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Bizarre Parosteal Osteochondromatous Proliferation (Nora Lesion) Mimicking Giant Cell Tumour of Tendon Sheath: A Case Report

Tsang Ming Kin Alan, MBChB and Lee Syn Yuk Michelle, MBChB, FRCSEd, FHKCOS, FHKAM*

Department of Orthopedics and Traumatology, Tseung Kwan O Hospital, 2 Po Ning Lane, Hang Hau, Tseung Kwan O, New Territories, Hong Kong SAR, China

Abstract

Nora lesion is a rare benign disease of parosteal osteochondromatous proliferation typically affecting the hand and feet, with notoriously high recurrence rates. Since its discovery, only few cases have been reported sporadically in the literature. No histological confirmed cases of this rare lesion have been reported yet in Hong Kong. We present a case of Nora lesion of the hand to demonstrate its clinical, radiological and histological features. Our case is highlighted by its diagnostic challenges pre-operatively as it mimicked a giant cell tumour of tendon sheath, as well as its correct identification intra-operatively followed by complete excision and decortication. There are no signs of recurrence detected at 1 year post-operatively.

Introduction

Bizarre parosteal osteochondromatous proliferation (BPOP), also known as Nora lesion, is a rare benign bony lesion affecting the extremities first described by Nora, et al. [1] in 1983. It often affects the small bones of the hands and feet, presenting as a parosteal mass. Although a benign entity, high recurrence rates close to 50% following surgical excision have been reported [2]. Since the time of its discovery, few cases have been reported as sporadic case reports or small series in the literature. No histological confirmed cases have been reported yet in Hong Kong. Here we present a case of Nora lesion of the hand in our locality to illustrate its clinical, radiological and histological features. Given the rarity of this lesion, diagnostic uncertainty pre-operatively was encountered as it mimicked a giant cell tumour of tendon sheath.

Case Presentation

A healthy 60-year-old Chinese gentleman presented to our clinic complaining of a right middle finger mass that has been gradually increased in size. The mass first became noticeable to him 2 years ago after a right middle finger sprain injury, followed by a brief period of pain at the proximal interphalangeal joint (PIPJ). Patient did not seek medical attention at that time as the symptoms were self-limiting. A vague prominence adjacent to the proximal phalanx was then noticeable to patient, slowly increased in size and causing restriction in finger motion. There was no numbness, skin discoloration, or constitutional symptoms.

Physical exam revealed a 2 cm × 1 cm firm nodular oval-shaped mass situated at the right middle finger proximal phalanx radial aspect without any skin changes. No wounds or surgical scars are present. PIPJ range of motion was limited by the mass with a resultant range of 5-60 degrees and tip to palm distance of 2 cm. Clinically the mass was close to and abutting onto the extensor tendon, causing reduced excursion. Dynamic tenodesis effect was preserved. The mass was non-tender, non-emptiable, and does not transilluminate. Tinel test and capillary refill test was normal. There are no other similar lesions in the hands and feet.

Plain radiographs showed soft tissue swelling and calcifications without obvious cortical and medullary connections (Figure 1). MRI showed the mass was hypointense on T1-weighted images and heterogeneous hyper intense on T2-weighted images, displacing the extensor tendon away from the phalanges (Figure 2). The reporting radiologist

***Corresponding author:** Tsang Ming Kin Alan, MBChB, Department of Orthopedics and Traumatology, Tseung Kwan O Hospital, 2 Po Ning Lane, Hang Hau, Tseung Kwan O, New Territories, Hong Kong SAR, China

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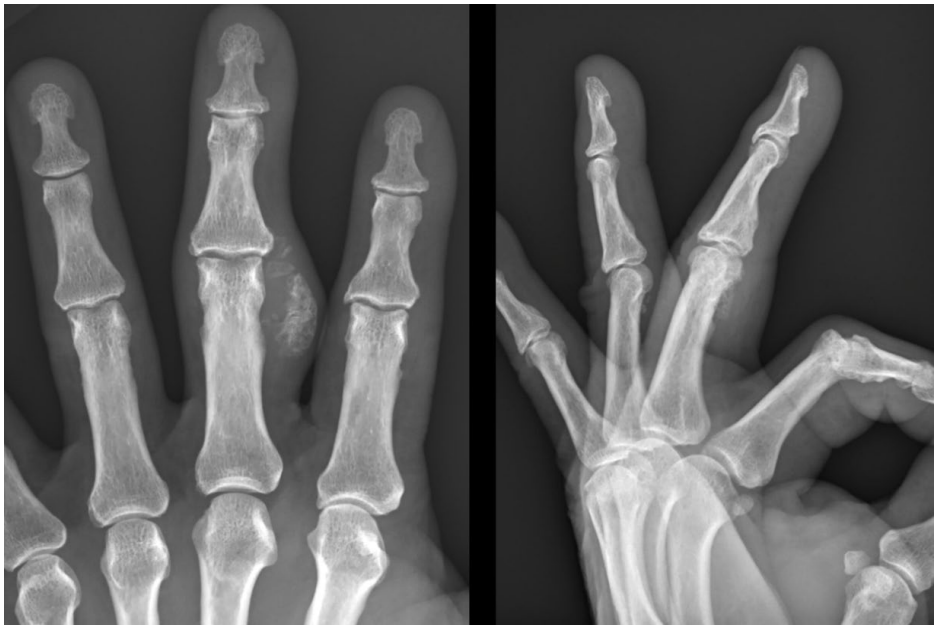

Figure 1: Plain radiograph in anteroposterior and lateral views of the right hand showing soft tissue swelling and calcifications adjacent to right middle finger proximal phalanx radial aspect. No obvious cortical and medullary connections are seen. Bony indentations noted at the proximal phalanx are suspected due to pressure erosion by the mass.

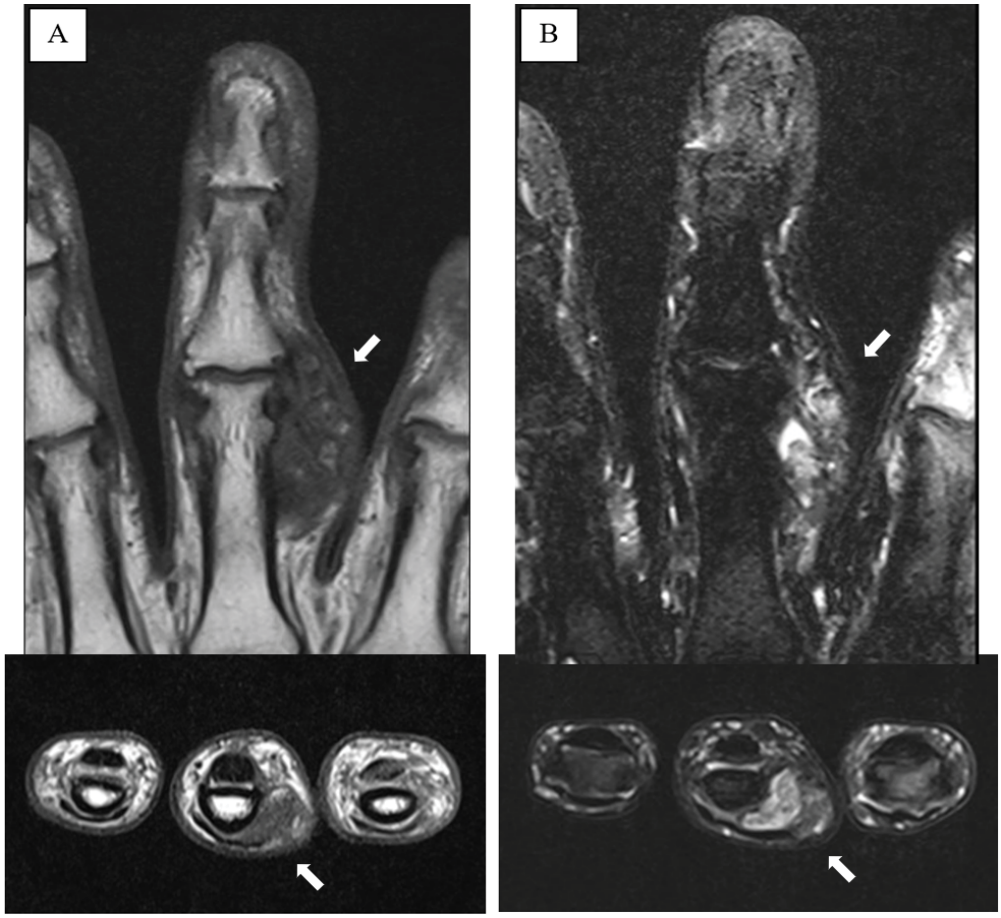

Figure 2: MRI images (coronal, axial) of an oval shaped soft tissue mass abutting the radial aspect of right middle finger proximal phalanx. A) T1-weighted images showed the mass was hypointense; B) T2-weighted images showed heterogeneous hyperintense signals. There is normal signal intensity of the cortex and bone marrow of the adjacent phalanges.

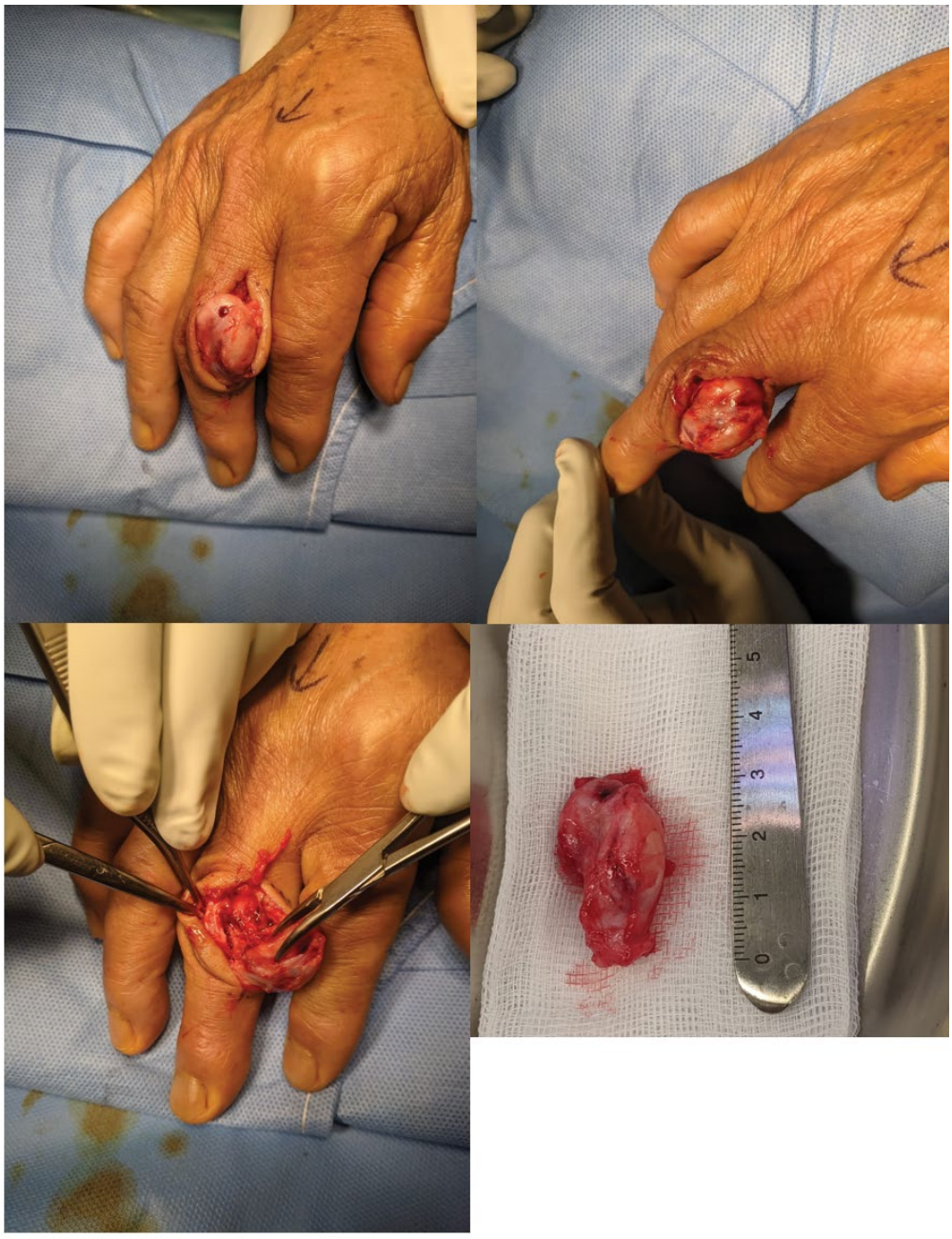

Figure 3: A 3 cm × 1 cm × 1 cm dome-shaped mass identified over right middle finger radial aspect, extending from middle to proximal phalanx. The encapsulated chondral mass was found in continuity with proximal phalanx cortical bone, displacing the extensor tendon ulnarly while having no connections to the tendon sheath.

also noted internal blooming foci within the mass that may be suggestive of haemosiderin deposition. At this juncture, the differentials included giant cell tumour of tendon sheath in view of clinical feature of an enlarging mass attached to tendon together with MRI signal characteristics. Other differentials included osteochondroma, although uncommon at this anatomical region.

We proceeded with excisional biopsy of the finger mass under general anesthesia via a mid-axial incision. A 3 cm × 1 cm × 1 cm parosteal encapsulated chondral mass in continuity with proximal phalanx cortical bone was identified (Figure 3). While the mass was noted to displace the extensor tendon to ulnar side, there were no connections of the mass to the tendon sheath. Given the intraoperative findings, BPOP is a more likely diagnosis. Complete excision of the mass with the capsule and periosteum was performed followed by decortication of the underlying cortical bone to reduce tumour recurrence. Histopathological analysis revealed typical features of BPOP (Figure 4), confirming its diagnosis. These features included a mixture of spindle cells, cartilage and bone tissue arranged in a bizarre pattern, and "blue bone" (trabeculae with basophilic staining pattern) was demonstrated.

Patient enjoyed an uneventful recovery post-operatively

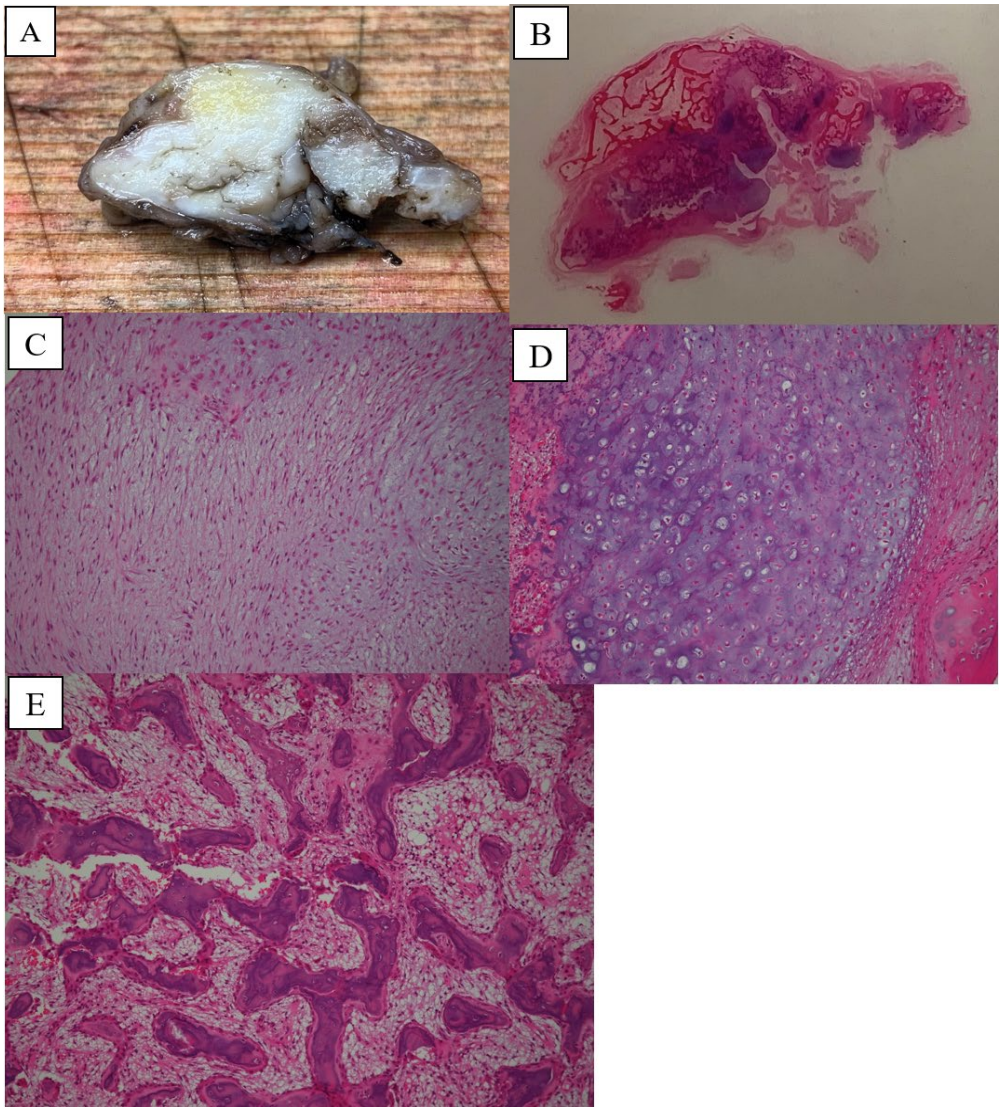

Figure 4: The lesion composed of a mixture of spindle cells, cartilage and bone tissue arranged in a bizarre pattern. (A) Cut surface of lesion; (B) Whole view of lesion on hematoxylin and eosin (H&E) stain; (C) Spindle cell area with myxoid stroma; (D) Chondroid area with hypercellular hyaline cartilage; (E) Osseous area with new bone displaying deep basophilia (blue bone).

and regained full range of PIPJ motion. There was no recurrence detected clinically and radio logically at 1 year after excision (Figure 5). Patient continues to follow-up with yearly monitoring.

Discussion

Nora lesion is a rare benign disease described as an exophytic mineralizing mesenchymal lesion, typically affecting the small bones of the hands and feet [3]. While the cause of BPOP remains uncertain, different hypotheses on its etiology have been described in the literature. Horiguchi, et al. proposed that BPOP may be a reparative process after periosteal trauma [4], while Zambrano, et al. cases of BPOP with chromosomal rearrangements suggested a neoplastic cause [5]. Our patient reported a history of trauma prior to the onset of mass at that region, resonating with trauma-related school of thought.

To date, no histological confirmed cases of this rare lesion have been reported in Hong Kong. The rare occurrence of this lesion along with its unusual clinical and radiological features contributed to a diagnostic challenge as illustrated in our case. Clinically, the mass mimicked giant cell tumour of tendon sheath in the hand, echoing with its rapid progression in size, a firm consistency, close proximity to and abutment onto the tendon, as well as its lack of transillumination. Being the second most common soft-tissue tumour in the hand (after ganglion) [6] also makes giant cell tumour of tendon sheath a higher and convenient differential. In plain radiographs, the mass showed intralesional calcifications with suspected pressure erosion to bone. Such features have also been reported in the literature for cases of giant cell tumour of tendon sheath [7]. Typical radiographical features of Nora lesion described in the literature include cortical bony growth projecting into the soft tissue while lacking connection with medullary cavity [2]. A lack of medullary involvement helps characterize BPOP from osteochondroma, its main differential. However in our patient, such radiographical

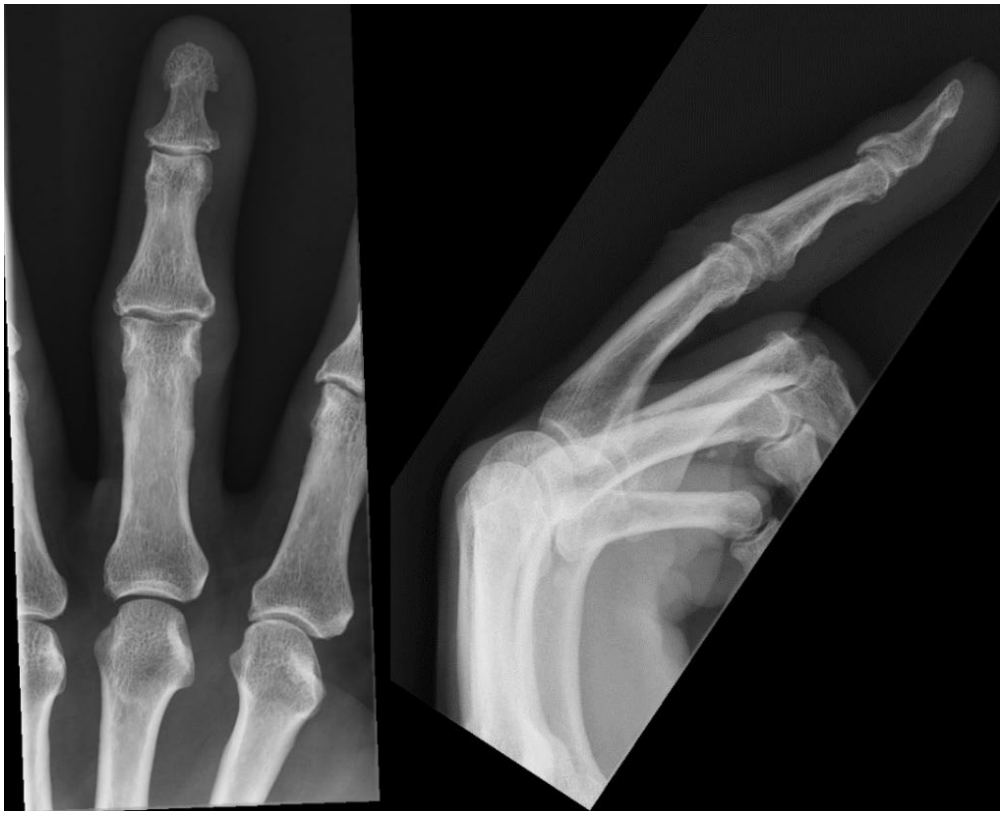

Figure 5: Plain radiograph in anteroposterior and lateral views of the right middle finger at 1 year postoperatively, showing no recurrence.

features are not obvious. MRI features of hypointense on T1-weighted and heterogeneous hyper intense on T2-weighted images demonstrated in our patient are in keeping with the classic descriptions of BPOP in the literature [3]. Interestingly, the reporting radiologist also noted internal blooming foci within the mass that may be suggestive of haemosiderin deposition, adding more speculation to giant cell tumour of tendon sheath as a differential.

Intraoperative findings revealed features of BPOP. A parosteal encapsulated chondral mass in continuity with cortical bone was noted. The mass was found to be different from giant cell tumour in that the colour it displayed was not typical of giant cell tumour (grey-white instead of orange-yellow tone), the consistency was firm to hard, it was just abutting but not extensively surrounding the extensor tendon, as well as its lack of multi-loculations. The suspicion of a chondral mass or NORA lesion was raised during the operation.

Nora lesion is notorious for having a high tendency to recur, with recurrence rates between 29% and 55% reported within 2 years after excision [1,2]. We proceeded with excision of the mass with the capsule and periosteal tissue beneath, followed by decortication of the underlying cortical bone via a small rongeur. This technique of en bloc negative margin excision followed by decortication is recommended to reduce recurrence rates [8,9]. Decorticating small areas of host bone does not jeopardize its stability. In a case series by Michelsen, et al. [9], 10 patients with Nora lesions in the hand had undergone surgical excision by this technique with a follow-up period of around 7 years. Only one had recurrence, occurring at 1 year after surgery. Our patient did not have recurrence detected clinically and radiologically during his latest follow-up at 1 year after excision.

Although a benign entity, Nora lesion can cause discomfort to patients, such as a rapidly increasing unpleasant mass, pain and restriction in motion in the extremities. Its unusual presentation that may mimic other differentials, and its rare occurrence especially in our locality makes correct identification difficult. However, Nora lesion is an important diagnosis to be kept in mind due to its high recurrence rate. Any chondral lesions that are firm to hard in consistency and attached to the bone should raise our clinical suspicion, no matter in the preoperative or intraoperative phase. Decortication should be performed in addition to complete excision. Further robust research is recommended to better understand its clinical behavior and pathogenesis.

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Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the publication of this article.

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