MANAGEMENT OF SEVERE KNEE FLEXION CONTRACTURES IN A CHILD WITH ARTHROGRYPOSIS MULTIPLEX CONGENITA

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ABSTRACT

Arthrogryposis multiplex congenita is a syndrome characterized by the presence of congenital contractures involving multiple joints usually with flexion deformities, with or without pterygia or webbing at the joints. The aim of this case report was to highlight the challenges of management of severe knee contractures in AMC and possible solutions in a resource-limited setting like ours. We presented a one-year-old child with bilateral severe knee contractures secondary to arthrogryposes. The involvement of knee is present in about 70% of cases with arthrogryposis, with flexion deformity more common than extension deformity. Knee flexion contractures, as seen in our patient, is one of the most disabling deformities. The surgical option adopted in the case presented were met with various challenges including abnormal muscles insertions, shortening and fibrosis of the muscles, abnormal position and shortening of the neurovascular structures in the popliteal fossa.

KEYWORDS Arthrogryposis, knee contractures, osteotomies

Introduction

Arthrogryposis multiplex congenital (AMC) is a syndrome characterized by the presence of congenital contractures involving multiple joints. Flexion contractures are more frequent than extension contractures. Pterygia or webbing at the joints may or may not be present [1]. AMC is due to reduce intrauterine fetal movements which occurs in conditions such as neuropathic and myopathic processes, maternal disease, abnormality of connective tissue, and impaired fetal or intrauterine vascularity [2].

In people with AMC, various joint of the extremities can be affected. However, knee joint involvement has been reported in 70% of the cases, with flexion more common than extension contractures [2, 3]. Knee flexion abnormality is hard to correct and has a high tendency to recurrence [3]. In this case report, the patient had contractures in both upper and lower limb. She received a posterior release and femoral osteotomy for her knee contractures. In a resource-limited setting, it is difficult to manage cases of severe knee contracture due to the risk of neurovascular damage. The aim of this case report is to highlight the challenges of management of severe knee contractures in AMC and proffer possible solutions.

Case Report

We present a nine months old female infant patient who presented at the age of 3 months with multiple joints anomalies. However, the anomalies were presented since the birth of the infant. The infant was born to a 28 years old para two (delivered twice before the index case). The pregnancy is at full term
and infant was delivered through vaginal delivery (vertex). The mother had full antenatal care though she had a brief febrile illness and rashes at first trimesters otherwise uneventful. There was no family history of any congenital anomaly, smoking, alcohol ingestion or drug use. Infant’s weight at delivery was 2.9 kg, within the normal range on the Apgar score and had neonatal jaundice that treated as an in-patient.

Examination revealed dysmorphic face and multiple joints anomalies of upper and lower extremities including webbing of elbow and axillary, fixed extension deformity, camptodactyly, absence of flexion creases and synphalangist of proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints of all the fingers, severe knee flexion contractures (120-degree on right and 120-degree on the left from full extension). There was also hypoplasia of the calf muscles, rigid bilateral club and significant widening of the sternal notch. [Figure 1 and Figure 2]

Plain X-ray revealed reduced space in the knee joint, and the absence of PIP and DIP joints in the fingers. Echocardiography was normal and complete blood count; electrolytes were all within normal values.

Surgically, the infant underwent soft tissue release and lengthening of the hamstring tendons and closed distal femoral extension osteotomy on the right side. It was observed intraoperatively that the soft tissues of the knee were thickened and fibrosed, and insertion of hamstring muscles was abnormal.

After surgeries, the knee gained 100 degrees (that is, achieved 20 degrees from full extension) on the right and left knee intraoperatively while the patient was still under general anesthesia with a muscle relaxant. However, severe reduction of vascular flow was compromised on Doppler assessment in lesser extension than on skin traction which is 80 degrees short of full extension. The limb was therefore kept at 80 degrees short of full extension, and a graded skin traction was then applied for three weeks. The wound was covered with a split-thickness skin graft after 130 degrees of the extension was achieved. The patient continued with physiotherapy; muscle strengthening exercise, tense, range of motion exercise and night splinting for six months. The infant had a 120-degree flexion range and -20-degree extension range, which was maintained up to 6 months. The left knee operated on four months (Table 1) and the wound was resurfaced with lateral and medial thigh flaps and split thickness skin graft.[Figure 3 and Figure 4]

<table>
<thead>
<tr>
<th>Events</th>
<th>Right</th>
<th>Left</th>
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<tr>
<td>Preoperative angle</td>
<td>120</td>
<td>120</td>
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<tr>
<td>Postoperative angle</td>
<td>80</td>
<td>60</td>
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<tr>
<td>Angle after postoperative traction</td>
<td>20</td>
<td>18</td>
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<tr>
<td>Night splinting</td>
<td>20</td>
<td>20</td>
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<tr>
<td>Six months postoperative</td>
<td>22</td>
<td>21</td>
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<tr>
<td>One year postoperative</td>
<td>35</td>
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Figure 1: Preoperative photograph lateral aspect of right lower limb.

Figure 2: Preoperative photograph medial aspect of left lower limb.
Discussion

The aetiopathogenesis of arthrogryposes is multifactorial. It may result from fetal malposition and crowding, oligoamnios, connective tissue anomalies or neurologic dysfunctions [4]. Genetic predisposition is likely. It is reported that mutations in at least five of the genes which encode a component of the contractile apparatus of skeletal muscles may be accountable [5]. Environmental factors affecting the mother and fetus such as infections, medications, radiation, traumas and chronic illnesses may also contribute to the development of AMC [6].

The infant presented in this case report had both upper and lower limbs affected. She had severe knees contractures for which a posterior release and femoral osteotomy were performed. The surgical option selected was one of the possible treatment modalities for consideration. We use this case to highlight the challenges of management of severe knee contractures in AMC and possible solutions for these challenges. Knee involvement was present in about 70% of the cases with arthrogryposis, with flexion deformity more common than extension deformity [2]. Knee flexion contractures, as seen in this case, is one of the most disabling of all deformities. Flexion contractures of the knee can restrict standing, walking, and any other weight bearing activities and make the bed or chair positioning difficult [7]. Patients with extension contractures, on the other hand, can have difficulty with squatting, sitting or crawling. Other knee abnormalities may include flatten femoral condyles, reduced joint space, thicken and contracted joint capsule, hypoplastic muscles, partial fibrosis of soft tissues and abnormal insertions [4]. The children with very severe arthrogryposis often have intra-articular deformities similar to the case presented. In such cases, soft tissue release alone is not sufficient to correct the deformity.

Previous methods for managing knee flexion contracture have included casting [8], bipolar traction [9], posterior soft tissue release [10], osteotomies [11], and femoral shortening [12]. These have been used alone or in combination. The outcomes of these methods are variable depending on the severity of the deformity and its etiology [13]. Treatment options are determined based on the seriousness of the flexion deformity. For the mild to moderate cases, serial casting, tractions or other physical therapy modalities starting early in life may be sufficient. However, nonoperative methods are very labor intensive and are ineffective in the presence of ankylosis [13]. For the severe to very severe cases, surgical interventions are necessary. It may include extensive soft-tissue release, Femoral, osteotomy or shortening, plastic, and reconstructive procedures, neural and microvascular reconstruction [14].

Posterior release and Ilizarov distraction have achieved good results. The major disadvantages of this method are the long fixation time and limitation of progress in the presence of intra-articular abnormalities [15]. Posterior release with femoral extension osteotomy, as presented in this case study, is better because the intra-articular abnormalities addressed surgically. However, recurrence of contracture because of skeletal immaturity is common following femoral extension osteotomies. Loss of joint range of motion after osteotomy at the rate of 10 per month has been reported. Therefore, long-term rigid night splinting is necessary [3].

Major challenges in soft posterior tissues release, as shown in the case presented, are abnormal muscles insertions, shortening and partial fibrosis of muscles. In this case, Z-lengthening of the hamstrings was done as per previous reports [16]. Other
reports have suggested that in the presence of intra-articular anomalies soft tissue release will only provide 30 to 70 degrees improvement in extension. In our case, distal femur extension osteotomy was done in addition to soft tissue release, and that resulted in a gain of 100 degrees in extension. Another challenge is the abnormal position and shortening of the neurovascular structures in the popliteal fossa which often requires Doppler or other imaging assessment before release. After the soft tissue release, intraoperative Doppler assessment of the distal arterial flow is crucial because of the possibility of vascular spasm from strain on the vessels. In the event of a significant reduction in blood flow in distal arteries, splinting should be avoided, and gradual distraction with skin tractions can be used with an aim to achieve maximum straightening within two weeks. Neurovascular structures can be exposed, and this often requires skin flap cover. In this case, thick sheet skin graft was applied two weeks after the first surgery to allow for maximum extension to be achieved first.

Postoperative rehabilitation is necessary to maintain the joint range gained during surgery [17]. For the infant presented, immediately after the surgery, the knee was kept in an extended position with a static splint for 3 to 6 weeks, followed by a rigid splint at night for six months. Physiotherapy was continue six months post-surgery. Review of literature has shown that children with significant movement in the distal parts and those who could do knee standing before corrective surgeries have a better prospect for independent ambulation. In our case, the patient is yet to achieve independent ambulation.

Conclusion

Our case illustrates the various challenges that may associate with the posterior release and osteotomy for the correction of joint contractures in people with AMC. The challenges may include abnormal muscles insertions, shortening and partial fibrosis muscles, abnormal position and shortening of the neurovascular structures in the popliteal fossa. Our case demonstrates the solutions that can be considered to overcome these challenges.

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Disclosure Statement

There were no financial support or relationships between the authors and any organization or professional bodies that could pose any conflict of interest.

Competing Interests

The authors declare no conflict of interest.

Ethical Approval

Informed consent was sought from the parents of the patient for this study. Ethical approval was also obtained ethical committee of the hospital.

References