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Excision of a bizarre parosteal osteochondromatous proliferation ("Nora lesion") in the hand: A case report

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Abstract
Bizarre parosteal osteochondromatous proliferation is a benign osseous tumor mostly affecting the small bones of the hand and feet. Literature solely contains case series. Differential diagnosis has to be made with some other benign lesions but in particular with malignant tumors. Diagnosis is assumed upon a combination of suggestive clinical, radiological and morphological features, although histopathology is decisive in most cases. Awareness of this rare lesion is necessary for correct treatment and follow-up, because misjudging this pathology may lead to extensive resection and associated morbidity. Recurrence must be taken into account after surgical resection. This report describes a single case localized in the first webspace of the left hand without recurrence up until 24 months after surgical resection.

Keywords: Bizarre parosteal osteochondromatous proliferation, Nora's lesion, osteochondroma, bone tumor

Introduction
Bizarre parosteal osteochondromatous proliferation (BPOP), also called 'Nora lesion', is a rare benign osseous tumor typically affecting the small bones of hands and to a lesser extent the metatarsals and phalanges of the feet [1]. Cases are described in patients aged 3 to 73 years without sex predilection [1-5]. Typically, BPOP grows aggressively and has a high recurrence rate. Literature consists of small case series and isolated cases. Atypical histological appearance and aggressive features on imaging make its diagnosis difficult. We report a case of a NORA lesion in the first webspace of the left hand of a 47 year-old female.

Case
A 47 year-old female presented at the outpatient clinic with a progressively growing palpable mass in the left first web space since 2 months. She didn't experience any pain, however complained from discomfort due to a mass effect. There was no history of trauma. The lesion presented as a palpable, spherical nodule loosely connected within the thenar musculature without impaired sensibility or functionality of the thumb.
Plain radiography revealed an abnormal density near the base of the index metacarpal extending towards the cortex without cortical irregularity. Subsequent MRI-scan showed a largely calcified soft tissue mass of mixed intensity signal on T1 and T2-weighted images with ulnar and palmar deviation of the index flexors and flexor pollicis longus tendon respectively. It was surrounded by the first dorsal interosseous, opponens pollicis, oblique head of adductor pollicis and deep head of flexor pollicis brevis muscle. There was little marginal contrast uptake (Fig. 1). The diagnosis of a Nora lesion was suggested following tertiary referral of MRI images.
Under tourniquet and brachial plexus block, the patient underwent an excision of the lesion. We performed an incision at the thenar crease and approached the mass while carefully protecting median nerve branches and the superficial palmar arch. The interval between the thenar musculature and first dorsal interosseous was opened and the lesion with overlying pseudo-capsule could be resected without signs of periosteal attachment. Intraoperatively the lesion was located within the oblique muscle belly of the m. adductor pollicis. An in toto, nerve sparing resection was obtained (Fig. 2). There were no postoperative complications. Immediate mobilisation of the left hand was initiated. The patient reported neither pain nor local swelling during follow-up with a normal sensibility and mobility of the thumb. The patient is free from local recurrence after 24 months follow-up.

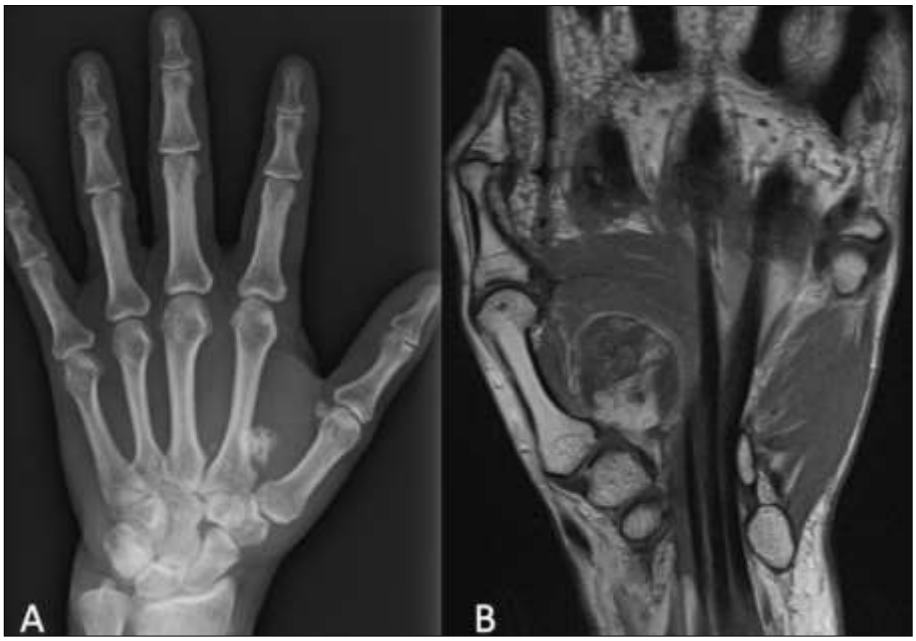

Fig 1: A. Plain radiograph demonstrates an abnormal density of 12 mm near the second metacarpal base without cortical irregularity. B. Coronal T1 weighted MR image visualises the mass with mixed intensity. It is well outlined and the surrounding soft tissues show no abnormality.

Histopathological analysis confirmed the diagnosis of a Nora lesion showing a mixture of fibrous tissue with bone, cartilage and spindle cells, each present in variable amount.

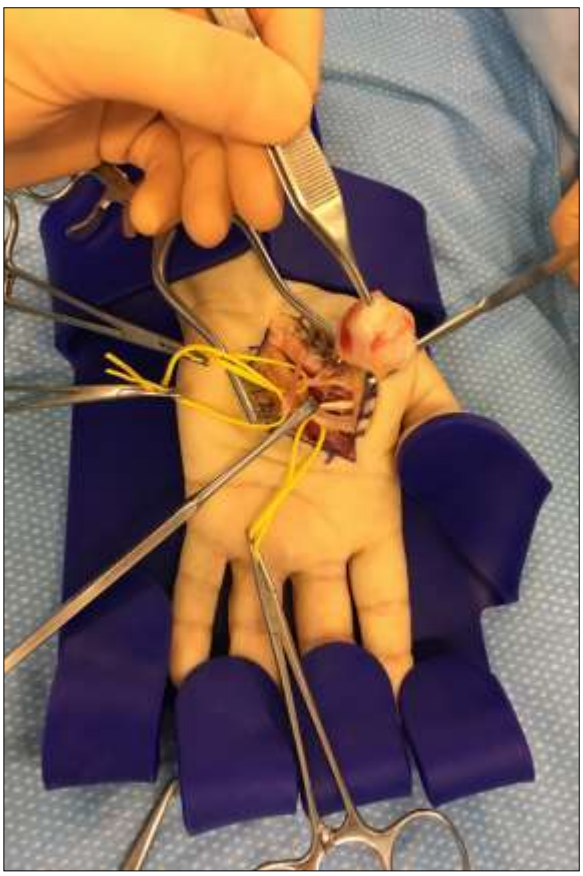

Fig 2: Intraoperative image shows the Nora lesion with pseudo-capsule at the palmar side of the second metacarpal bone after resection

Discussion

In this case we performed an in toto resection without exciding adjacent periosteal tissue or bone cortex. This case showed that in toto resection is a valid option in absence of malignancy in order to minimize morbidity and avoid local recurrence, although treatment and recurrence risk should be discussed with the patient prior to surgery [3]. Since our patient experienced discomfort due to the mass, we decided to perform a surgical excision. She was free of local recurrence after a follow-up period of 24 months. Recurrence rates range from around 50% in early case series to less than 30% in more recent articles but there are no reports of metastatic lesions [2, 3, 5].

Our approach is in contrast with Michelsen and colleagues who advocated excision of the pseudo capsule over the cartilage and any periosteal tissue or abnormal bone cortex in order to prevent recurrence even if there was a clear cleavage plane between lesion and host bone. In their series of 10 cases, they had a recurrence rate of 10% [5]. Berber and colleagues showed no statistically different recurrence rate in marginal or incomplete resection compared with complete resection, although the number of cases was small [3].

Nora and colleagues were the first in 1983 to describe 35 patients affecting metacarpals, metatarsals or phalanges, proximally being more affected than distally [1]. Meneses and colleagues added another 65 cases ten years later, in which 27% was localized in the long bones and 1 in the skull [2]. A more recent case series also confirmed around 23% involvement of long bones [3]. Later it has been reported in sesamoid bones, clavicle, zygoma and even in head and neck area [3, 6, 7]. Additional series are limited to a handful of cases.

Our patient suffered from a painless swelling, which is also documented in case series [3, 5]. Other presentations include pain, tenderness, limited range of motion or absence of any symptom. Although aetiology remains unclear, preceding trauma is often postulated as causative mechanism. This was observed in 30% of previous case series [2]. Furthermore, recent molecular investigation showed recurrent and unique translocations involving 1q32 in five cases of BPOP [8].

Diagnosis is made upon a combination of suggestive

clinical, radiological and morphological features, although histopathology is decisive in most cases. A biopsy may be considered as next step if there is a pre-operative concern about possible diagnosis. Differential diagnosis has to be made with some other benign lesions such as an osteochondroma although in the latter the cancellous bone of the stalk is continuous with the cancellous bone of the host bone near its base [9]. Parosteal osteogenic sarcoma should also be differentiated from BPOP but this malignancy is uncommon in hands and feet. Moreover, a suggestive history and location of the lesion in combination with the absence of soft tissue, cortical and medullary abnormalities may allow to distinguish it from subungual exostosis, stress fracture or florid reactive periostitis.

As particular features are suggestive on plain radiographs, Torregiani and colleagues showed that MRI is a useful guide in further characterizing the diagnosis [9]. MRI imaging of our patient showed partly increased T1 signal and variable signal on axial T2 image with fat saturation. The mass was well outlined without any soft tissue abnormality. Based upon these findings, a benign origin of the lesion was assumed. The radiological features of our case are not completely consistent with Torregiani and colleagues who reported decreased T1 signal intensity and high signal on T2 and STIR imaging in a case series of three patients, although these observations only reflect a phase during evolution [9]. A radiological spectrum going from periosteal thickening over a calcified periosteal mass lesion to an exostosis-like bone formation within one year has been postulated, confirming the assumption by pathologists made earlier [4]. This natural evolution identifies the lesion's distinct appearance and enables us to differentiate it from malignant masses and direct us towards a more conservative management.

Histologically, a Nora lesion is a mixture of fibrous tissue with bone, cartilage and spindle cells, each present in variable amount. A recent case series identified the presence of blue-staining osteocartilaginous tissue ('blue bone') as a helpful feature, since its absence may exclude this type of lesion from the differential diagnosis [10].

Conclusion

In conclusion, bizarre parosteal osteochondromatous proliferation is a benign condition that must be recognized and differentiated from malignant osseous lesions. MRI can be very useful in further characterizing the diagnosis which is usually based upon histopathology. Awareness of this rare lesion is necessary for correct treatment and follow-up. This case showed that in toto resection is a valid option in order to minimize morbidity and avoid local recurrence, although treatment and recurrence risk should be discussed with the patient.

Conflicts of interest statement

The author declare that there is no conflict of interests regarding the publication of this paper

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