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short report

Bizarre Parosteal Osteochondromatous Proliferation (Nora's Lesion): A report of the first case originating in soft tissue

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Abstract
Many tumors are unique to the organs from which they arise. Over the last 20 years, however, most tumors that were thought to be primary in soft tissues (derived from the primitive mesenchyme) and thought not to have counterparts in bone, were found to, in fact, rarely arise as unique lesions from bone. Some examples include synovial sarcoma, rhabdomyosarcoma and leiomyosarcoma, to name but three. We now have begun to see the reverse with lesions that were initially thought to be unique to bone arising in soft tissue. While this has been well reported with osteosarcoma and Ewing's sarcoma, it has never been reported with Bizarre Parosteal Osteochondromatous Proliferation (BPOP), also known as Nora's lesion. This study explores the first reported case of Nora's lesion originating in soft tissue. While BPOP was originally thought to be a reactive (non neoplastic) process, molecular genetics suggest it has a non-random unique molecular signature. Moreover, it is clonal, which many consider the ultimate indicator of a true neoplasm. In this study, clinical, histopathological, and radiological studies help in further clarification of this entity.

Introduction
Bizarre parosteal osteochondromatous proliferation (BPOP) mostly commonly presents as an exophytic outgrowth from the cortical surface of bones. It was described in 1983 by Nora and colleagues, resulting in the eponym, Nora's lesion. Since then, approximately 160 cases have been presented in the peer-reviewed literature with a wide age range peaking in the 4th decade (1). BPOP usually affects the metacarpal or metatarsal bones, or the proximal and middle phalanges; however, long bones, skull, maxilla and metatarsophalangeal sesamoid have been reported to be affected as well (2-4). The first line of treatment is generally surgical excision (5). The frequency of recurrence of this benign lesion as well as its clinical presentation may cause it to be mistaken for malignant processes (5-6). While BPOP was originally thought to be a reactive (non-neoplastic) process, molecular genetics suggest it has a non-random unique molecular signature (7). Moreover, it is clonal, which many consider the ultimate indicator of a true neoplasm.

There has been a trend building over the last 2 decades, to recognize tumors that were thought to arise primarily in soft tissues (derived from the primitive mesenchyme) to, in fact, rarely arise as unique lesions of bone. Examples include synovial sarcoma, rhabdomyosarcoma and leiomyosarcoma. While still exceedingly rare, the recognition of such primary bone lesions is now well accepted. Synovial sarcoma arising in bone for example, has been reported in three cases since its first report in 1997 (8). Even rarer is the recognition of the reverse, with lesions that were initially thought to be unique to bone now identified as having arisen in soft tissue. While this has been well documented with osteosarcoma and Ewing's sarcoma, it has never been reported for Bizarre Parosteal Osteochondromatous Proliferation. This study explores the first reported case of Nora's lesion originating in soft tissue.

Case Report
The patient, a 48 year old hypertensive, dyslipidemic African American woman, status post hysterectomy, was first referred to our institution because of coronary artery disease with unstable angina. After admission, acute myocardial infarction was ruled out and she underwent cardiac catheterization and right coronary artery percutaneous transluminal coronary angioplasty (PTCA) with stent placement in her right ventricular marginal branch coronary artery which initially showed 90% stenosis and 70% stenosis of the right ventricular marginal branch. The 90% RCA lesion was reduced with a stent to 0%. The right ventricular marginal stenosis was reduced to 10% utilizing a balloon. There were no complications. She did well but only for several months when she started having recurrent episodes of retrosternal chest pain with shortness of breath on minimal exertion. After a sudden increase in shortness of breath and pain radiating to the left arm, she went to her local emergency room. Again an acute infarction was excluded and she was transferred to our institution for left heart catheterization with coronary angiogram and left ventriculogram showing end stent restenosis of her RCA. This was dilated to 0% successfully. Several months later a third episode occurred with similar symptomatology and the addition of intermittent claudication in her right groin area and calf with walking but with no prior history of peripheral vascular disease. As before, the patient underwent left heart catheterization with coronary angiogram the same day. The left main stem artery was patent while the LAD and left circumflex demonstrated minor irregularities. Again, the RCA showed 90% restenosis and this stenosis was reduced from 90% to 20%.

Three years later she presented because of near complete incapacitation by pain in her right wrist progressing over a three month period and a mass on the lateral volar aspect of the ulna at the wrist joint identified on physical examination. It was very tender to touch resulting in lack of full supination, keeping her awake at night. Conventional radiography revealed calcification at the rim of a cystic appearing lesion adjacent to the distal ulna. MRI had both fluid and solid characteristics and was suspicious for contact with, and invasion of, the cortex of the ulna. Microbiologic cultures at the same time of excision were negative.

She had an uneventful recovery without sequela Seven years have now past following removal of her wrist lesion. She has had no problems related to the lesion or its excision.

Results

Radiographic Imaging

Best seen on CT was a 1.5 x 0.9 cm soft tissue mass in the volar aspect of the wrist immediately anterior and separate from the distal ulna at the level of the distal radial ulnar joint. The lesion was slightly cephalad and anterior to the pronator quadratus muscle. It demonstrated curvilinear ossification peripherally and proximally that was well defined while a more irregular calcified component was observed on its radial aspect (Figure 1).

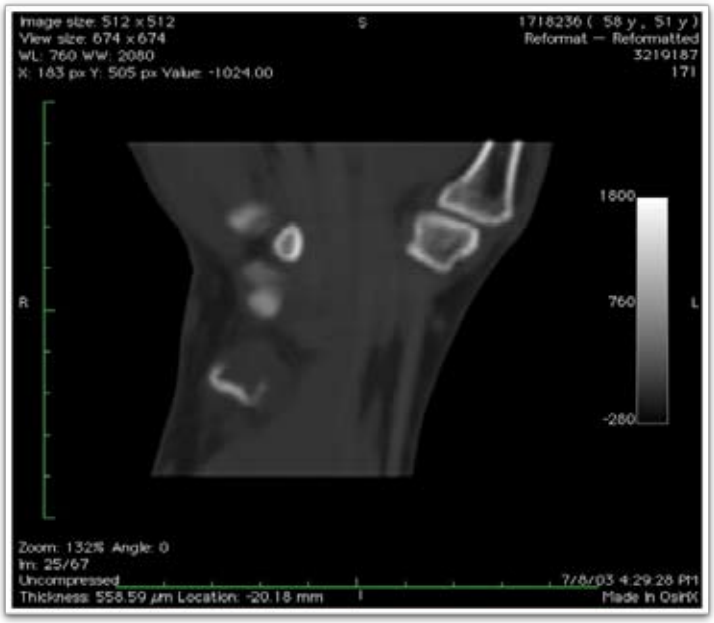

Figure 1. Coronal view

Pathology

On gross examination, the specimen was a glistening, tan-white, 2.0 x 1.7 x 1.5 cm portion of bony tissue that appeared to be partially covered by a glistening blue-gray cartilaginous cap. This cartilaginous cap measured up to 1.3 cm in maximal thickness. Upon sectioning, only minute portions of bone admixed with mottled tan-yellow cartilage was noted throughout the specimen.

The specimen was entirely submitted following fixation and decalcification. The lesion demonstrated interdigitation of fibroblastic, cartilaginous and osseous elements with focal atypia. At higher power, a transition from one tissue type to another was evident (chondroosseous metaplasia). Still higher power demonstrated large number of stromal cells with nuclear hyperplasia, hyperchromasia and contour membrane irregularities abutting bizarre fibroblastic cells, osteoclastic giant cells, osteoblasts and vascular elements (Figure 2). Other fields showed less worrisome zones of hyaline cartilage and fibrocartilage.

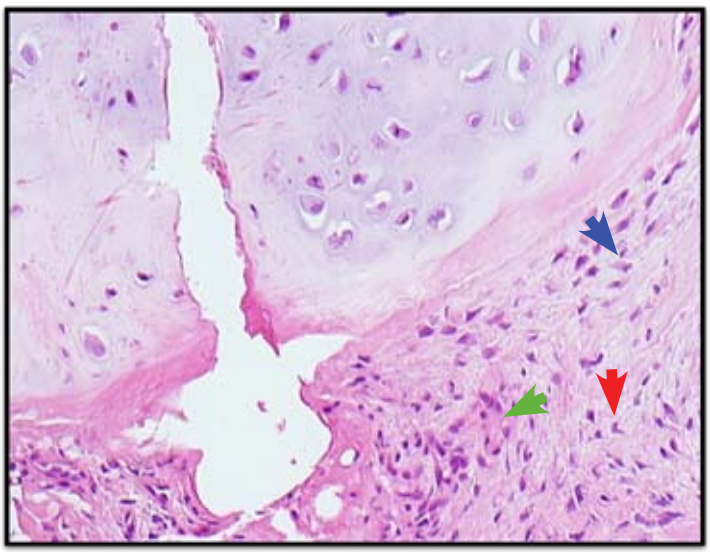

Figure 2. High power pictomicrograph (H & E 400x). Red arrow indicates stromal cells with hyperchromasia and nuclear contour abnormality. Blue arrow indicates bizarre fibroblastic cells. Green arrow indicates osteoclastic giant cell.

Discussion

Histologically, BPOB demonstrates well formed trabecular bone centrally surfaced by hyaline cartilage. The bone has blue tentorial properties by conventional H & E staing (blue bone). The cartilage is often lobulated and the connective tissue stroma hypervascular and admixed with atypical (bizarre) fibroblasts & mineralized bone mimicking sarcoma. This case fulfilled all these criteria and was considered a benign myxocartilagenous proliferation with non-zonal ossification separated by small amounts of bizarre spindled cells of fibroblastic lineage surrounding rare osteoclastic giant cells. No abnormal mitoses were appreciated and necrosis was not seen to be present. The differential diagnosis was broad and included malignant tumors such as osteosarcoma, reactive lesions such as periosteal chondroma and benign conditions including soft tissue chondroma. However, all were rejected because of clinical, radiologic or histologic features incompatible with those options leaving Nora's lesion the most reasonable possibility.

Conclusion

In conclusion, we, for the first time, report here that Nora's lesion can originate in soft tissue. While we explored here the histological and radiological demonstration of this uncommon entity, molecular genetics may help to further characterize the pathogenesis of this entity.

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