SPASTIC QUADRIPLEGIC CEREBRAL PALSY WITH EXPOSED MALUNION BONE AT BILATERAL PROXIMAL HUMERUS AND DISTAL FEMUR

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ABSTRAK

Anak dengan cerebral palsy berisiko mengalami fraktur kerapuhan akibat rendahnya densitas tulang yang disebabkan oleh berbagai etiologi. Status ambulasi pada pasien cerebral palsy merupakan salah satu faktor yang dapat memengaruhi densitas tulang. Penatalaksanaan fraktur pada kondisi ini sering kali berakhir dengan malunion, baik dengan terapi non-operatif maupun operatif. *Spastic quadriplegic cerebral palsy* (SQCP) merupakan tipe yang berat, menyebabkan kelumpuhan dan kekakuan otot pada kedua lengan dan tungkai, serta dapat memperberat proses penyembuhan fraktur tulang. Seorang gadis berusia 9 tahun telah mengeluhkan luka terbuka di bahu kanan dan kiri serta lutut kiri sejak 4 bulan yang lalu. Pemeriksaan radiografis pada kedua bahu dan lutut menunjukkan malunion pada humerus proksimal bilateral dan femur distal kiri. Pasien didiagnosis menderita cerebral palsy spastik quadriplegia GMFCS V dengan tulang malunion yang terekspos pada humerus proksimal bilateral dan femur distal. Kemudian ia diobati dengan antibiotik ampicillin sulbactam, luka dibersihkan (debridement), dilakukan osteotomi, dan penutupan luka primer. Penderita cerebral palsy GMFCS IV dan V memiliki insidensi fraktur tulang yang lebih tinggi, dan hal ini terkait langsung dengan densitas mineral tulang (BMD). Beban mekanis, baik akibat kontraksi otot maupun beban gravitasi, menyebabkan deformasi tulang dan meningkatkan kekuatan tulang. Spastisitas ditandai dengan peningkatan tonus otot, refleks yang hiperaktif, dan kehilangan gerakan sendi yang menyebabkan kontraktur.

Kata kunci : *cerebral palsy, malunion, osteotomi, quadriplegia,* tulang terekspos

ABSTRACT

Children with cerebral palsy are at risk of fragility fracture due to low bone density from various etiologies. The ambulatory state of cerebral palsy patients is one of the factors that can influence bone density. Treatment of fractures for this condition often results in malunion, whether treated non-operatively or surgically. Spastic quadriplegic cerebral palsy (SQCP) is a severe type that make paralysis and muscle stiffness of both arms and legs, and will aggravate the healing process of bone fractures. A girl 9-year-old has been complaining of open wounds on the right and left shoulder and left knee since 4 months ago. Initially in the form of a protrusion from under the skin, it becomes an open wound with the proximal humerus bone and distal femur bone coming out. Then she was treated with the antibiotic ampicillin sulbactam, the wound was debridement, osteotomy shortening, and primary wound closure. GMFCS IV and V cerebral palsy sufferers have a higher incidence of bone fracture fragility, and this is directly related to bone mineral density (BMD). Mechanical loading, either due to muscle contraction or gravitational loading, causes bone deformation and increases bone strength. Spasticity is characterized by increased muscle tone, hyperactive reflexes, and loss of joint motion which is leading to contractures. These will cause high rate of fractures in low bone density, disrupt bone healing process, and can make exposed bones as in this case. The treatment of spastic quadriplegic cerebral palsy must be adjusted to the spasticity of the muscle tone and the patient's medical comorbidities. The action that can be undergone in conditions of exposed malunion bone are administering antibiotics, osteotomy debridement, closing the wound to reduce the risk of bone infection.

Keywords *: cerebral palsy, malunion, exposed bone, quadriplegic, osteotomy*

INTRODUCTION

Cerebral palsy (CP) is a collection of conditions impacting movement, balance, and posture, making it the primary motor disability in childhood. It arises from irregular brain development or damage during brain development, leading to difficulties in muscle control. It is affecting approximately 2.0 to 3.5 per 1000 live births (Sadowska et al., 2020). The Gross Motor Function Classification System (GMFCS) is commonly utilized to evaluate gross motor skills, particularly focusing on sitting and walking capabilities in CP children (Haberfehlner et al., 2020). This classification system categorizes children based on their capacity for mobility and sitting, focusing on their ability to perform specific functional tasks and their requirement for assistive devices like canes, crutches, walkers, or wheeled mobility aids (Nylén & Grooten, 2021). Cerebral Palsy is classified into spastic, dyskinetic, and ataxic based on the type of movement disorder. This disease is also divided into four types based on the area of presentation, namely quadriplegic, hemiplegic, diplegic and monoplegic. Diplegic occurs most frequently, followed by hemiplegic (20–30%) and quadriplegic (10–15%). Spastic presents with hypertonicity and hyperreflexia, it may be unilateral or bilateral. Dyskinetic presents with involuntary, uncontrolled, repetitive, and sometimes stereotype movements with altered muscle tone. Ataxic is in coordination with a decreased muscle tone (Paul et al., 2022).

In a previous study, 1,451 children with CP and 787,159 without CP were examined. Fracture rates per 1,000 person-years were similar between females and males with and without CP. However, males, epilepsy, and anti-seizure medication were associated with higher fracture rates, while CP diagnosis or GMFCS-level were not had any association. Non-ambulant children with CP had more lower extremity fractures. Upper extremity fractures were more common in children with lower GMFCS levels, while femoral fractures were more prevalent in those at GMFCS level V (Granild-Jensen et al., 2022). A significant number of children with cerebral palsy experience heightened muscle tone, leading to potential complications such as increased shortening or improper healing at fracture sites. Malunion, characterized by excessive shortening exceeding 2 cm, angulation surpassing 30° in the sagittal plane or 10° in the frontal plane, or rotation beyond 10°, is a concern. In cases of distal femur fractures, varus/valgus angulation or flexion/extension angulation up to 10° is typically tolerated (Linton et al., 2022).

Hence, when managing bone fractures in children with cerebral palsy, it's crucial to employ strategies that minimize the need for prolonged immobilization. This may involve utilizing casts or splints that enable the child to resume standing or considering surgical interventio ns. Furthermore, children with cerebral palsy often exhibit lower bone density, which, when coupled with immobilization, can predispose them to additional fractures at various sites, including the same or opposite extremities (Mughal, 2014). In the following case study, we'll examine a child with a history of cerebral palsy who presented to the emergency department with injuries to both shoulders and the left knee, along with visible protrusion of bones.

CASE PRESENTATION

A 9-year-old child was brought to Emergency Department with complaints of open wounds on the right and left shoulder and left knee since four months ago. Initially in the form of a protrusion from under the skin, it becomes an open wound with the proximal humerus bone and distal femur bone coming out. The wounds were discharging blood and pus. Over the past week, the wounds have increased in size and bones have become visible within the wounds. There were no history of child abuses. The patient has Cerebral Palsy and unable to walk with weakness in all extremities, sometimes she experienced convulsions for 1-2 minutes. The patient is the first child of four siblings, there were no reported maternal illnesses during

pregnancy. The mother received regular prenatal care from a midwife. The patient was delivered via Cesarean section with a full-term gestational age, weight was 3400 grams. The patient cried immediately after birth, and there were no congenital defects noted.

Figure 1. Clinical Manifestations of Exposed Bone in The Proximal Right and Left Humerus

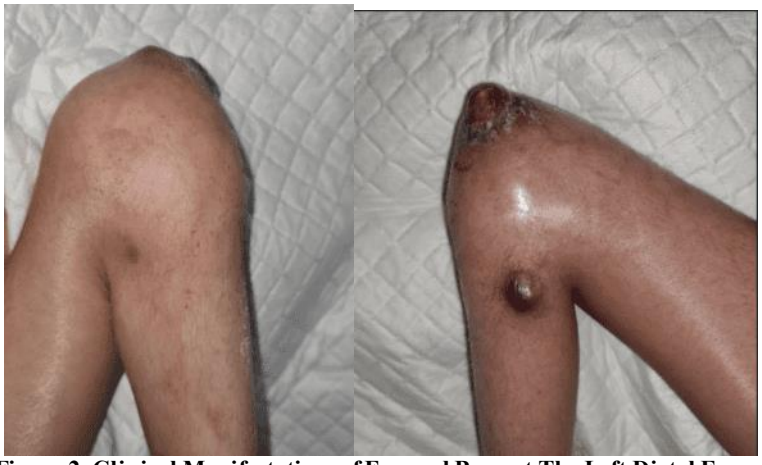

Figure 2. Clinical Manifestations of Exposed Bone at The Left Distal Femur

Examination of vital signs were within normal limits. Her body weight was 15 kg. tachycardia, and other examinations within normal limits. On physical examination, open wounds were found on the right and left shoulders measuring 5x3 cm and 4x2 cm, and on the left knee measuring 3x2 cm. Prominent bone was visible in all wounds, distal blood vessels were normal, and all range of motion were reduced. Laboratory tests revealed anemia, thrombocytosis, and hypoalbuminemia.

Table 1. Laboratorium Examination

	Value	**Unit**
Hb	8,9	g/dL
Wbc	6.300	sel/cm3
Plt	114.000	sel/cm3
HCT	37	%
CRP	42,4	mg/L
Alb	2,4	g/dL
Na	130	Mmol/L
K	3,5	Mmol/L
Ca	1,04	Mmol/L

The patient underwent imaging examinations on the right and left humerus and femur. At the left and right humerus anterior-posterior (AP) and lateral radiographs, malunions were found in the proximal 1/3 of the humeral bone with callus formation, along with a defect and soft tissue swelling in the shoulders region. Meanwhile, in the left femur anterior-posterior (AP) and lateral radiographs, malunion was observed in the distal 1/3 of the femoral bone with callus formation, soft tissue defect and swelling in the knee region.

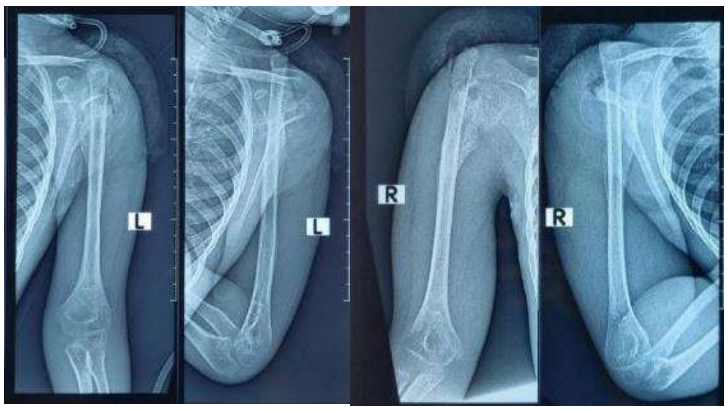

Figure 3. Left and Right Humerus AP/Lateral Rontgens

Figure 4. Left Distal Femur AP/Lateral Rontgen

Figure 5. Post Operative Clinical Manifestation of Right and Left Proximal Humerus

The patient was diagnosed as spastic quadriplegic cerebral palsy GMFCS V with exposed malunion bone at bilateral proximal humerus and distal femur, bicytopenia, and hypoalbuminemia. Then, she was treated with the injection antibiotic of 150 mg/kg/day Ampicillin Sulbactam and 150 mg Metamizole every 8 hours, immediate surgical debridement,

osteotomy shortening on the right and left proximal humerus and distal left femur, and primary wound closure in the operating theater. The patient was planned for general condition improvement and wound culture.

Figure 6. Post Operative Clinical Manifestation of Left Distal Femur

DISCUSSION

Cerebral Palsy (CP) encompasses a set of enduring challenges affecting movement and posture, leading to limitations in activity. These issues stem from non-progressive disruptions that occur during fetal brain development or early infancy. Motor difficulties associated with CP frequently coincide with disruptions in sensation, perception, cognition, communication, and behavior. Additionally, individuals with CP may experience epilepsy and secondary musculoskeletal diseases. Cerebral Palsy (CP) is marked by a diverse range of risk factors, underlying causes, clinical presentations, functional impairments, associated conditions, management approaches, and progression over an individual's lifespan. Recent research by Shevell et al. suggests viewing CP as a spectrum disorder rather than distinct clinical entities. CP classification typically involves a combination of motor type and topographic distribution. Motor types encompass descriptors such as spastic, ataxic, dyskinetic, or mixed, while topographic classification indicates which body parts are affected, such as diplegic, triplegic, tetraplegic, quadriplegic, or hemiplegic. Severity levels of CP are often categorized subjectively as mild, moderate, or severe (Shevell et al., 2019). In the case presented, the child's history indicates cerebral palsy with long-standing limb immobility since childhood.

According to the updated Gross Motor Function Classification System (GMFCS), individuals are categorized based on their level of mobility and functional abilities. GMFCS level I signify unrestricted walking abilities. Those in GMFCS level II may walk with some limitations, such as balance issues or the need for hand-held aids before age four, possibly requiring wheeled mobility aids for longer distances. GMFCS level III individuals can walk indoors with hand-held aids but rely on wheelchairs for community and longer distances, with sufficient sitting and standing abilities for transfers. GMFCS level IV individuals can sit with support but have limited independent mobility, often utilizing manual wheelchairs or powered mobility devices. Children in this group may require assistance for mobility aids, and their abilities may remain similar from ages 6 to 12. GMFCS level V represents the most severe impairments, with individuals relying solely on powered wheelchairs for mobility.

These individuals typically lack independent head and trunk control before age two and may require assistance for rolling. While their abilities may stabilize after age four, full assistance in transfers becomes necessary after age six (Löwing et al., 2015). In this case, the patient's limited body movement since birth aligns with the classification criteria for cerebral

palsy at GMFCS level V. Malunion in cerebral palsy, while less common compared to other complications, can still occur. Due to the underlying motor impairments and musculoskeletal issues associated with cerebral palsy, fractures may heal improperly, leading to malunion in some cases. However, the prevalence of malunion in cerebral palsy is relatively low compared to other conditions. Nonetheless, it remains an important consideration in the management of fractures in individuals with cerebral palsy, particularly given the potential impact on mobility and overall function (Wahyuni, 2023). Cerebral palsy often presents with muscle spasticity and imbalance due to the underlying brain injury. Spasticity can lead to abnormal forces acting on bones during movement or weight-bearing activities, predisposing them to fractures. Imbalance in muscle tone can further contribute to abnormal stress distribution around the fracture site, affecting healing. Individuals with cerebral palsy may have impaired bone healing mechanisms due to factors such as poor blood supply to the bone, reduced osteoblast activity, or impaired signaling pathways involved in bone repair. This delayed healing process can increase the likelihood of malunion if the fracture fragments do not adequately realign during the healing process (Enslin et al., 2020).

Immobility or reduced weight-bearing due to motor impairments in cerebral palsy can prolong the healing process and contribute to malunion. Limited mobility may prevent normal stress distribution on bones, leading to inadequate remodeling and alignment during the healing phase. Contractures and joint deformities commonly observed in cerebral palsy can further complicate the management of fractures. These musculoskeletal abnormalities may interfere with the application of casts or splints, leading to suboptimal immobilization and subsequent malunion. Individuals with cerebral palsy may have nutritional deficiencies or difficulties with feeding, which can impair bone health and hinder the healing process (Han et al., 2017). Poor nutritional status can lead to decreased bone density and impaired bone remodeling, increasing the risk of malunion. Overall, the pathophysiology of malunion in cerebral palsy is complex and involves a combination of factors related to the underlying neurological condition, musculoskeletal abnormalities, healing mechanisms, and nutritional status. Effective management requires a comprehensive approach addressing these various factors to optimize outcomes and prevent long-term complications (Alvarez Zaragoza et al., 2018).

The association between quadriplegic spasticity and bone-exposed fractures in CP patients is multifactorial. Quadriplegic spasticity, characterized by severe muscle tightness and stiffness affecting all four limbs, can significantly impair mobility and increase the risk of falls or trauma, which may lead to fractures. Additionally, individuals with quadriplegic CP often have poor bone quality due to factors such as decreased weight-bearing activities, limited mobility, and altered bone metabolism, further predisposing them to fractures. The combination of spasticity-induced muscle imbalance, reduced bone strength, and increased susceptibility to trauma increases the likelihood of bone-exposed fractures in quadriplegic CP patients (Wahyuni, 2023).

Clinical evaluation plays a crucial role in assessing the functional impact of malunion in cerebral palsy and guiding treatment decisions. By carefully evaluating the individual's mobility, range of motion, and overall functional abilities, healthcare providers can determine the necessity for corrective measures. Imaging studies, such as X-rays or CT scans, are often utilized to further evaluate the extent of malunion and aid in developing an appropriate management plan. In cases where corrective action is deemed necessary, surgical intervention may be recommended. Corrective surgery, particularly in the metaphysis region, aims to realign the affected bone, such as the proximal humerus, to its correct position. This procedure can help restore proper anatomical alignment, alleviate pain, and improve functional outcomes for individuals with cerebral palsy affected by (Javvaji et al., 2023; Vigni et al., 2020). There was a notable decrease in pain scores and significant improvement in radiographic parameters following the corrective osteotomy. The authors recommend performing the corrective

osteotomy within one year of the malunion, as it results in greater gains in range of motion (Selles et al., 2020; Tarabishi et al., 2023).

For pediatric patients requiring emergency surgery to address an exposed bone or traumatic amputation, it is advised to administer peri-operative prophylaxis with cefazolin intravenously at a dose of 30 mg/kg (with a maximum dose of 2 gram) within 30 minutes prior to the procedure. This dosage may be repeated if the surgery extends beyond 4 hours, unless the patient is already receiving broad-spectrum antibiotic therapy (Opri et al., 2022). Based on antimicrobial stewardship (AMS) in our hospital, we used an injection antibiotic of Ampicillin Sulbactam for open wound with exposed bone, muscle or tendon more than 6 hours. Debridement and irrigation of contaminated wounds with ample saline solution are crucial initial steps in managing exposed bone in children. It's important to employ a more conservative approach to debridement in children compared to adults, preserving bone fragments with uncertain viability and soft tissue attachment, as they may aid in bone healing. Although surgical site infections are a common complication in orthopedic trauma surgery for adults, they occur less frequently in children. Children generally have a better prognosis than adults due to their superior soft tissue recovery capacity (Aulisa et al., 2023; Chen et al., 2019).

CONCLUSION

The management of spastic quadriplegic cerebral palsy necessitates tailored approaches considering the degree of spasticity in muscle tone and any concurrent medical conditions present in the patient. When dealing with cases of exposed malunion bone, treatment options typically involve the administration of antibiotics, performing osteotomy procedures for realignment, debridement to remove damaged tissue, and wound closure to mitigate the risk of bone infection. These interventions aim to address both the underlying musculoskeletal issues and the potential complications associated with bone exposure, ensuring optimal outcomes for the patient's overall well-being.

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