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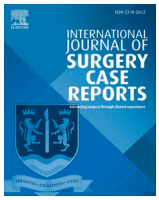

Case report

Presentation of polymelia in conjugation with spinal dysraphism: A case report

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ABSTRACT

Introduction and importance: Polymelia is a rare congenital deformity characterized by an extra limb connected to a different part of the body. The Incidence of this condition in humans is scant but relatively higher in animals. Hereditary and genetic factors are linked to the pathogenesis of this condition, but the relationship is not clearly understood. In this article, we highlight the presentation of polymelia and the approach used to treat the patient.
Case presentation: A 3-month-old male was brought to medical attention by his parents, who expressed concern regarding an extra limb attached to his back. He was born via spontaneous vaginal delivery after an uncomplicated pregnancy, with the only significant finding being the additional limb. After a thorough evaluation by a multidisciplinary team, he was diagnosed with Polymelia associated with spinal dysraphism. The child underwent surgery to excise the extra limb and to seal the defect in the vertebrae.
Clinical dissection: Polymelia is associated with multiple congenital malformations. Although it can be diagnosed in the antenatal period, most cases are identified after birth. The extra limb is usually functionless, but sensation may be preserved. It may be a well-developed limb or just a bud. A comprehensive evaluation is mandatory to explore the associated hidden malformation.
Conclusion: The expression of polymelia differs among individual case presentations, and the related congenital abnormalities pose significant management challenges. Surgical intervention is always essential, yet some complications are inevitable.

1. Introduction

Polymelia is a spectrum of rare congenital disorders characterized by accessory limbs and associated with several forms of congenital malformations. The occurrence of this condition in human remains is infrequent, leading to a reliance on case studies in the literature, many of which showcase exceptional traits. The supernumerary limb can be either a fully formed or partially developed limb, which may be comparable in size to a normal limb or manifest as a smaller appendage or bud. The exact causes of this rare condition are still not fully understood, although various theories have been proposed to explain certain facts [1,2].

When multiple congenital anomalies are present, the primary objective is to address any pathology that could jeopardize the child's life. In less severe cases, where significant deformities do not pose a health risk, the focus shifts to reducing the social stigma associated with atypical appearances. A multidisciplinary team is essential to customize the management plan based on the patient's needs and to implement rehabilitation strategies for any complication developed. Radiological evaluations play a crucial role in this process, offering detailed structural insights and assessing the risks associated with potential interventions. Surgical procedures are often necessary to rectify structural abnormalities while enhancing the child's appearance. In this article, we highlight the presentation of Polymelia and the approach used to reconstruct deformity [3,4].

2. Case presentation

A three-month-old male infant was brought by his parents with concerns regarding an accessory limb located on his lower back. He was delivered at 39 weeks of gestation at home through a spontaneous vaginal delivery, following an uncomplicated pregnancy. This child is the third in his family, and there is no history of similar conditions or skeletal anomalies among siblings. Following his birth, the infant cried promptly and showed no difficulties with breastfeeding or defecation. The patient's mother stated that she did not consume any toxins or drugs

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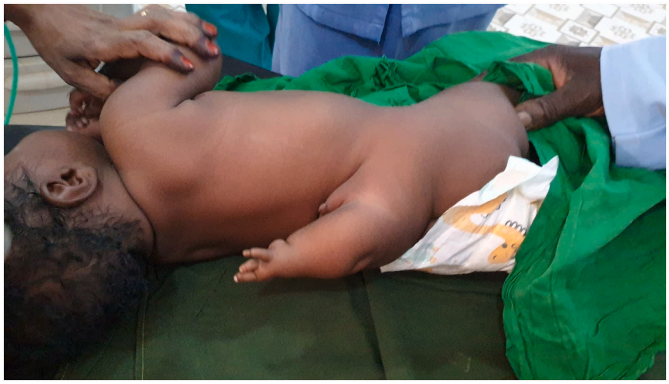

Fig. 1. Pre-operative: showing extra limb at the lower back of patient.

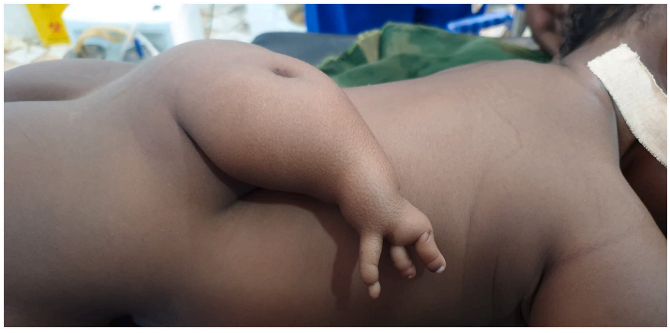

Fig. 2. Showing details of limb (toes with nails).

while pregnant and did not adhere to antenatal care guidelines. When the patient was 10 days old, his parents sought medical evaluation, during which a pediatric physician assessed him and determined that he was within the normal growth centile, showing no neurological or digestive abnormalities based on clinical examination and abdominal ultrasound. Also, he was assessed by an orthopedic surgeon who revealed no abnormalities apart from an extra limb in his lower back. X-ray showed a normal skeleton according to the patient's age, but there is tissue shadow at the back and bone not attached to the vertebral column. Then, he was brought for a plastic surgery consultation when he was 29 days age. Upon examination, the lower and upper limbs appeared well-developed and exhibited normal movement without deficits. The patient also demonstrated a typical response to pain stimuli. Notably, an accessory limb was observed in the lumbar region, oriented upward and laterally (Fig. 1) (Fig. 2) The proximal section of this limb contained a hard structure consistent with bone, while the distal end featured three toes. Surrounding the base of the limb was a bulging mass of soft tissue, accompanied by a skin tag at the proximal end. This accessory limb did not exhibit spontaneous movement or respond to stimulation, although the patient did react to painful stimuli. A CT scan was ordered for further evaluation due to the unavailability of an MRI in the city. The radiologists identified a multilevel loss of posterior elements at the L2, L3, L4, and L5 vertebrae. Additionally, soft tissue was observed at the sacral region, extending iso-dense soft tissue containing neural elements from the spinal canal through the posterior vertebral defect. These findings are indicative of spina bifida (lipomyelocele) (Fig. 3). In a multidisciplinary meeting involving a pediatric physician, cardiologist, anesthetist, neurologist, radiologist, and orthopedic and plastic surgeons, the decision was made to proceed with the excision of an extra limb and repair of the defect. However, the intervention was postponed until the patient reached three months of age. Preoperative assessments, including a complete blood count, urea and electrolytes, and coagulation profile, were all within normal limits. Also, blood grouping and saving were completed. In the operating room, following the administration of general anesthesia, the patient was placed in a prone position. A transverse incision was made along the previously marked line, just proximal to the base of the extra limb. The dissection proceeded to the base of the limb, which was found to be connected to the underlying adipose tissue by several fibrous bands. The limb was supplied by a single small artery, and the associated veins were ligated and dissected. Upon releasing the fibrous bands, the limb was freed, remaining attached by a distal cutaneous flap. Dissection continued through the adipose tissue until the dural sac was identified, at which point all fatty tissue was excised, and the neural elements were untethered from the surrounding structures (Fig. 4). The neural tissue was carefully retracted through the vertebral defect, after which bilateral lumbar fascial flaps were raised and inverted to effectively seal the defect. Any excess skin and additional limbs were excised. Following the achievement of hemostasis, a drain was inserted at the base of the wound. The wound was then closed in layers, leading to a transverse linear appearance (Fig. 5). The patient recovered from anesthesia without complications. Limbs movement ware normal post-operatively, and sensation was preserved. The recovery phase was uneventful, and the patient remains under consistent follow-up.

This work is under SCARE guideline [5].

3. Discussion

Polymelia is a rare congenital condition characterized by the presence of extra limbs in humans. Although it is uncommon, various literary sources have documented its different manifestations. The limited understanding of the underlying causes of this disorder complicates the comprehension of its pathogenesis and subsequent management strategies. Limb development occurs early in embryonic growth and is regulated by a variety of genes and molecular factors. Disruptions in the maternal environment due to factors such as trauma, infection, radiation, or exposure to toxins have been linked to limb deformities [6,7].

Polymelia frequently occurs alongside a variety of congenital abnormalities, including anorectal malformations, spinal dysraphism, and genitourinary anomalies. These related conditions can range from mild to critical, often requiring urgent surgical intervention during the neonatal phase. The classification of Polymelia is based on the attachment site on the body, which includes cephalomelia (attached to the head), thoracomelia (attached to the thorax), pygomelia (attached to the pelvis), and notomelia (attached to the vertebral column). Spinal dysraphism is a congenital defect of the neural tube that manifests in various forms, with approximately 14 case reports investigating its link to polymelia [8,9]. Saaiq M, et al. proposed a new classification system based on the characteristics of the supernumerary limb. This system categorizes accessory limbs into two main groups: polymelia associated with spinal dysraphism and polymelia not associated with spinal dysraphism. The first group is further divided into four subcategories: a. Well-developed extra limb, b. Moderately developed extra limb, c. Mildly developed limb, and d. Poorly developed extra limb. In contrast, polymelia without spinal dysraphism is classified into three categories: a. Well-developed limb, b. Moderately developed limb, and c. Poorly developed limb [10].

In this context, polymelia associated with spinal dysraphism presents several differential diagnoses, including teratoma, spinal hamartoma, parasitic twin, and disorganization-like syndrome. Various theories have been proposed to elucidate the pathogenesis of this condition; however, none have gained universal acceptance. Teratomas, which typically encompass all embryonic layers and can differentiate into various tissue types, are unable to develop organized mature limbs, rendering this

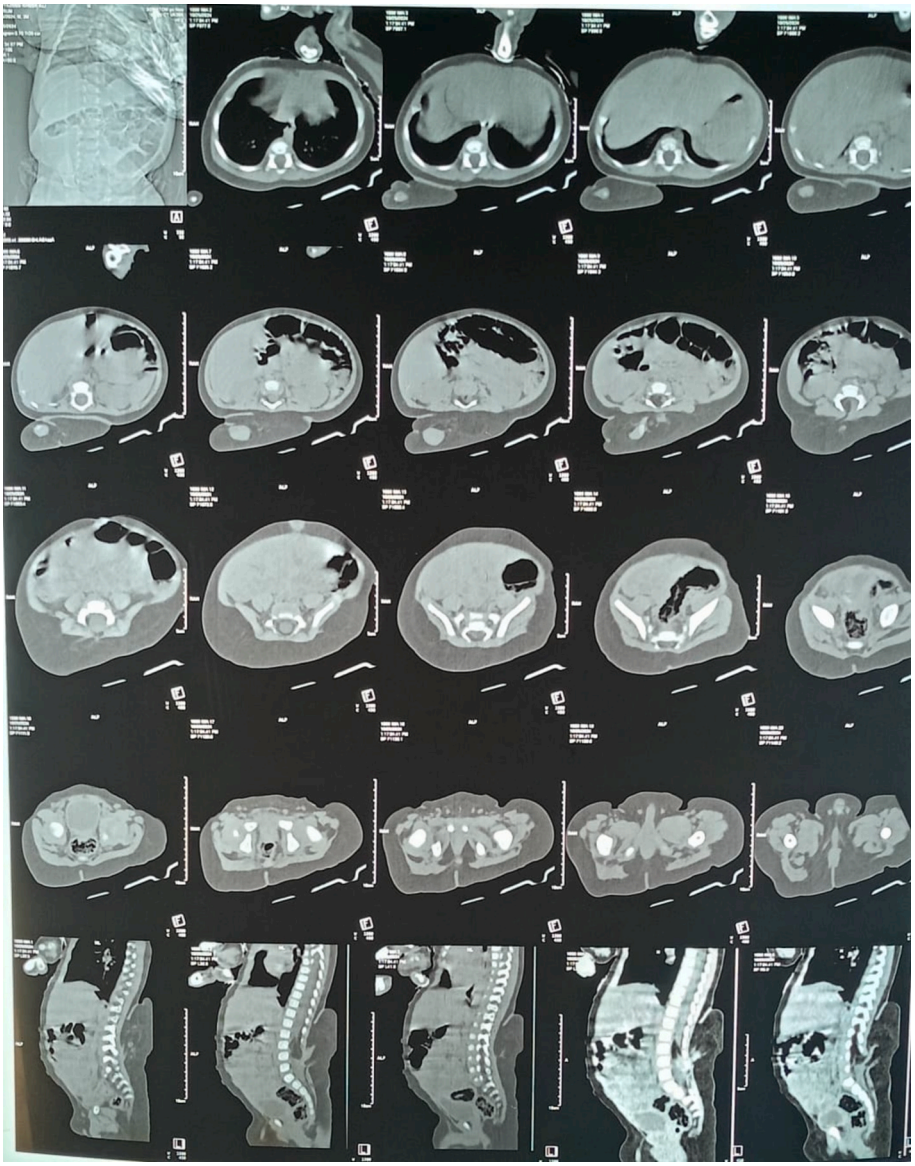

Fig. 3. Ct scan: showing spina Bifida and fatty deposition (lipomyelocele).

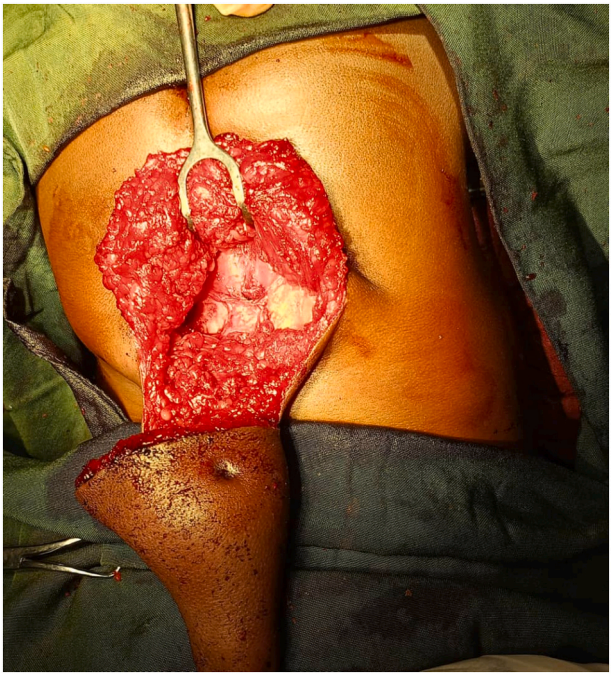

Fig. 4. Intra-operative: showing spinal defect (arrow) and limb remained attached by distal flap.

theory untenable. The fusion theory suggests that the presence of an additional limb results from the failure of regression of underdeveloped twin limb buds. In such instances, there is often an increased number of limbs, frequently accompanied by genitourinary and anorectal anomalies. Another hypothesis attributes the formation of the extra limb to neural tube disruption, which may cause a leakage of neural fluid that prompts mesodermal elements to differentiate and create an accessory limb. Additionally, the duplication hypothesis links the premature splitting of the limb bud to the emergence of an extra limb [8,11,12].

Polymelia can be diagnosed during antenatal care by qualified sonographers, though most cases are typically identified after birth. Among the various imaging modalities, MRI is the most effective for detailing the anatomical characteristics of the extra limb and for providing comprehensive insights into neural tube defects and the protrusion of neural elements. It also evaluates the vascular supply to the redundant limbs, while CT scans are more effective for visualizing bony structures. For accurate surgical planning, a multimodal imaging approach is recommended to thoroughly assess anatomical relationships. In our situation, an orthopedic specialist initially requested a plain X-ray, but after a multidisciplinary discussion, MRI was recognized as the optimal investigation. Unfortunately, due to the lack of availability in the city, a CT scan was requested to obtain additional information [12,13].

Surgical procedures are designed to address congenital malformations and involve the excision of excess tissue to enhance aesthetic outcomes. However, these interventions carry inherent risks, including complications such as bleeding, damage to adjacent structures, infection, and wound dehiscence. Additionally, specific complications may arise from neural injury, potentially resulting in limb weakness, urinary incontinence, and tethered spinal cord. Fortunately, our patient did not experience any postoperative complications [14].

4. Conclusion

The presentation of polymelia can differ significantly among cases, and the associated congenital abnormalities pose further challenges in management. Surgical intervention is often essential, though some complications cannot be prevented. It is advisable to implement long-term follow-up to identify any complications that may arise later.

Author contribution

Conceptualization, Data Curation, Formal Analysis,Methodology Momen Mohamed, Amin, Yassin, Moutaz Hamidr; Software and Writing – Review & Editing: Momen Mohamed.

Consent

Written consent was obtained by patient to be part of this job and provide permission for publication includes photography. A copy of consent available but written in Arabic to allow patient to understand purpose of study.

Ethical approval

Patient confidentiality will be maintained and data will be collected after full explanation of purpose of research and obtaining of an informed consent. Ethical approval was obtained from ethical committee of hospital.

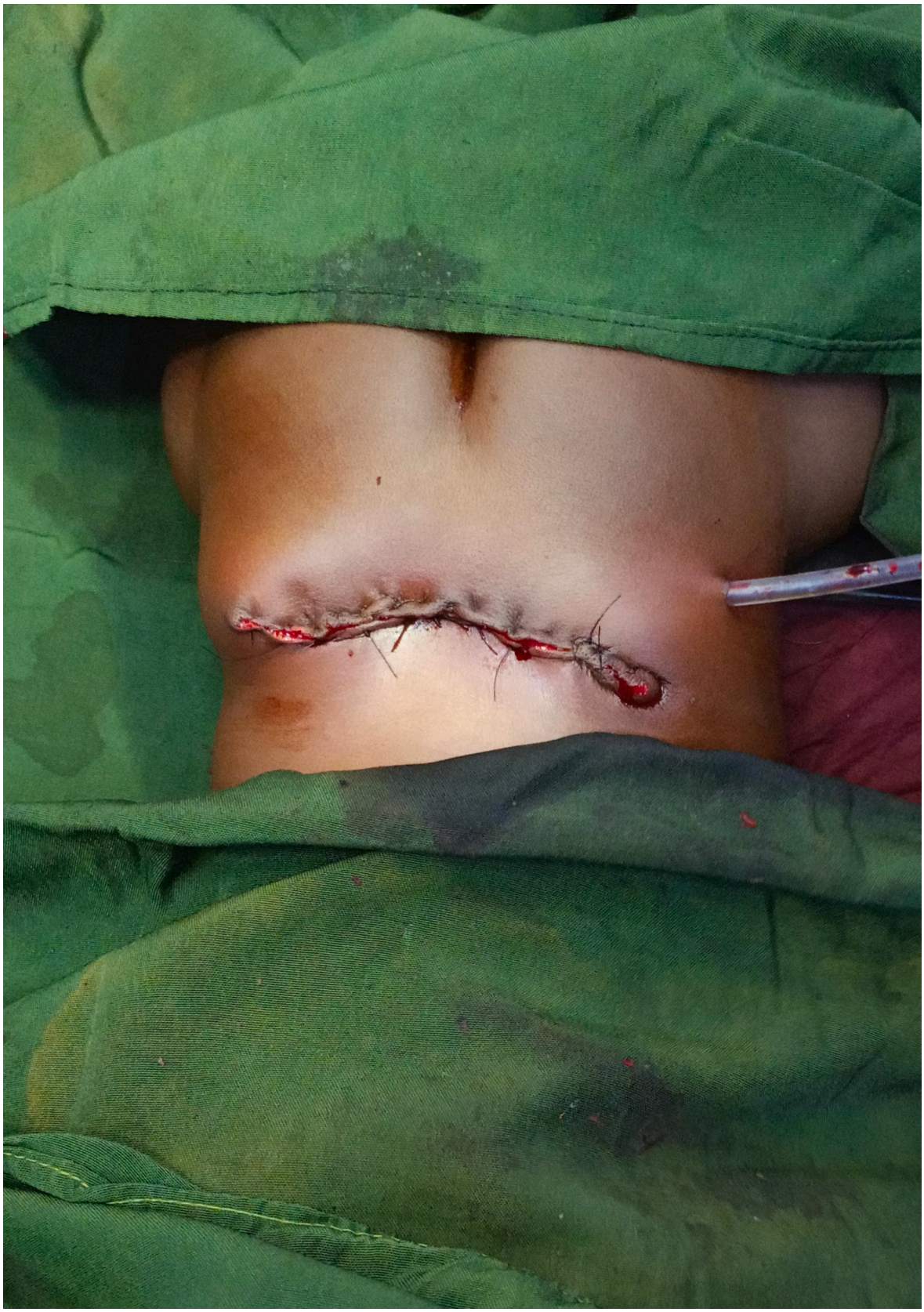

Fig. 5. Intra-operative: after excision of extra limb.

Guarantor

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Conflict of interest statement

All authors disclose no conflict of interest.

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