DOI: https://doi.org/10.30795/jfootankle.2024.v18.1784

Case Report

Bizarre parosteal osteochondromatous proliferative lesion of the hallux

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Abstract

Foot tumors are a rare occurrence, which can make diagnosis and management difficult. Certain benign tumors can be locally aggressive, with a rapid progression resembling a malignancy. We present a case of bizarre parosteal chondromatous proliferative (BPOP) lesion, also known as Nora lesion, arising from the sesamoid. This is a benign lesion with overlapping clinical and radiological features of a malignant neoplasm. Highlighting classic BPOP characteristics is crucial in avoiding potentially incorrect management and resulting morbidity.

Levels of evidence IV; Therapeutic studies; Case Report.

Keywords: Foot tumor; Hallux.

Introduction

Primary bone tumors account for 2% to 4% of all neoplasms. With regard to soft tissue tumors, 8% of benign tumors and 5% of malignant tumors occur in the foot and ankle[1].

Common benign soft tissue tumors found in the foot include plantar fibromatosis, ganglion cyst, giant cell tumor of tendon sheath, pigmented villonodular synovitis, and lipoma. More prevalent benign osseous lesions include osteoid osteoma, enchondroma, aneurysmal bone cyst, and chondroblastoma[1].

The possibility of any lesion being malignant should always be considered, as it has been shown that pain, lesion size, and symptom duration are not reliable in differentiating benign and malignant lesions[2]. Although malignant bone tumors are rare in the foot, osteosarcoma, Ewing sarcoma, and chondrosarcoma do occur[1]. Malignant melanoma is also fairly common in the foot, with potential life-threatening consequences if left untreated[3].

The low incidence of large soft tissue tumors in the foot makes the diagnosis and management of these lesions challenging. We present a rare case of bizarre parosteal osteochondromatous proliferation (BPOP) in the foot. This case study aid the reader in making this difficult diagnosis and managing the lesion appropriately. Informed consent was obtained from patient.

Case report

Clinical history

We present the case of a 56-year-old female patient who presented with a progressively slow growing mass under the left hallux. She reported a previous soft tissue injury to this left hallux three years prior. Wound healed uneventfully with local dressings. Eighteen months ago, she noticed a mass growing under the left hallux. The mass increased significantly in size over time, causing difficulties in wearing and walking in closed shoes. She consulted her local podiatrist, who recommended having the mass assessed by a surgeon. She reported no night pain.

Examination

Examination revealed a generally well Caucasian female. She walked on the lateral border of the left foot with supination of the forefoot to avoid pressure on the mass. Clinically, she had a 3 cm x 5 cm mass under the left first metatarsal head. The mass had a firm rubber-like consistency and was not adherent to the overlying soft tissue. There were no skin changes or increased local temperature. There was no tenderness on palpation. Sensation was altered over the plantar aspect of the hallux. Range of motion was limited in

Study performed at the Netcare Linksfield Hospital, Johannesburg, South Africa.

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How to cite this article: Siyo Z, Saragas NP, Ferrao PNF. Bizarre parosteal osteochondromatous proliferative tumor of the hallux. J Foot Ankle. 2024;18(2):255-60.

the metatarsophalangeal joint (MTPJ) due to impingement by the mass.

Special investigations

Radiographs of the foot showed a soft tissue lesion containing calcifications below the first metatarsal head (abutting the medial sesamoid), with no bony changes (Figure 1).

Magnetic resonance imaging (MRI) of the hallux showed a 56 mm x 27 mm x 30 mm heterogeneous septate mass with calcifications. Rim was calcified and enhanced with contrast (Figure 2). The mass enveloped the medial sesamoid and communicated with the MTPJ, displacing the flexor hallucis longus (FHL) medially. Differential diagnosis on MRI was tenosynovial giant cell tumor (calcification is rare in this type of tumor) and tumoral calcinosis (rare in the foot).

Computed tomography (CT) scan confirmed calcification within the mass. The mass enveloped the medial sesamoid but did not breach the cortex (Figure 3).

Management

Patient was consented for an excisional biopsy of the soft tissue mass. An L-shaped incision was made in the flexor crease of the first MTPJ, extending along the medial border of the lesion (Figure 4). Lesion was found to be well encapsulated and excised en bloc, while protecting the sensory nerves (Figure 5). The FHL was intact and not adherent to the lesion (Figure 6). The lesion seemed to originate from the medial sesamoid. The mass was sent for histology. Wound was closed in layers. Patient was mobilized in a heel weight-bearing wedge shoe. Stitches were removed at three weeks, by which time the wound had healed well. Patient was placed in supportive shoes at four weeks. At the 12-week follow-up, patient was back to normal shoe wearing and performing normal daily activities without any discomfort (Figure 7).

Histopathology

Histology reported a 49 mm x 35 mm x 20 mm specimen, showing a well-circumscribed, multilobulated lesion (Figure 8). Within the lobules, there was irregular and immature cartilage with enlarged bizarre binucleated chondrocytes. Chondro-osteoid tissue stained characteristically blue using hematoxylin and eosin stain (H&E), with enlarged bizarre binucleated chondrocytes. There was spindle cell proliferation between the bone trabeculae (Figure 9). Diagnosis of BPOP was reached. Specimen was reported to have clear margins.

Discussion

Bizarre parosteal osteochondromatous proliferation is a rare benign, non-infiltrative bone surface lesion. It was originally described by Nora et al.[4] in 1983, hence the term Nora lesion. This type of lesion is mostly found in tubular bones of the hands and feet, with a 59% to 27% distribution respectively[4]. Infrequently, they occur in long bones and the skull. Atypical presentations have been reported in the calcaneus and talus[5]. These lesions commonly occur in middle-aged patients, but have been reported in patients ranging from 12 to 81 years of age, with no specific gender predilection[6]. Its

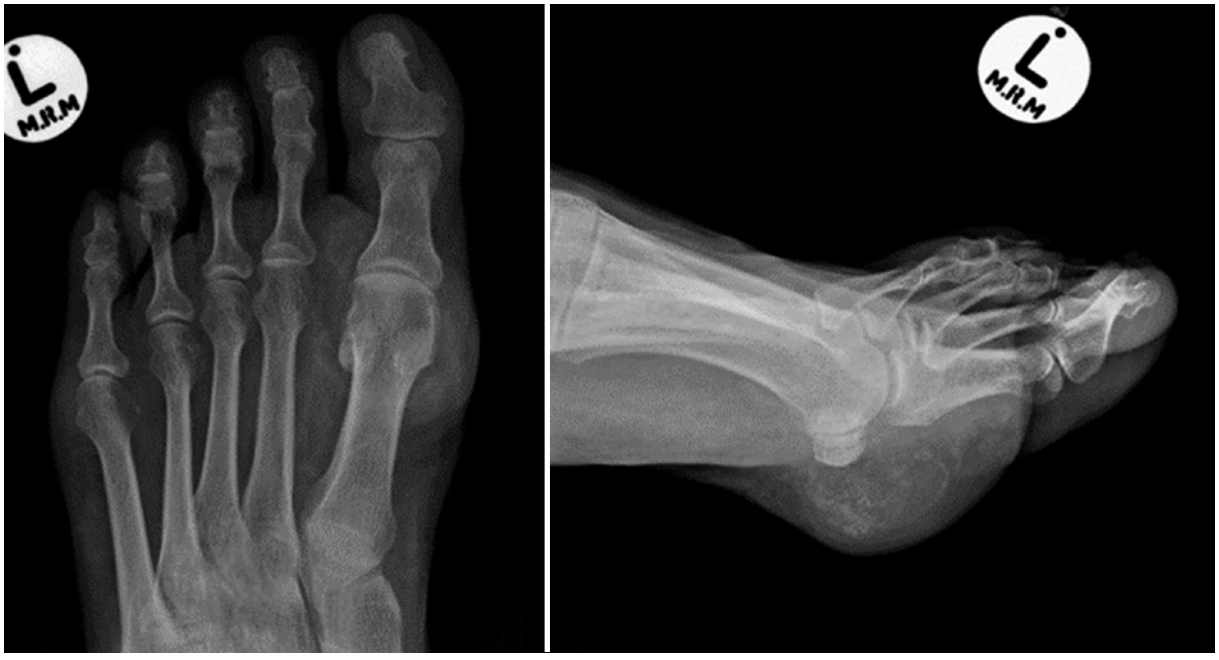

Figure 1. Antero-posterior and lateral radiographs showing the soft tissue mass with calcifications below the first metatarsal head.

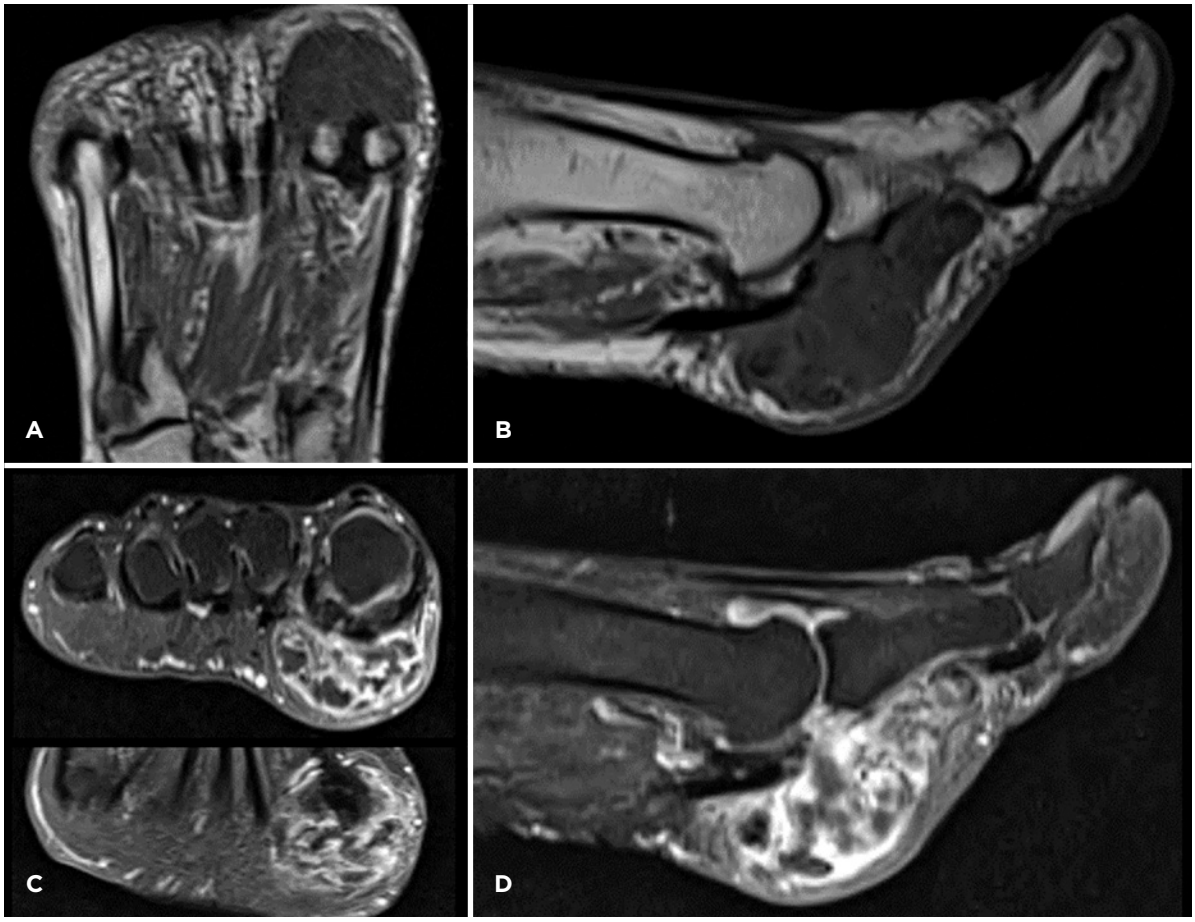

Figure 2. (A) T1-weighted axial MRI image showing a mass with uniformly low signal. (B) T1-weighted sagittal MRI image showing a mass with uniformly low signal. (C) Fat-saturation with contrast axial MRI image showing increased signal with calcification and rim enhancement. (D) Fat-saturation with contrast sagittal MRI image showing increased signal with calcification and rim enhancement.

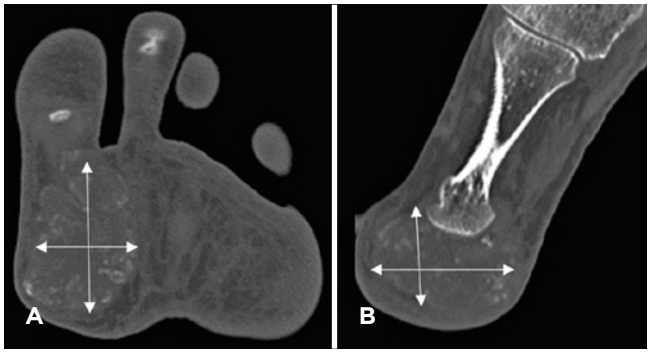

Figure 3. (A) CT scan axial image showing calcification within the lesion. (B) CT scan sagittal image showing no cortical breach of the metatarsal head by the lesion.

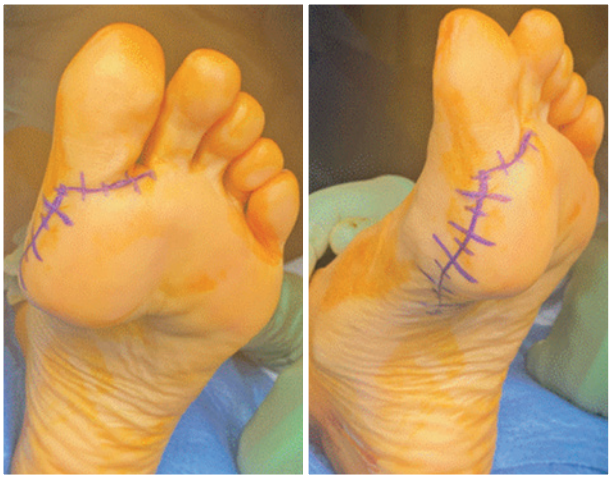

Figure 4. Lesion was approached using an L-shaped incision in the flexor crease of the first MTPJ extending along the medial border of the lesion.

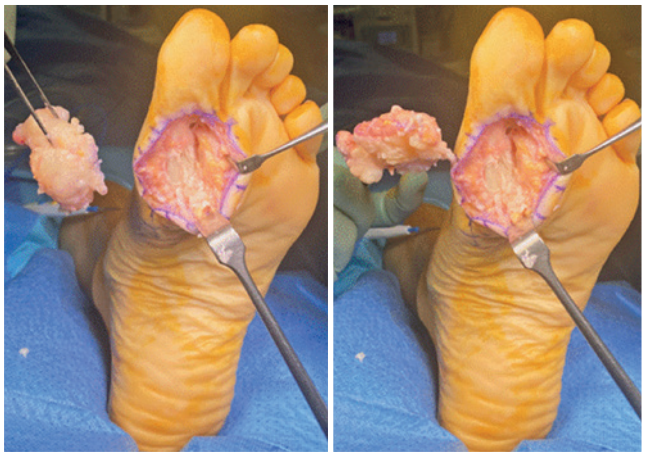

Figure 5. Lesion was found to be well encapsulated, being excised en bloc.

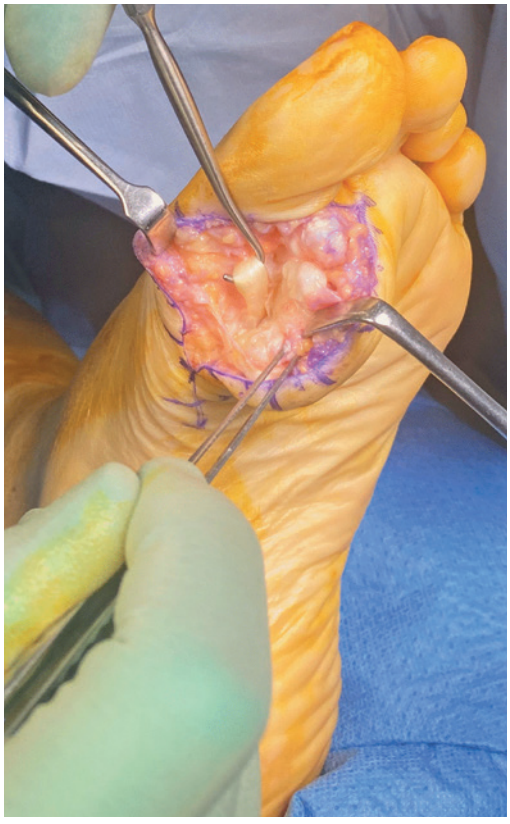

Figure 6. The FHL was intact and not adherent to the lesion.

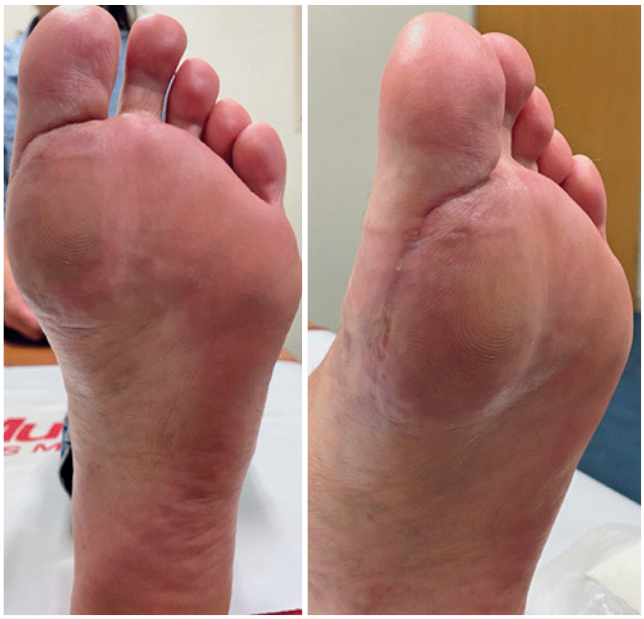

Figure 7. Wound had healed well in six weeks, with no painful scaring.

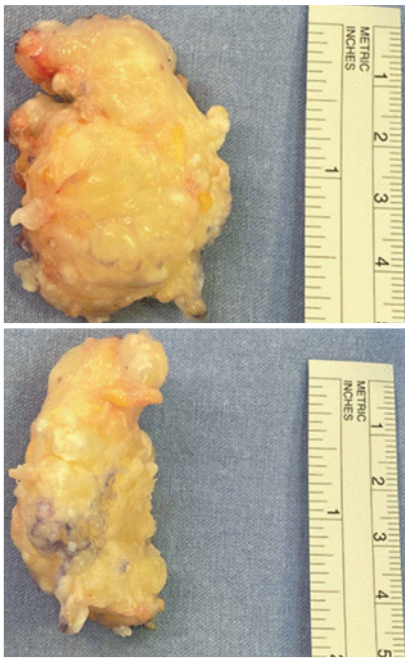

Figure 8. Macroscopic image showing a well-circumscribed, multilobulated lesion.

features resemble a malignancy, such as rapid growth, high rate of recurrence, and an atypical histological appearance. No case of malignant transformation has been reported, but these lesions can become aggressive and locally invasive.

Bizarre parosteal osteochondromatous proliferation can either be a slow or rapidly growing lesion which is inherently not painful. Overlying soft tissue is generally not affected by the lesion. As presented in our case, patient had no tenderness

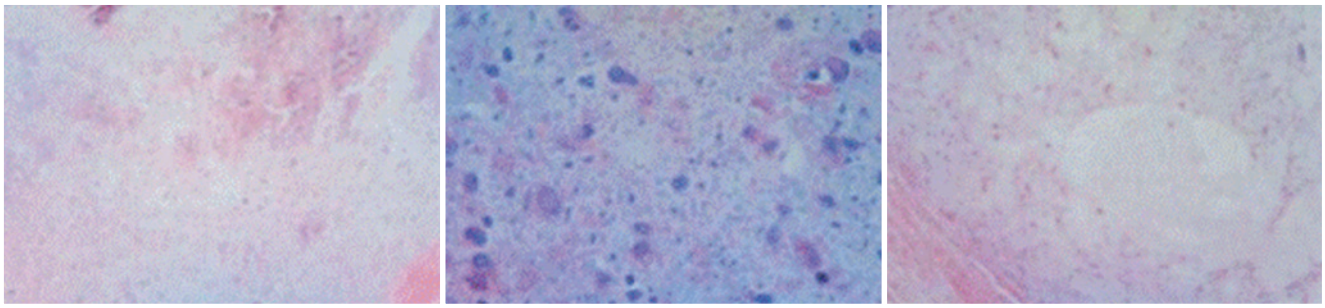

Figure 9. Histological images showing a chondro-osteoid tissue which stained characteristically blue using hematoxylin and eosin stain. There is a spindle cell proliferation between the bone trabeculae and enlarged bizarre binucleated chondrocytes.

over the lesion but struggled with activity and shoe-wearing problems related to the increasing lesion size. There is no specific known etiology, but previous traumatic periosteal damage has been suggested to be a predisposing factor, as seen by histological similarities to callus and subungual exostosis[4]. Edoardo et al.[7] suggested that these lesions have a multifactorial pathogenesis, with trauma being a high-risk factor[7]. Our patient reported a significant soft tissue injury to her left hallux prior to developing the lesion. Nilsson et al.[8] detected a translocation between chromosome 21 and 32 t (q32;21) in five cases, and also found balanced translocation between chromosome 1 and 17 t (1;17) in four of five cases, indicating a genetic predisposition[8]. Similar findings of balanced translocation of chromosomes t (1:17) and q (32;21) in BPOP cases have been identified by other authors[9].

Radiological features of BPOP have close resemblance to those of bone reactive lesions and benign neoplasms such as florid reactive periostitis, myositis ossificans, osteochondroma, and periosteal chondroma[10]. Malignant lesions like parosteal osteosarcoma and chondrosarcoma are part of the differential diagnosis in cases of rapid growing BPOP lesions. Plain radiographs show a classic well-circumscribed, mineralized lesion that abuts the cortex of the neighboring bone. This radiographic feature differentiates a BPOP lesion from myositis ossificans, which has characteristic islands of calcification not arising from the adjacent bone. The CT scan of BPOP lesions clearly demonstrates the absence of medullary communication between the adjacent bone and the lesion, which distinguishes it from an osteochondroma. Lack of cortical erosion and periosteal reaction differentiates this lesion from malignant tumors[11]. Current case's CT scan showed the lesion originating from the medial sesamoid, but not compromising the cortex. Typical BPOP MRI findings are that of uniformly low signal in T1-weighted images, while on T2-weighted and STIR images the lesion has intermediate signal in the center, with high signal intensity around the periphery[6,11].

Histological confirmation must always be attained when managing tumors. Microscopic features of BPOP lesions are bizarre cartilage with hypercellular large binucleated chondrocytes blending with woven bony trabeculae on a fibrous background comprised of reactive-looking spindle cells. Thus, the lesion is a mixture of cartilaginous, osseus, and fibrous tissue. The research conducted by Cocks et al.[12] on the histological variability of 16 BPOP cases reported a characteristic finding of "blue bone" in all specimens. "Blue bone" has been defined as an unusual mineralized cartilaginous matrix. Presence of "blue bone" and significant fibrous tissue is highly suggestive of a BPOP lesion[12]. There is no presence of cellular atypia, which would be suggestive of malignancy[4].

Such BPOP lesions are best managed by performing a wide local excision with clear margins as definitive treatment. It is important to get clear margins, as these lesions have a local recurrence rate of 37.4%[7]. In a case series of 22 Nora lesion cases by Berber et al.[13], 21 patients had surgical excision and one patient had amputation of the toe due to the size and local infiltration of the lesion. Recurrence occurred in six patients (27.3%), with a mean time to recurrence of 49 months. Two of eight patients with clear margins on histology had recurrence (25%), compared to 4 of 14 patients with marginal or incomplete resection (28.6%)[13]. There is no mention in the available literature regarding the possible use of radiation therapy to try to prevent recurrence. There has been no recurrence in our case at one-year follow-up after management with local excision.

Atypical BPOP lesions have been reported. Lesions are considered as being atypical according to either their location or radiological appearance. Atypical locations include facial bones, spine, clavicle, radius, fibula, calcaneus, talus, and sesamoids. Atypical imaging features are erosion of the cortex by the lesion and extension into the medullary canal. These atypical BPOP lesions are difficult to differentiate from malignant tumors without histological confirmation[7,14].

In conclusion, BPOP is a rare benign lesion that mimics both benign and malignant processes. It can be differentiated from other tumors by its typical radiological and histological characteristics. This atypical presentation of a BPOP lesion arising from the sesamoid highlights the need for a high index of suspicion and good clinical acumen when dealing with this condition so as to avoid inappropriate and destructive management.

Authors' contributions: Each author contributed individually and significantly to the development of this article: ZS *(https://orcid.org/0000-0002-8420-9244) Wrote and reviewed the successive versions and participated in the reviewing process; NPS *(https://orcid.org/0000-0002-5566-7588) Wrote reviewed the successive versions and participated in the reviewing process; PNFF *(https://orcid.org/0000-0003-4639-0326) Conceived and planned the activities that led to the paper, wrote or reviewed the successive versions and participated in the reviewing process. All authors read and approved the final manuscript. *ORCID (Open Researcher and Contributor ID).

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