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CASE REPORT
Reconstructive

A Rare Sequela of Constriction Band Syndrome: Case Report

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Summary: The patient in this case report is a 19-year-old man who presented with left foot cauliflower lesion. He complained of an inability to wear proper shoes, in addition to an unpleasant appearance of his foot. The lesion was present since his birth. Based on history and physical examination, the top 2 differential diagnoses at this stage were pediatric neurofibroma and constriction band syndrome (CBS). Laboratory investigations and x-ray were ordered for the patient. X-ray showed absence of most of the phalanges of the first, second, and third toes, with swelling of the overlying soft tissues of the foot. CBS was confirmed. Excision of the lesion was done along with skin graft applied on the area. Biopsy showed skin with dermal fibrosis and extensive adipose tissue infiltration without any sign of atypia or malignancy. The patient was discharged with regular follow-up appointments. *(Plast Reconstr Surg Glob Open 2021;9:e3564; doi: 10.1097/GOX.0000000000003564; Published online 10 May 2021.)*

Constriction band syndrome (CBS) is a relatively rare congenital anomaly that occurs when the fetal parts adhere to and become entrapped by a fibrous band of the amniotic membrane. Several terms have been used to describe this condition. However, constriction band or amniotic band syndromes are commonly used by experts. The incidence of CBS ranges from 1/1200 to 1/15,000 live births and 1/70 still births.[1,2] CBS has high variable presentations and mostly affects the distal portion of the extremities.[3,4] It might occur in any tissue or structure depending on the depth of the band and the site of constriction. Therefore, the specific management strategy must be individualized upon patient's functional and aesthetic needs.[5] In this article, we report a rare presentation of CBS as a cauliflower-like lesion affecting the forefoot that has not been previously described in the literature. It was managed surgically with good final aesthetic and functional outcomes. We aimed to shed light on the various presentations of CBS and provide surgeons a clue that might help in managing similar cases.

CASE PRESENTATION

A 19-year-old Nepalese man presented to our plastic surgery clinic with a left foot cauliflower lesion (Fig. 1). The lesion was present since birth. The patient's mother reported an uneventful pregnancy with no postpartum complications. The patient had no previous medical or surgical history and no previous admission to the hospital. His main complaints were an inability to wear proper shoes and ugly disfigurement of foot. On clinical examination, a cauliflower-shaped lesion with crypts was found, which had a foul smell due to difficulty in cleaning the area. Thus, skin secretions and normal flora were active in the depths of the crypts. The differential diagnoses that were considered at this stage were pediatric neurofibroma and CBS. Left foot x-ray was done, which showed no visualization of most of the phalanges of the first, second, and third toes that was associated with swelling of the overlying soft tissues of the foot (Fig. 2). The x-ray result along with the clinical assessment showed a clear demarcation line between normal skin and the malformation. Based on history, clinical examination and investigations, a diagnosis of CBS was confirmed. The malformation is a sequela of disruption of lymphatic drainage of the distal area to the band that causes localized lymphedema, which can lead to hyperkeratosis and growth of affected tissues. The patient was booked for surgical intervention. Excision of the lesion reaching the plane of plantar

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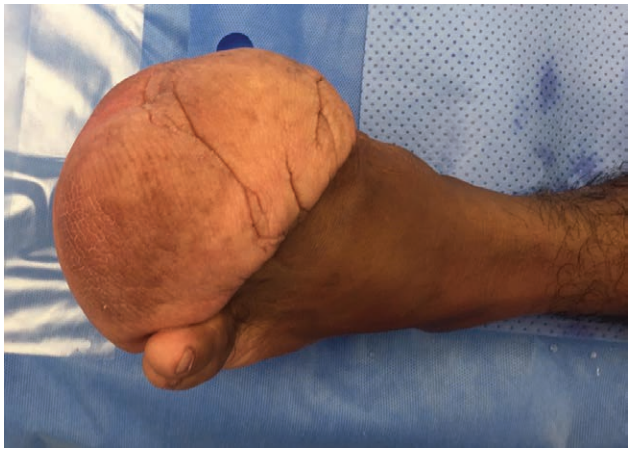

Fig. 1. Preoperative photograph of the patient's left foot showing the cauliflower-shaped lesion, with crypts found across the lesion.

fascia on the plantar surface and to healthy looking subcutaneous tissues (proximal to the band) on the front and dorsum of the foot was done to prevent any recurrence through removal of all lymph-containing soft tissues (Fig. 3). A split-thickness skin graft was applied in the area and fixed with staples. A biopsy was taken from the mass, which showed skin with dermal fibrosis and extensive adipose tissue infiltration without any sign of atypia or malignancy. The first dressing was done on the fifth day, followed by change of dressing on alternative days. After 3 months, an assessment was done, and the patient was discharged from the clinic. The patient reports a significant improvement in the quality of life and was satisfied with the outcome reached. He was asked to visit the clinic at 6 and 9 months postoperative to ensure the absence of any complications. During the follow-up visits, the patient was walking freely, using normal shoes, and reported satisfactory aesthetic outcome (Fig. 4). Assessment showed intact pain and temperature sensation, which is adequate to protect the foot from injuries. Touch sensation is better in the normal foot, as reported by the patient.

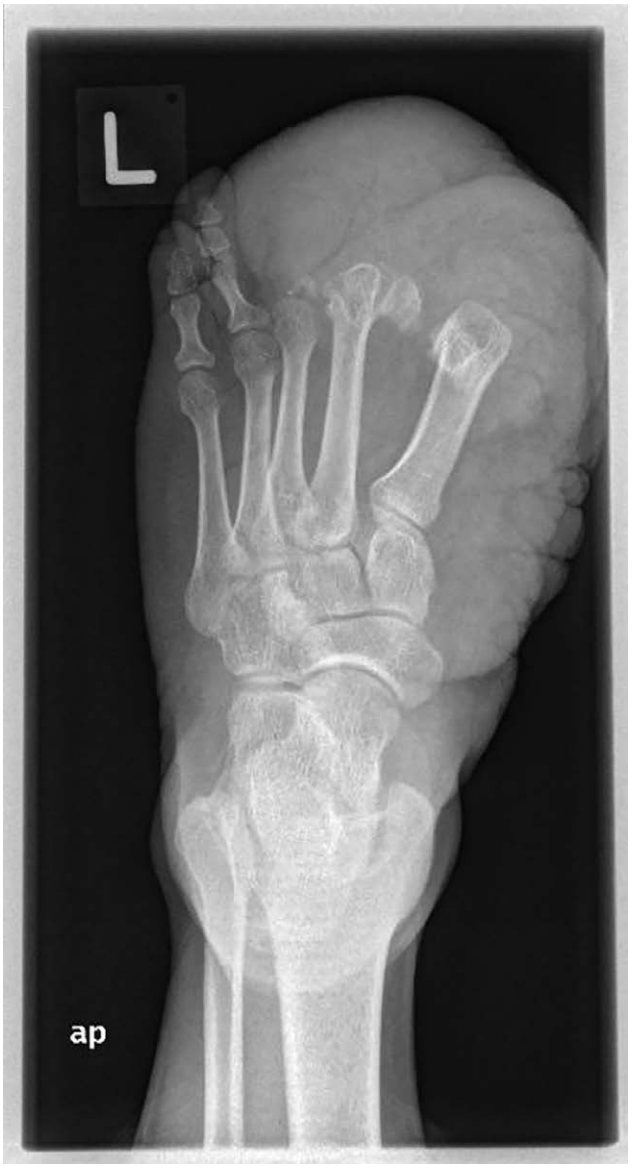

Fig. 2. X-ray image of the patient's left foot (anteroposterior view), showing the absence of most of the phalanges of the first, second, and third toes. Also, swelling of the overlying soft tissues of the foot is shown.

DISCUSSION

CBS is characterized by extremely varying presentation depending on the position of the band, which has been classified into 4 main categories, including constrictive rings, limb defects, neural defects (spine, brain), and craniofacial abnormalities.[6] The constriction rings can be limited to the skin and soft tissue or may invade deeply to obstruct the vascular and lymph supply, which can lead to malformations, amputations, and lymphedema at the side of the constriction of the affected limbs.[7,8] The prevalence of CBS Sequela is approximately 1.08 per 10,000 births.[1] The etiology of this syndrome remains unknown, and its occurrence is mainly sporadic without genetic or hereditary predisposition. Although there is no clear etiology, one theory suggests that early rupture of the amnion results in multiple loose strands. These strands can entangle the fetus parts and eventually lead to malformation and deformation. Another theory suggests that fetus blood flow compromise due to misoprostol exposure or chorionic villus sampling can explain internal visceral organ involvement. Moreover, genetic mutations are hypothesized to be the cause of the remaining CBS cases that were not well explained by the previously mentioned theories.[9]

Diagnosis of CBS can be established prenatally or after birth. Prenatally, it can be established by an ultrasound, which may detect the constriction bands, amputations, or lateralization of defects that are usually in the midline.[10] CBS present at birth can be diagnosed based on clinical features and appearance, which may suggest the best management plan. In regions where the medical service is not well developed or not approachable for all

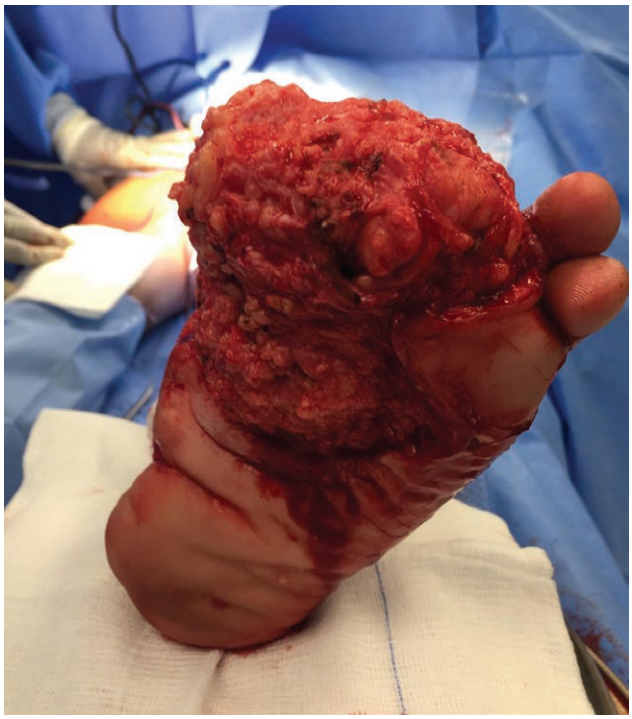

Fig. 3. Intraoperative photograph of the patient's left foot, showing the excision of the lesion reaching the plantar fascia on the plantar surface and the healthy-looking subcutaneous tissues (proximal to the band) on the frontal and dorsal surfaces.

of the population, a diagnosis and management may be delayed for years until the patient seeks proper medical advice, as described in this case report.

Although many sequences of this syndrome have been described previously, our case has a unique clinical presentation that has not been previously described in the literature and therefore requires a detailed report. This report, for the first time, describes a foot cauliflower lesion as a presentation of CBS Sequela. Therefore, this report will help surgeons to identify and manage similar cases in the future. Nevertheless, a multidisciplinary approach for management of CBS with a proper treatment plan must be tailored to each individual to optimize function and improve aesthetic appearance.

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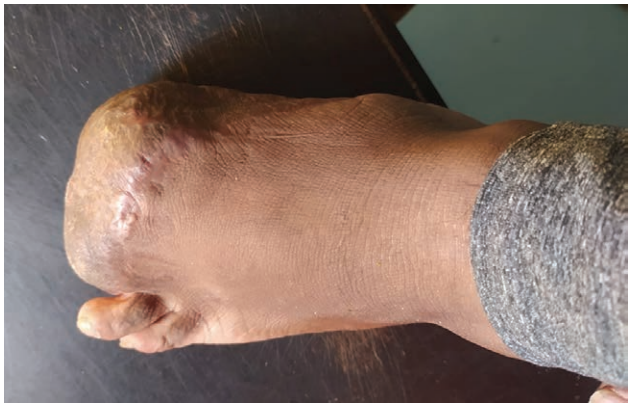

Fig. 4. Postoperative photograph of the patient's left foot, showing the final outcome of the reconstructed region.

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