10]. In our literature review also 29 cases were female and 31 male patients, in the present study.

This tumoral lesion has a benign pathology, although it may be mistaken for malignant lesions. Because it occasionally shows a rapid growth and a high incidence of recurrence [7].

In addition, the differential diagnosis includes parosteal osteosarcoma because of its parosteal localization [11]. Furthermore, it may be confused with osteochondroma because of being localized on the bone surface and having cartilaginous component. It can be distinguished by not showing continuity with medullary canal [12].

Horiguchi et al., suggested that mechanism of BPOP development is a lesion secondary to injury [9]. However, most cases in the literature had no history of trauma.

Whereas malignancy or metastasis has not been reported in almost all studies about BPOP, Choi et al. described a case of BPOP associated with fibrosarcoma in 2001 [13]. Treatment method is resection with wide margins in aggressive benign and malignant tumors, while marginal excision is performed in benign tumors [14, 15]. No malignancy was found in our recent literature review and in the 4 patients in the current study [2-13, 16-18].

The current treatment method recommended for BPOP is marginal excision of symptomatic lesion. Intralesional excision is highly associated with recurrence [16]. We applied marginal excision in all of our patients after a careful preoperative planning.

High recurrence rates between 29% and 55% at 2-year follow-up have been reported in the literature, and even it has been reported that about half of these recurrent cases were referred again due to a second recurrence [1, 17, 18]. In our study, one of four patients developed recurrence at follow-up, and thus the rate of recurrence was 25%.

This study has several limitations. The study included data of a single center, and the number of patients was low because of the lesion being seen rarely. In addition, we evaluated the cases retrospectively. Further multicenter prospective studies with a larger number of patients are warranted.

**CONCLUSION**

Bizarre parosteal osteochondromatous proliferation (BPOP) is a rare bone lesion which may be localized in many different localizations of the body, and although being of benign character, it may be encountered with frequent recurrence. A careful marginal excision performed with a good preoperative planning, is the most important key in prevention of recurrence.

**REFERENCES**

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