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Case Report

Aggressive atraumatic myositis ossificans in a toddler ☆,☆☆,★

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ABSTRACT

Myositis ossificans (MO) is a benign disorder where bone forms within muscles or other soft tissues. This condition usually follows trauma and is rare in pediatric patients. Here we present the case of a 2-year-old male who developed MO of his right elbow without obvious trauma to the area. Imaging of MO in the initial phase is highly unspecific and obtaining tissue samples through a biopsy can render misleading reports. In most cases MO is a self-limited process with complete resolution, however, some cases may present a diagnostic and therapeutic challenge.

Introduction

Myositis ossificans (MO) is a benign condition where bone heterotopically forms within muscles or other soft tissues such as tendons and ligaments [1]. MO is more common in young adults in the second to third decade of life and is rare in the pediatric population [2,3]. Males are affected more commonly (3:2) and its etiology is remains not clearly understood [4,5]. In most cases it occurs 4-12 weeks subsequent to a traumatic event, in other cases mechanical stress or minimal trauma can also be associated [6]. The most common location is the lower limb (73%) and within that region, the quadriceps muscle; areas prone to trauma such as the elbow and shoulder follow in frequency [6,7]. MO is classified according to its presentation into 3 different types: myositis (fibrous) ossificans progressiva, traumatic, and circumscripta without history of trauma [8].

MO undergoes 3 different stages, initial development (<4weeks), mid-stage (4-8 weeks) and finally mature stage (>8 weeks); features noted on imaging studies as well as histologic examination correlate with the distinctive phases [9–11].

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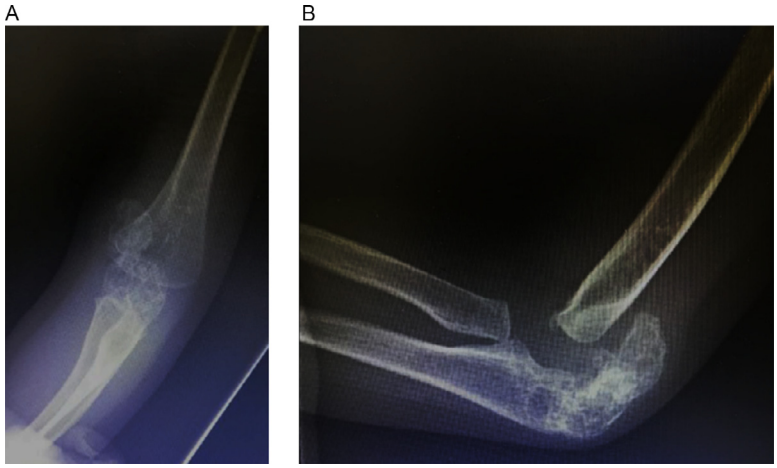

Fig. 1 – Right elbow AP (A) and lateral (B) radiograph, obtained at 2 years and 4 months of age, demonstrating an ossified mass protruding from the proximal aspect of the ulna.

On radiographic images, in the early phase a flocculent opacity with no ossification is initially noted, however, during the subsequent phase a peripheral ossification rim develops until it reaches its mature image of "eggshell calcification" in the final stage [10,12]. When present in tendons or ligaments, the shape of the MO follows that of the involved structure [12]. Obtaining a biopsy of the mass can be highly deceiving given the cells in the center of the lesion with high mitotic rate can be misinterpreted for a malignancy [13,14]. Furthermore, puncturing the lesion to obtain the sample can stimulate even more MO production and worsen the patient's condition [15]. Here we describe the case of a 2-year-old boy who developed MO around his right elbow without any obvious trauma and presented a diagnostic and therapeutic challenge for the multidisciplinary medical team.

Case report

A 2-year-old a male was brought for orthopedic consultation due to a complaint of inability to fully extend his right elbow that had started in the previous weeks without any prior trauma to the area. On examination, the patient did not have any swelling, redness or warmth in the area and was able to fully flex, pronate and supinate the right elbow, however, could only extend up to -30 degrees. Additionally, a hard enlarged area was palpated on the posterior distal aspect of the right elbow. Due to the patient's age, there was difficulty in assessing local tenderness. At the time radiographs were obtained which showed an ossified mass connecting to the proximal aspect of the right ulna (Fig. 1) and a decision was made to obtain a Tc99 bone scan (Fig. 2) and an MRI with and without contrast (Fig. 3) which reported an ossified mass of unclear etiology extending from the proximal aspect of the ulna into the triceps tendon and muscle and abnormal mild uptake in the bone scan. There is no medullary continuity between the solid ossified mass and the ulna to suspect an osteochondroma. Following, given contradictory differential diagnoses the patient underwent a percutaneous core needle imaging-guided biopsy which informed mature and immature bone tissue with osteoblast and intramedullary components noted, areas of calcification, proliferative small vessels and scattered inflammatory cells (Fig. 4). Unfortunately, zonal architecture was not demonstrated which may have been in part due to fragmentation of the sample (Fig. 4B). The slides were shared for a second opinion at an international high-volume center, with a congruent suggestion of a diagnosis of myositis ossificans. In the period of 2 months that followed the biopsy, the ossified area grew double its original size, and at that point the patient lost his ability to flex his right elbow as well maintaining a fixed position at 90 degrees of flexion (Fig. 5). A clinical international consultation was then pursued, and the outside team of orthopedists recommended resection of the ossified area to regain elbow range of motion. Prior to surgery the myositis ossificans was noted to have hastily grown once again, therefore, after a multidisciplinary discussion, a decision was made to observe and reschedule surgery at a later time once the lesion had stabilized (Fig. 6). The patient then underwent a period of 2 years of observation. Unfortunately, an MRI without contrast obtained at that point demonstrated that the lesion continued to be actively growing and was now extending up to the proximal triceps; surgery was once again cancelled (Fig. 7). Given the aggressive behavior and continuous growth of the lesion, the patient was then referred to a team of medical specialists to investigate genetic causes of heterotopic ossification. The patient underwent genetic testing for progressive ossifying fibrodysplasia and progressive osseous heteroplasia which returned negative. Following, the patient was put on a trial of oral corticosteroids by the medical team, which failed to produce any improvement. Additionally, the parents noted that the ossification process would stop to only resume once new minor trauma to the area had occurred. After exhausting many alternatives, now the patient is undergoing treatment with intravenous pamidronate, with mild improvement (Fig. 8).

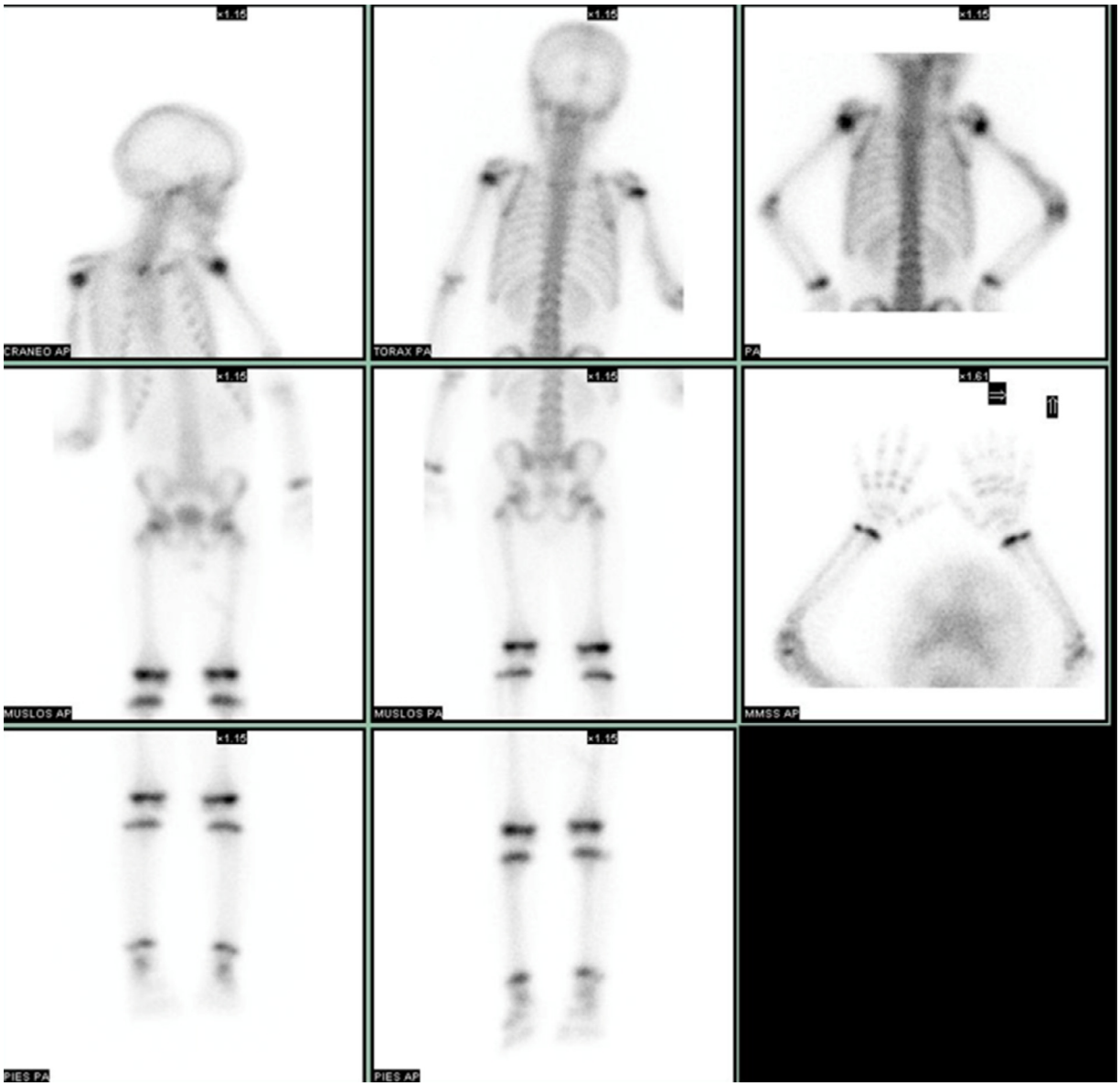

Fig. 2 – Tc99. Bone scan with Tc99, obtained at 2 years and 4 months of age, showing active lesion with abnormal mild uptake in the right elbow. No other abnormalities were noted.

Discussion

MO is a rare condition in children and infants with limited cases described in the literature [16]. When present MO manifests as a painful growing mass and limited range of motion if in proximity to a joint [5,17]. Besides trauma several other, less frequent, risk factors for heterotopic bone formation have been identified such as spinal cord and brain injuries, burns, hip replacement and fractures, especially acetabular ones [12]. Additionally, sporadic cases of MO have been linked to genetic mutations. Fibrodysplasia ossificans progressiva is an extremely rare condition, presenting as painful flares with bone formation in muscles, tendons and ligaments as well as congenital deformity of bones, caused by a mutation of the ACVR1 gene [18–20]. This condition carries a dark prognosis due to increasing immobility as a consequence of the uncontrolled heterotopic bone formation [21]. Progressive osseous heteroplasia is an additional disease characterized by heterotopic bone formation, initially subcutaneously with progression to deeper tissues, caused by a genetic mutation of the GNAS gene [22]. In the case presented, given the unusual age presentation, the lack of traumatic history and the aggressiveness of the bone formation, the patient was tested for both genetic alterations, with negative results.

Radiographic images of this disorder will vary depending on the MO stage. In initial stages unspecific images such as soft tissue opacities or even no finding can be encountered if taken too early in the development of the MO. As the MO progresses increased mineralization is noted progressing in a centripetal fashion, emphasizing the importance of serial images to diagnose this disorder [23]. More detail of the lesion

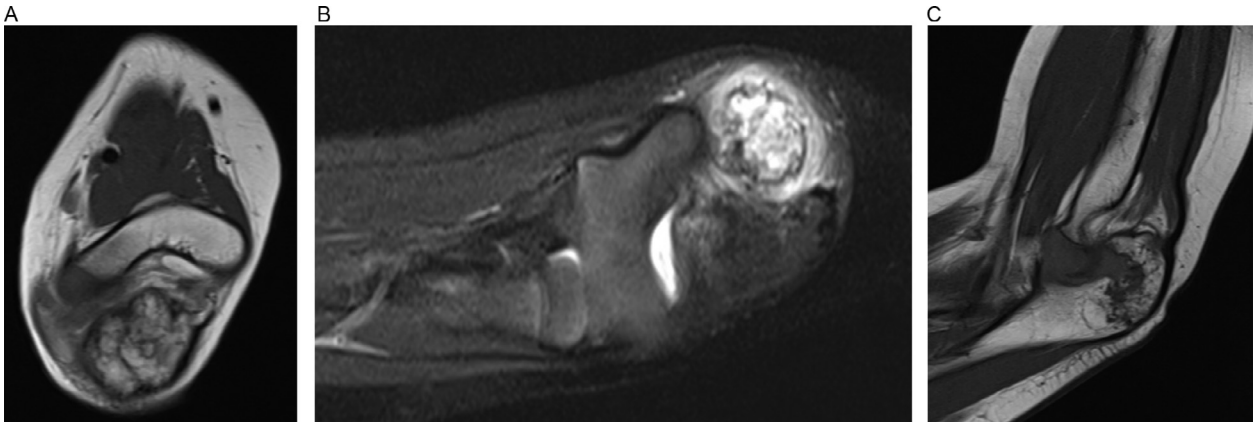

Fig. 3 – MRI without Gadolinium enhancement of the right elbow obtained at 2 years and 4 months of age. Axial (A), coronal (B), and sagittal (C) views showing an ossified mass with surrounding edema, extending from the proximal aspect of the ulna into the triceps muscle.

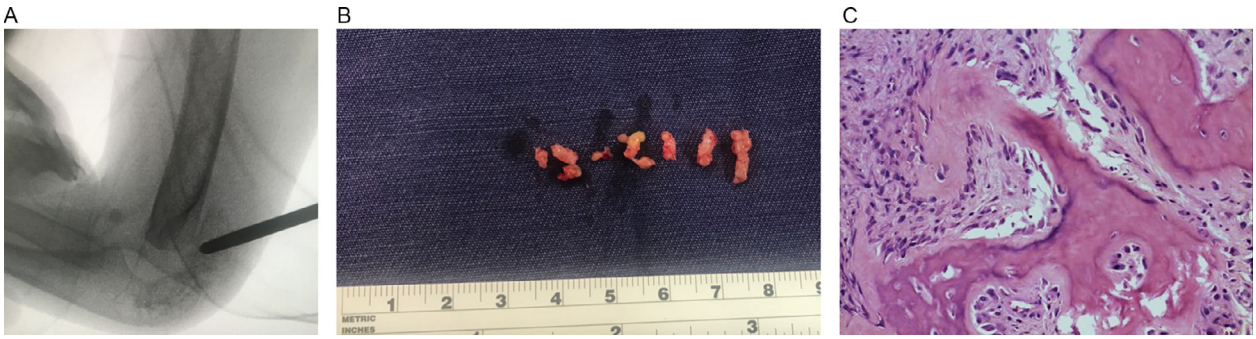

Fig. 4 – Images from the percutaneous imaging-guided biopsy, obtained at 2 years and 5 months of age. Fluoroscopic image (A) demonstrating the correct placement of the needle. Macroscopic images of the sample (B). Microscopic image H&E ×40 (C) showing immature bone tissue, small vessels and scattered inflammatory cells.

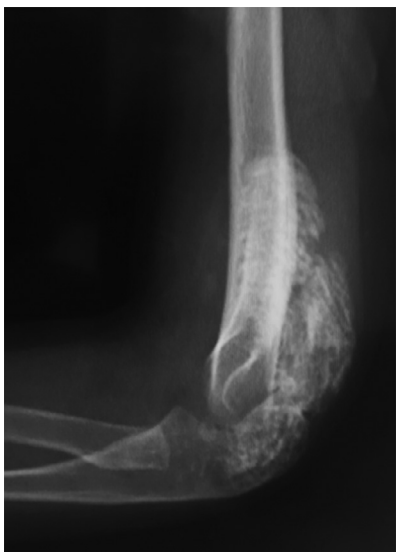

Fig. 5 – Lateral radiograph of the right elbow, at 2 years and 7 months of age, demonstrating increased in the ossification volume post-biopsy.

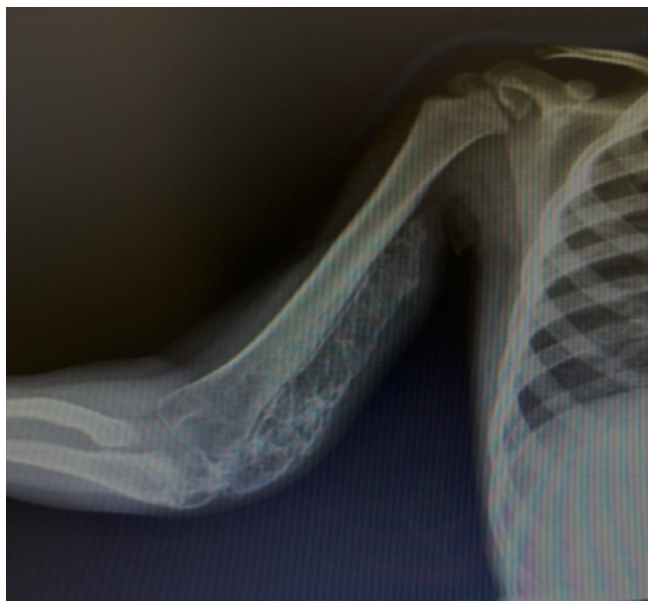

Fig. 6 – Radiograph of the right humerus, at 4 years and 8 months of age, demonstrating significant increase of the ossified area up to the proximal third of the triceps.

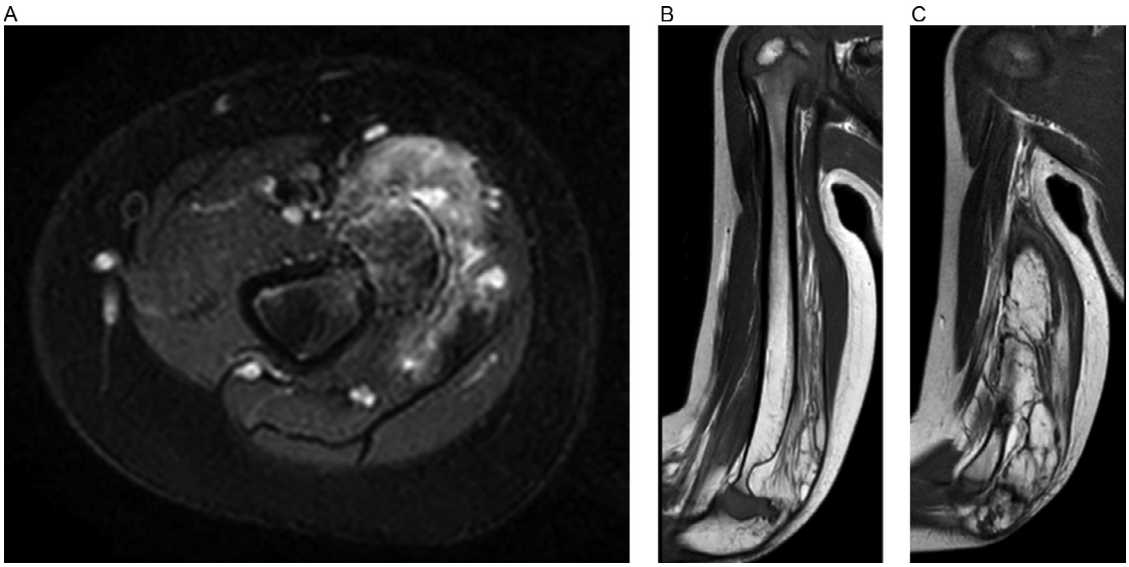

Fig. 7 – MRI without Gadolinium enhancement of the right elbow after 2 years of observation, at 4 years and 8 months of age. Axial stir (A) and 2 cuts of sagittal T1-weighted sequences (B, C) demonstrating the ossified mass replacing the fibers of the triceps muscle.

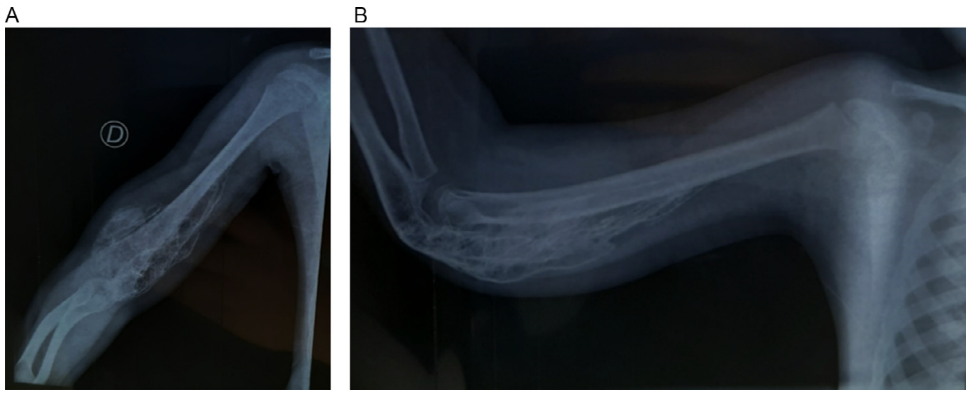

Fig. 8 – Radiographic images of the right elbow, AP (A) and lateral (B) views, obtained at the last follow-up at 5 years and 11 months of age, showing mild improvement of the myositis ossificans.

can be obtained with a CT scan which demonstrates a peripheral ossification zone with a radiolucent center and a "string sign" as lucency that separates the lesion from the adjacent bone [24]. Even though the "string sign" is often the rule, in longstanding cases the MO can adhere and fuse to the periosteum of the neighboring bone, either as a patch area or more broadly, as seen in our case [12,25]. Such circumstances warrant caution as to not being misinterpreted for a parosteal osteosarcoma. MRI images can add to the confusion if obtained too early in the development of the MO. Under MRI studies MO presents as a heterogeneous mass iso or hyperintense in T1 and hyperintense in T2-weighted sequences with surrounding edema [10,12]. On contrasted enhanced sequences a hyperintense rim, also called "zone phenomenon," can be observed, however, heterogeneous gadolinium enhancement is not infrequent, likewise advising caution as to being differentiated from soft tissue sarcomas [13,23]. In the presence of concerning signs, possibly indicating a more worrisome diagnosis, a biopsy could be warranted. For the tissue biopsy the sampling should involve an area from the center to the periphery to demonstrate the "zonal pattern," obtaining tissue solely from the center risks obscuring the diagnosis even more [5,26].

In most cases MO is a self-limited process with complete resolution, however, the genetic forms and aggressive or resistant cases can present a therapeutic challenge [14,15]. For uncomplicated MO, the majority of the patients can be managed successfully with observation and NSAIDs [26]. In situations where the MO remains painful, is restricting range of motion, causing a nerve impingement or there is a diagnostic conundrum, resection of the ossified mass is recommended [5,17]. Surgical removal of the MO should be performed completely and once the mass has shown features of mature ossification, otherwise risking recurrence [27]. In the case of our patient, his condition was still actively growing and more concerningly being exacerbated under minor trauma or even a needle

biopsy, therefore, a decision was made to maintain observation.

Conclusion

Myositis Ossificans (MO) is a benign condition where bone heterotopically forms within muscles or other soft tissues such as tendons and ligaments. This disorder is particularly rare in toddlers, and even though, in most patients can be limited process, more aggressive cases can present as a diagnostic and therapeutic challenge for the treating team.

Patient consent

Per the local institutional review board, consent was exempt due to this being the case of research involving the collection or study of existing data, documents, records, pathological specimens, or diagnostic specimen with the information being recorded by the investigator in such a manner that subjects cannot be identified, directly or through identifiers linked to the subjects. Nevertheless, the patient was informed and consented to publication.

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