Bizarre Parosteal Osteochondromatous Proliferation: a case report

SHIH-TING CHEN1, WEN-SHENG TZENG2, CHIEN-FENG LI3, CHIEN-HUNG LIN2, CLEMENT KUEN-HUANG CHEN2

Department of Medical Imaging1, Chi Mei Medical Center, Liou-Ying Campus, Tainan, Taiwan
Department of Medical Imaging2, Department of Pathology3, Chi Mei Medical Center, Tainan, Taiwan

ABSTRACT

Bizarre parosteal osteochondromatous proliferation (BPOP) is a benign disorder within a spectrum of reactive proliferative processes of tubular bones, particularly the phalanges of the fingers or toes. The cause is unknown, and it is difficult to distinguish BPOP from a bone or soft tissue tumor. The radiographic and magnetic resonance imaging manifestations of reported BPOP cases are mostly nonspecific. We present herein a case of BPOP with unique computed tomography features that enabled us to make a prospective diagnosis of BPOP for appropriate surgical planning.

CASE REPORT

A 31-year-old man suffered from painful swelling of his right second toe for 6 months. He had no history of major systemic disease, surgery, trauma, or hyperuricemia. A physical examination disclosed a swollen second toe with tenderness and limited range of motion. Clinical impressions included tophaceous gout, osteomyelitis, or a tumor growth. Laboratory data including routine blood, urine, stool studies, and serum uric acid level were all within normal ranges.

Frontal and medial oblique radiographs of the foot (Fig. 1) showed a juxtacortical radiolucent lesion with an incomplete sclerotic margin in the medial aspect of the proximal phalanx of the second toe and smooth periosteal reaction circumscribing the affected proximal phalanx. Focal and subtle cortical sauceration was noted. To the best of our knowledge, computed tomography (CT) features of BPOP have not been described until date. We report herein a case of BPOP, in addition to a non-specific MRI, presenting with unique CT features of benign solid periosteal reaction reminiscent of myositis ossificans (MO), which enables us to make a specific imaging diagnosis of BPOP.
**Figure 1.** Frontal radiograph of the right foot shows periosteal new bone formation (arrowheads) around the proximal phalanx of the second toe. Focal absence of periostitis (arrow) and overlying soft tissue lesion (asterisk) are seen over the medial aspect of the proximal toe. The cortex is preserved (open arrow).

**Figure 2.** Magnetic resonance image shows a mass-like lesion (asterisk) around the proximal phalanx of the right second toe. The lesion manifested as intermediate signal intensity on T1WI (a. and d.), heterogeneous low and intermediate signal intensity on T2WI (b. and e.), and heterogeneous enhancement after gadolinium administration (c. and f). The bone marrow was normal in signal intensity on these images. A periosteal reaction (arrowheads) circumscribing the affected phalange and the bony cortex was intact (open arrow). Focal absence of periosteal reaction (arrow) around the soft tissue lesion is seen.
Bizarre parosteal osteochondromatous proliferation

A noncontrast CT scan was also performed for better depiction of possible internal calcification, ossification, and osseous structure and showed solid periosteal new bone formation circumscribing the intact cortex of the proximal phalanx of the second toe in addition to a juxtacortical mass (Fig. 3). The CT scan was reminiscent of MO. The combination of focal disruption of periostitis and overlying soft tissue lesion on CT images lead us to the diagnosis of BPOP.

The patient underwent curettage of the lesion without difficulty or adhesions. Pathology reported prominent reactive fibroblasts in a storiform pattern with hypercellularity and increased mitotic activity without cellular atypia (Fig. 4). Focal areas of osteoid formation were observed between the fibroblastic proliferative areas. The histological diagnosis was consistent with BPOP.

A regular plain film follow-up for about 1 year and a clinical follow-up for about 2 years revealed resolution of the soft tissue lesion without local recurrence or metastasis.

DISCUSSION

BPOP is a rare benign disease first reported by Nora in 1983. It manifests as an uncommon reactive mineralizing mesenchymal lesion with an undetermined nature or trigger factor [1, 2]. This lesion is usually seen in young adults, mostly between 20 and 40 years of age, and involves the short tubular bones of the hand or foot, particularly the proximal phalanges. Antecedent trauma history, which was present in 12% of the cases in Michelsen’s series, does not appear to be a definitive etiological factor [3]. The clinical presentation includes progressive swelling, pain, and limited range of motion of the affected phalanx. In typical cases, plain radiographs may show tumor-like ossifications with internal lucent areas abutting on the periosteal aspect of an intact cortex without evident cortical destruction or medullary changes [4]. MRI reveals a soft tissue mass with low signal intensity on T1 weighted sequences and high signal intensity on T2 weighted or short τ inversion recovery sequences. The cortex and medulla appear normal [1]. The radiographic and MRI manifestations usually suggest a nonspecific soft tissue or bone mass making it difficult to arrive at a definitive diagnosis.

Another closely related benign lesion to BPOP is FRP. FRP was first described by Mallory in 1933 as a metaplastic and neoplastic soft tissue lesion containing bone and cartilage in the phalanges of the hands and toes [5]. The histological and radiological features of FRP are currently well-established. It is a benign fibro-osseous pseudotumor in the digital soft tissue. Similar to BPOP, there is usually no definite antecedent history of trauma. FRP demonstrates fusiform periosteal new bone formation around the affected phalanx on radiographs [6]. Before 1980, FRP and BPOP were considered....
distinctive entities and had a major differential diagnosis. Later studies found considerable overlap between FRP and BPOP. In 2001, Murali reported that FRP can be self-limiting or can evolve into BPOP [2] and proposed that each represents the opposite end of the spectrum of the same entity. In 1992, Yuen proposed that if the reaction remains contained within the periosteum, localized fusiform periostitis with combinations of fibrosis, cartilage, and bone elements develops and fits the description of FRP. If the periosteum is breached, the reactive process can then extend into the loose areolar tissue around the phalanges, producing the lobular BPOP lesion. Yuen et al advocated a unitary hypothesis among FRP, BPOP, and turret exostosis and proposed a general term, proliferative periosteal processes of phalanges (PPPP), for the entire spectrum. The specific appearance of each component in the spectrum depends on factors related to breaching of the periosteum, the stage of lesion evolution, maturation of the process, and local anatomic factors [7].

BPOP has been regarded as a variant of heterotopic ossification, arising from the cortical surface of the affected bone with focal disruption of the periostium and localized osseosecondral proliferation in the soft tissue. Chaabane et al. and Murali also reported that BPOP is a well-defined juxtacortical ossified mass without continuity with the medulla, which is close to the end stage of PPPP, i.e., turret exostosis [4, 8]. At this stage, the diagnosis is much easier on radiographs, MRI, or CT. However, in our case, plain radiographs and MRI revealed a prominent periosteal reaction and a nonspecific soft tissue mass suggesting malignancy. The solid periostial reaction with focal defect and heterotopic ossification-like appearance on CT images are reminiscent of a breached periostium with lobular reactive proliferation, characteristic of BPOP.

The differential diagnoses of BPOP include thopaceous gout, osteomyelitis, parosteal/periosteal osteosarcoma, juxtacortical chondrosarcoma, giant cell tumor of the tendon sheath and fibromatosis, and so on. In the absence of a clinical history, hyperuricemia, bone erosion, soft tissue calcifications, characteristic MR signals, or gout are scarcely likely. Osteomyelitis is easily excluded in the absence of clinical toxic signs, abnormal marrowsignal, bone destruction, symmetric soft tissue swelling, or abscess formation. The MR appearances of our case apparently did not fit the specific features of GCTTS or fibromatosis such as low T1 and T2 signal and presence of hemosiderin in the former [9, 10].

Juxtacortical tumors are a major concern. Lack of tumor bone and mineralization, features of chondral matrix, characteristic calcifications, subjacent cortical destruction, or aggressive patterns of periostitis are helpful for excluding tumors. Treatment of BPOP is surgical excision. Similar to heterotopic ossification, surgery is best performed after maturation of the reactive tissue proliferation for easier removal of the lesion and less local recurrence [2].

In summary, although BPOP is rare, some imaging features allow a prospective imaging diagnosis. A solid periosteal reaction with or without focal breach of periostitis, preservation of normal cortex and bone marrow, eccentric lobular nonspecific soft tissue mass, and clinical presentations are helpful. Similar to the diagnosis of heterotopic ossification, CT is more sensitive to discern these features allowing an early specific diagnosis of BPOP.

REFERENCES