Orthopaedic Case of the Month

A 16-year-old Boy with a Recurrent Mass of the First Toe

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History and Physical Examination

A 16-year-old boy was referred to our outpatient clinic with a painless, recurrent mass on the metatarsophalangeal joint of his right first toe. He first noticed the swelling approximately 6 months earlier after experiencing minor trauma while playing soccer, which resulted in a hematoma affecting the first toe. Owing to increasing swelling he had surgery under local anesthesia 3 months later to remove the tumor. Histopathologic examination classified the lesion as osteochondroma. The mass continued to grow and when the patient was referred to us approximately 3 months after the first operation, the mass was nearly twice as large as before. The patient denied constitutional symptoms, such as fever, night sweats, recent weight loss, or fatigue. The patient's physical status was good. Aside from an operation for hexadactyly on the left foot, his medical and family histories were noncontributory.

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Physical examination revealed a well-defined, compact, fixed, and firm mass approximately 2.5×2 cm. There were no signs of local infection or lymphadenopathy. The scar from the previous operation appeared without hypertrophy or redness (Fig. 1).

Plane radiographs and MRI were performed (Figs. 2, 3).

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

Radiographs at presentation showed a surface-based, densely mineralized lesion along the proximal phalanx of the first toe without obvious intramedullary extension. There was no extrinsic erosion or periosteal reaction of the underlying bone (Fig. 2).

MRI showed a heterogeneous lesion. The tumor appeared predominantly isointense to muscle on T1 (Fig. 3A) and hyperintense on T2 with serpiginous areas of persistent low signal intensity centrally (Fig. 3B–C). The adjacent cortex of the phalanx was somewhat indistinct, but there was no evidence of medullary invasion. Small surrounding soft tissue edema was noted (Fig. 3).

Differential Diagnosis

Osteochondroma
Parosteal osteosarcoma
Turret exostosis
Bizarre parosteal osteochondromatous proliferation
(Nora lesion)
Florid reactive periostitis
Surface chondroma (periosteal chondroma).

An en bloc marginal excision was performed. Superficially, the pseudocapsule enclosing the tumor was not violated (Fig. 4A). Separation from the underlying bone could be performed easily (Fig. 4B), and resection included the periosteal tissue beneath the lesion and decortication of the underlying host bone (Fig. 4C).

Histologic evaluation was performed (Fig. 5).

Based on the clinical history, physical examination, radiographic images, and histologic examination, what is the diagnosis and how should this lesion be treated?

Histopathologic Interpretation

The resected specimen was hemispheric and 2 cm in greatest dimension (Fig. 5A). Macroscopically, the lesion

resembled an osteochondroma, including a cartilage cap and a stalk composed of bony trabeculae. Microscopically, three components (cartilage, bone, and spindle cells) were observed in differing amounts. The cartilage and bone interface showed irregular endochondral ossification, the bone trabeculae formed were characteristically stained blue with hematoxylin and eosin (“blue bone”) and showed prominent osteoblastic rimming. Cartilage revealed partially hypercellular with moderately enlarged nuclei and contained large chondrocytes. The spindle cells were uniform without any atypia. They were loosely arranged between the trabeculae. Mitotic figures were seen occasionally, but none was atypical (Fig. 5B–C). The margins were negative in the 10 examined sections.

Diagnosis

Bizarre parosteal osteochondromatous proliferation (Nora lesion).

Discussion and Treatment

In addition to the bizarre parosteal osteochondromatous proliferation (BPOP), the differential diagnosis based on clinical and radiographic data included benign (osteochondroma and surface chondroma) and malignant tumors (parosteal osteosarcoma), and reactive lesions (Turret exostosis, florid reactive periostitis). The patient’s medical history and the localization and typical imaging of the tumor strongly argued toward BPOP. As with the current patient, patients frequently report precedent trauma before they recognize a painless, growing mass, typically affecting the small bones of the hands or feet, requiring them to consult a physician. BPOPs show a striking tendency to

Fig. 1 The clinical appearance reveals a solid mass along the dorsolateral aspect of the first ray without overlying skin changes. It is fixed to the adjacent bone.

Fig. 2A–C (A) AP and (B) oblique plain radiographs taken before reoperation show a surface-based, densely mineralized lesion along the proximal phalanx of the first toe, measuring approximately 2×2 cm. The mass extends from the dorsolateral aspect of the phalanx without obvious intramedullary extension. There was no extrinsic erosion or periosteal reaction of the underlying bone. (C) An AP plain radiograph taken before the first operation shows the initial status: a slightly smaller lesion with identical appearance.

Fig. 3A–C (A) A coronal T1-weighted MR image shows a lesion with low signal intensity (white arrow), predominantly isointense to muscle. The bone marrow signal of the adjacent proximal phalanx (star) is normal. (B) On a sagittal image, a heterogeneous pattern is shown. The lesion (arrow) appears hyperintense with serpiginous areas of persistent low signal intensity centrally. (C) On the axial fat-saturated image, the adjacent cortex of the phalanx is somewhat indistinct (maybe owing to previous surgery), but there is no evidence of medullary invasion of the lesion (arrow). Small surrounding soft tissue edema is noted.

recur; the most important anamnestic clue in our patient may have been the short period to recurrence. On radiography, the tumor showed a surface-based, radiodense appearance without obvious intramedullary continuity, which would have been seminal for osteochondroma. There was no evidence of periosteal reaction pointing toward malignancy. On MRI, a clear delineation of the tumor could be drawn and alterations of the underlying bone were not evident, both underlining the benign appearance. Definitive diagnosis was established by histologic examination, showing the typical composition of cartilage, bone trabeculae—characteristically stained blue with hematoxylin (“blue bone”)—and a spindle cell containing stroma.

As with our patient, the lesion usually is mistaken for osteochondroma because of its surface location and cartilaginous component. Solitary osteochondromas are extremely uncommon in the small bones of the distal extremities [1]. In contrast to calcified masses attached to but not in continuity with the underlying cortex as in BPOPs, osteochondromas show a typical continuity with the medullary canal radiographically. The absence of such a communication has been singled out as a critical imaging feature for BPOP, although reports of corticomedullary

continuity to the underlying bone are sparse [19]. Histologically, the cartilaginous cap in osteochondromas is uniform and better organized; chondrocytes usually are arranged in parallel rows lacking atypia [1].

Surface chondromas (juxtacortical or periosteal chondromas) are cartilaginous tumors emanating from the periosteum of small bones. Corresponding to BPOPs, they most frequently are diagnosed in the hands and feet of patients younger than 30 years [2]. They typically show pressure erosions at the cortical surface of bone with remodeling of the cortex and overhanging osseous margins (“saucerization”) on plane radiographs, signs that never were observed with BPOPs. Sclerotic rims or thin cortical shells also have been described and alleviate distinction between these two lesions [24]. Histologically, surface chondromas lack the spindle cell stroma and the typical “blue bone” pathognomically for BPOPs [2].

A BPOP must be distinguished from a parosteal osteosarcoma, which rarely is found in the hands and feet [10, 21]. Parosteal osteosarcomas contain atypical mitotically active fibrocytic cells and variable amounts of osteoid within their abundant fibrous stroma. BPOPs never show atypia of the fibrocytes, although they contain small amounts of proliferative fibrous tissue [17]. The periphery

Fig. 4A–C Intraoperative photographs show the en bloc negative margin excision. **(A)** Superficially, a pseudocapsule encloses the tumor and is not violated. **(B)** Separation from the underlying bone can be performed easily by resecting the adjacent periosteum. **(C)** After removal of the tumor, decortication of the underlying host bone is performed.

Fig. 5A–C **(A)** Presenting with a cartilage cap and a stalk composed of bony trabeculae, the lesion resembles an osteochondroma macroscopically. **(B)** Microscopically, the tumor is composed of a cartilage cap (star), covered by a fibrous capsule superficially (thin arrows) and a bony stalk (Stain, hematoxylin and eosin; original magnification, $\times 50$). **(C)** The cartilage cap (star) is partially hypercellular with

moderately enlarged nuclei and contains large chondrocytes. The cartilage-bone interface shows irregular endochondral ossification. Bone trabeculae (arrow) are characteristically stained blue (blue bone). The spindle cells (triangle) of the stroma are loosely arranged between the trabeculae (Stain, hematoxylin and eosin; original magnification $\times 100$).

of parosteal osteosarcomas may contain bizarre cartilage intermixing with the surrounding soft tissue, which indicates malignancy and is missing in BPOPs [17].

Benign, nonneoplastic lesions such as florid reactive periostitis (reactive periostitis or periostitis ossificans) also may simulate BPOPs. Potentially present in the whole skeleton, the florid reactive periostitis frequently affects the bones of the hand and therefore corresponds to the main localization of BPOPs [7]. Turret exostosis is a dome-shaped parosteal bone proliferation located on the dorsal aspect of the phalanges. BPOPs, florid reactive periostitis, and turret exostosis were proposed to be part of the same lesion spectrum, often related to an initial stimulus such as trauma, that leads to hemorrhagic subperiosteal proliferation, which then matures [25]. The lesion may represent an intermediate state between florid reactive periostitis and turret exostosis. This hypothesis has been supported through case studies showing a relationship between antecedent trauma and development of Nora's lesion [11] and findings suggesting processes occurring in the cartilaginous cap of a BPOP are similar to those of enchondral ossification in the growth plate, indicating a BPOP may be a reparative process after periosteal injury [9]. However, chromosomal rearrangements found in BPOPs [26] and the seemingly specific balanced translocation t(1;17)(q32;q21) [6, 16], strongly suggest a BPOP is a neoplastic rather than a reactive process.

A BPOP is an uncommon mineralizing mesenchymal lesion that typically affects the surfaces of bones in the hands and feet, frequently the metacarpal and metatarsal bones or the proximal and middle phalanges [5, 12, 17]. The hands are four times more commonly affected than the feet; 50% of the lesions involve the proximal phalanges [23]. Although BPOP has a characteristic clinical and histologic appearance, it may be confused with other benign and malignant lesions.

BPOPs have a remarkable tendency to recur, almost representing a clinical feature of the lesion. Recurrence rates of 30% to 50% and a great potential for rerecurrence [5, 12, 17] are difficult to explain if BPOPs were reactive alterations. A large series (35 cases) of BPOPs by Nora et al. [17] reported the recurrences between 2 months and 2 years after surgery. Dhondt et al. [5] reported all recurrences were evident within 6 months postoperatively. Choi et al. [4] reported on a case of BPOP in combination with an associated fibrosarcoma, although it was unclear whether this was an incidental finding or a transformation. However, despite a high tendency to recur and a sometimes atypical histologic appearance, no clear malignant transformation, metastases, deaths, or associated systemic diseases have been described so far in patients with BPOPs [1, 3, 5–23, 25, 26].

A BPOP shows a benign periosteal proliferation. It normally affects small bones of the hand or the feet;

manifestations in other long bones are exceptional. The radiographic picture is typical and has a surface-based, radiodense lesion without obvious intramedullary extension. Histologically, the tumor is composed of cartilage, appearing partially hypercellular and containing large chondrocytes, characteristic blue-stained bone trabeculae (blue bone), and a uniform spindle cell stroma. Since Nora et al. established and described BPOPs as an independent diagnosis in 1983 [17], fewer than 100 cases have been reported in the literature. As the tumor frequently is mistaken for an osteochondroma, the estimated number of unreported cases might be much higher.

Excision is the recommended therapy of a symptomatic BPOP. Intralesional excision has great potential for local recurrence, but it preserves stability without decortication of the affected bone. En bloc negative margin excision including the pseudocapsule, the periosteal tissue beneath the lesion, and decortication of the underlying host bone appears beneficial in preventing local recurrence [13]. Wide resection could lead to segmental amputation because of the anatomic conditions in the long bones of the fingers and toes and cannot be recommended as first-line surgical treatment [7, 13].

Our patient was treated by en bloc negative margin excision. He now has been followed for 10 months and is free of recurrence.

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