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Case report

A simple management of massive bone defect after en-bloc resection of osteofibrous dysplasia of tibial shaft: A case report

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ABSTRACT

Introduction: Osteofibrous dysplasia is a relatively rare disease, exclusively found in children, affecting the tibial diaphysis. Various management approaches are already available, but an internationally approved management guideline is not yet established. There is a major concern in the current management of wide excision technique as it frequently results in massive bone defect.

Case presentation: Here we present a case of osteofibrous dysplasia on a 10-year-old girl in Cipto Mangunkusumo Hospital with chief complaint of mild persistent pain of her lower leg since two years before with slight bowing deformity. The radiograph and histopathological examination support the diagnosis of osteofibrous dysplasia. She was managed with en-bloc resection (wide excision) of the tumor, followed with reconstruction using biomaterials substitute; combination between demineralized bone matrix (Bonegener^{R}) and bone substitute "hydroxyapatite and calcium sulphate" and internal fixation using plate and screw.

Results: Clinical and radiological evaluation showed successful improvement and outcome. The patient showed progressive functional outcomes and achieved functional score of 100% LEFS at 3 years follow-up. The plate and screw was removed at 48 weeks after adequate callus formation andradiological union was achieved.

Conclusion: Simple reconstruction using biomaterial bone substitute not only created new bone formation with good stability, but also enabled patient to have an improved quality of life. This method is recommended to overcome the massive bone defect after tumor resection in osteofibrous dysplasia patient.

1. Introduction

Osteofibrous dysplasia (OFD) is a neoplasm of the bone commonly occurs in the tibial diaphysis. The incidence of OFD is approximately 0.2% of all bone neoplasms [1]. Cases are mostly found in younger patients in the first two decades of life. The main cause of OFD is still unknown and sometimes is associated with adamantinoma [1–4]. The most common presenting symptom of OFD is a painless enlargement of the tibia with varying degree of tibial bowing deformity [3].

There is no consensus on the standard treatment of OFD. One approach is by performing resection of the affected region and replacing the defect with bone grafts along with fixation to ensure stability. Other approaches include extra-periosteal excision, autologous free fibular graft, and bone graft application; and other less-invasive procedures e.g. curettage or even the conventional watchful waiting [1,5–7]. However, tumor resection for OFD can cause massive bone defect which increases risk of fractures, unstable graft, and longer bone healing period [8].

There are several factors required to stimulate the process of bone union. They are osteo-inductive (mesenchymal cell and growth factors), osteogenic cells, osteoconductive (scaffolds), and mechanical stability [5,6]. The main concern for orthopaedic surgeons are reconstruction of the massive bone defect in OFD can stimulate the bone healing cascade by promoting an initial vascular in-growth from periosteum [7].

One of surgical approaches to manage the massive bone defect in pediatric patients is bone graft application either using autologous or allogeneic bone graft [8]. However, the procedure resulted in a larger bone defect at the donor site or a larger amount of bone graft from bone bank, prolonged severe pain, nerve damage, bleeding, infection, fracture, hematoma, and sensory loss [9]. Hence, reconstruction of massive bone graft should be simpler and safer particularly for OFD in pediatrics. In this report, we presented a case of OFD in which we treated with wide excision of the tumor, followed with reconstruction using bone material; substitution "hydroxyapatite and calcium sulphate" and internal fixation with plate and screw. This case report has been reported in line with

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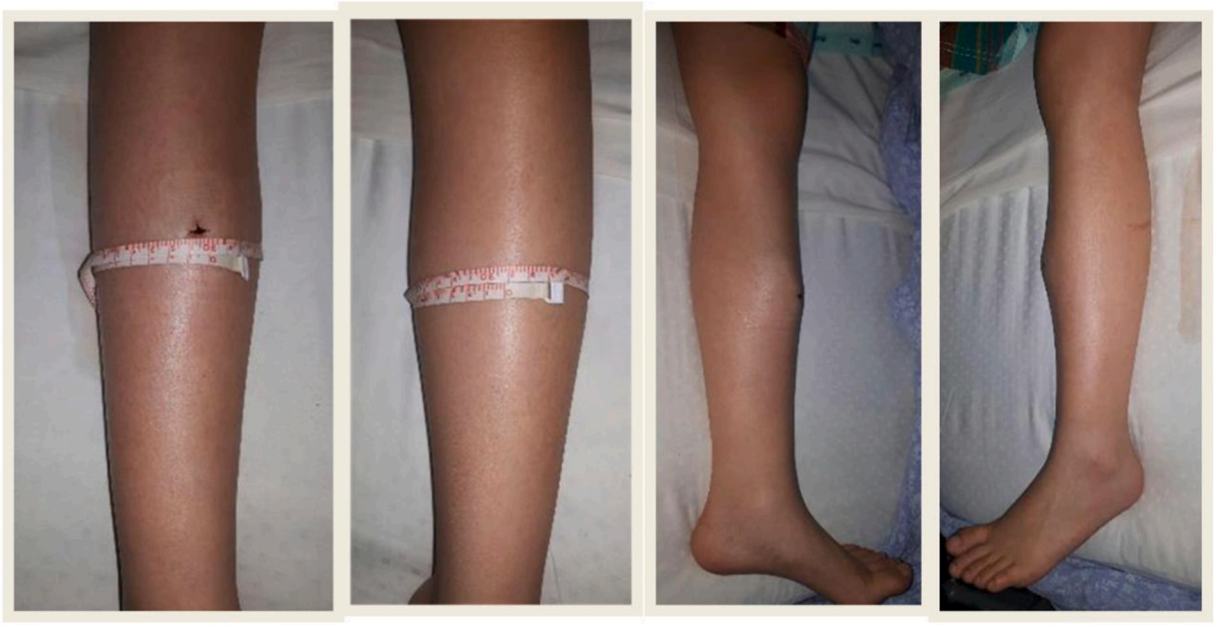

Fig. 1. Pre-operative clinical condition. A mass on the anteromedial side of the tibia with bowing deformity.

the SCARE Criteria [10].

2. Case presentation

A 10-year-old girl was referred to our hospital with chief complaint of mild persistent pain of her lower leg since two years before with slight bowing deformity. Her past medical history showed that she had experienced an injury on the left lower leg. Three months later, a hard mass appeared on her injury site. Physical examination found a painless solid mass with anterolateral bowing deformity of the left tibia. No sign of inflammation or skin discoloration was found [Fig. 1].

Blood analysis results showed normal range of white blood cell and erythrocyte sedimentation rate, but an increased alkaline phosphatase and lactate dehydrogenase. The radiographs showed typical presentation of OFD that's a bubbly appearance of geographic lytic lesion with no periosteal reaction [Fig. 2A]. Magnetic resonance imaging (MRI) with contrast revealed a hyperdense lesion with sclerosis of the internal cortical surface with no evidence of soft tissue involvement [Fig. 2B].

Histopathological examination from specimen retrieved through core biopsy was consistent with osteofibrous dysplasia, showing C-shaped bony spicules, immature bone trabeculae line with osteoblastic rimming [Fig. 3]. Daily functionality measured by lower Extremity Functional Scare (LEFS) was 15%.

Single surgeon, AFK, had performed the surgery at our hospital. This study has not involved multidisciplinary team. The patient was in supine position while applying a tourniquet on the left thigh. An anterolateral approach was performed to expose the lesion. The periosteum was preserved and en bloc resection of the tumor 10 cm in length was performed to ensure removal of neoplastic tissue, thus leaving massive bone defect. The massive bone defect was filled by combination between demineralized bone matrix (Bonegener^{R} 5 cc) and bone substitute "hydroxyapatite and calcium sulphate" (PerOssal^{R} 50 pellet). The periosteum was sutured and had internal fixation application using plate and screw on the medial site of the tibial shaft [Fig. 4]. The patient suffered no acute complication and was discharged on the fifth day after surgery. The outcome measured were subjective complaints, functionality based on LEFS, radiological assessment based on callus formation/bone formation [Fig. 5]. Partial weigh bearing was allowed at fourth week post-operative and full weight bearing at eighth week post-operative. No significant complaint was reported during 3-years follow-up. The patient's LEFS gradually improved, significantly since eight weeks post-operative.

On the third year post procedure, the patient was able to carry out daily activities and regularly participated in high intensity exercise with the score 100% of LEFS [Fig. 6]. Progression of post-operative outcomes is summarized in Graph 1.

3. Discussion

Osteofibrous dyplasia is a benign bone dysplasia in which lytic lesions occur in the cortex of the tibia and fibula among pediatric patients [1–3]. The presented case is a pediatric patient with OFD who had a painless mass with bowing deformity of tibia. OFD is typically located at the tibial diaphysis [9].

Based on radiographs and MRI, our study is consistent with the classic pathologic findings in OFD as a cortical thickening interspersed with multiple roundish lytic areas with a "bubbled" appearance. Neither periosteal reaction nor a transitional zone is generally found in OFD. Meanwhile, histologic evaluation of tissue from the lesion revealed a fibrous stroma with spicules of trabeculae which is characteristically rimmed with osteoblasts [1–3,9].

One of current treatment approaches for OFD is wide excision with various techniques of bone reconstruction which may lead to a massive bone defect [3,4,11]. There are several alternatives to reconstruct the massive bone defect including bone graft, bone transfer, biomaterial substitutes, bone transport, or mesenchymal stem cell implantation [3]. Bone graft is more suitable for a large bone defect because it creates a stronger and more stable construction despite its longer operating hours,

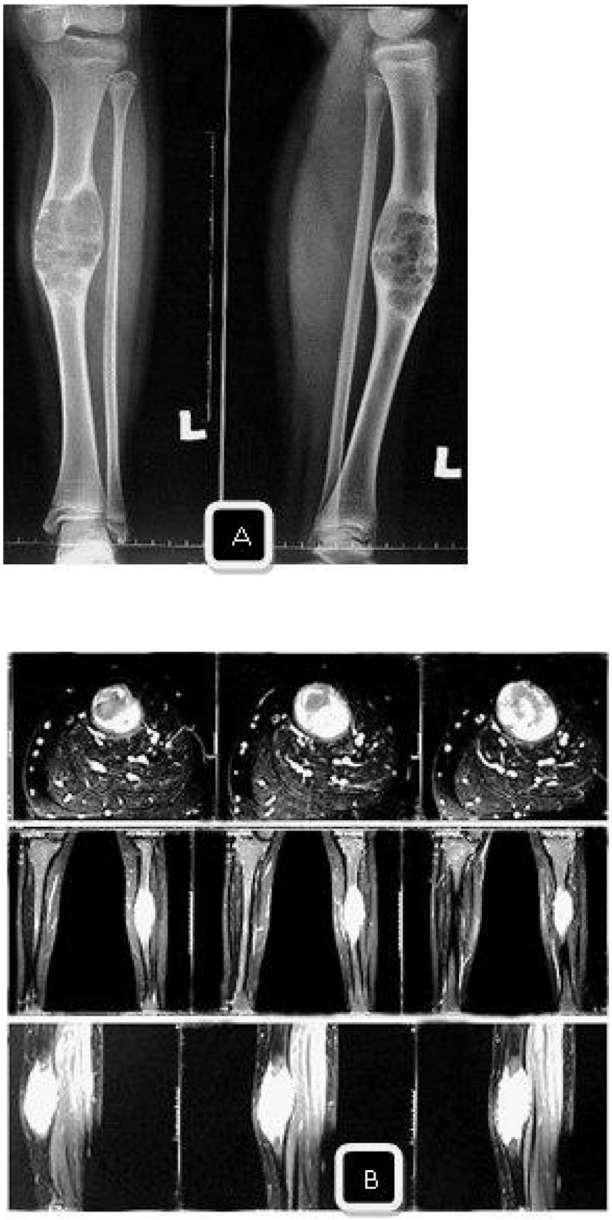

Fig. 2. A. Radiographs showed bubbled appearance of the shaft tibia.
B. MRI revealed hyperintens lesion of the shaft tibia.

higher on site morbidity, and several complications including infection, nerve injury, prolonged pain, etc. [11]. Furthermore, the autologous material does not always carry a high potential to generate bone, depending on the donor's age and comorbidities [12]. Bone transport requires a skillful technique; however it sometimes causes malunion or non-union due to mal-alignment during healing process [13]. Mesenchymal stem cell technique is successful to overcome the great bone defect, but a higher cost to create stem cells are needed [3,14]. Therefore, the simplest, the safest and the most effective treatment is needed in management of massive bone defect after tumor resection.

We revealed technique using reconstruction of bone defect with biomaterial substitutes; combination between demineralized bone matrix and bone substitute "hydroxyapatite and calcium sulphate". This technique is very simple as it only needs biomaterial substitutes to fulfill the existing bone defect with no cut to the bone. Biomaterials are synthetic or non-vital natural materials which are used in clinical practice for therapeutic aims and placed within the human body or in contact with the patient's tissue [12]. As these materials often remain within the body, their biocompatibility is a necessary prerequisite [12].

Biomaterials also have ability to decompose when new bone formed (biodegradability). Their characteristics are appropriate with osteo-inductive/osteo-conductive properties to stimulate cellular proliferation and osteogenic differentiation in the healing site [13].

The role of biomaterials in osteo-conductivity is to provide an ideal

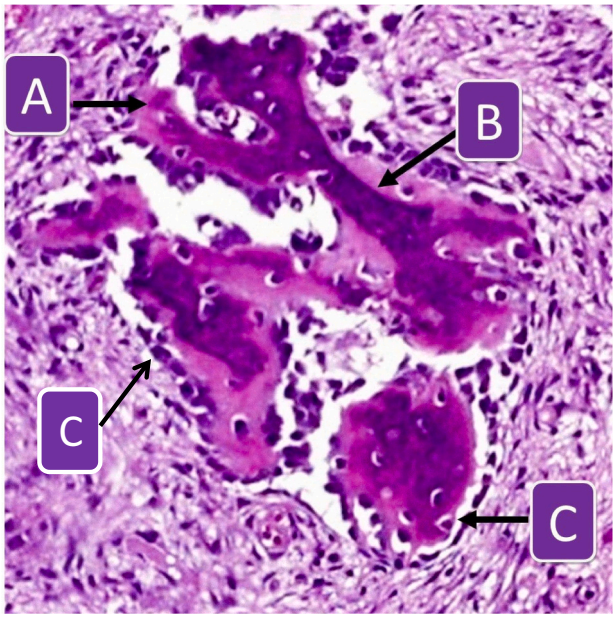

Fig. 3. Histopathological examination of the specimen confirmed for osteofibrous dysplasia; A. c-shaped bony scapulae; B. immature bone trabecuale; and C. osteblastic rimming.

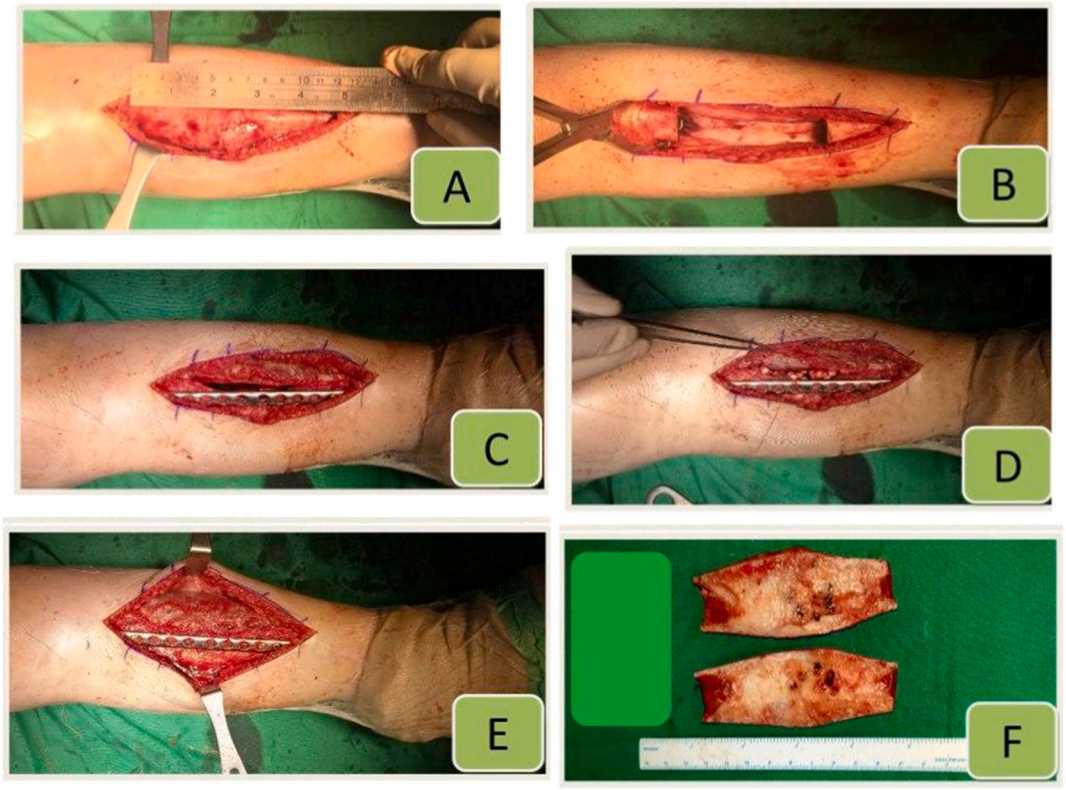

Fig. 4. A. The tumor exposed.
B. En bloc resection with 10 cm bone loss, periosteum was preserved.
C. Internal fixation using plate and screw to maintain the alignment.
D. Application of combination of Bonegener and PerOssal.
E. Suture of periosteum and final construct reconstruction.
F. Macroscopic gross pathology of the tumor.

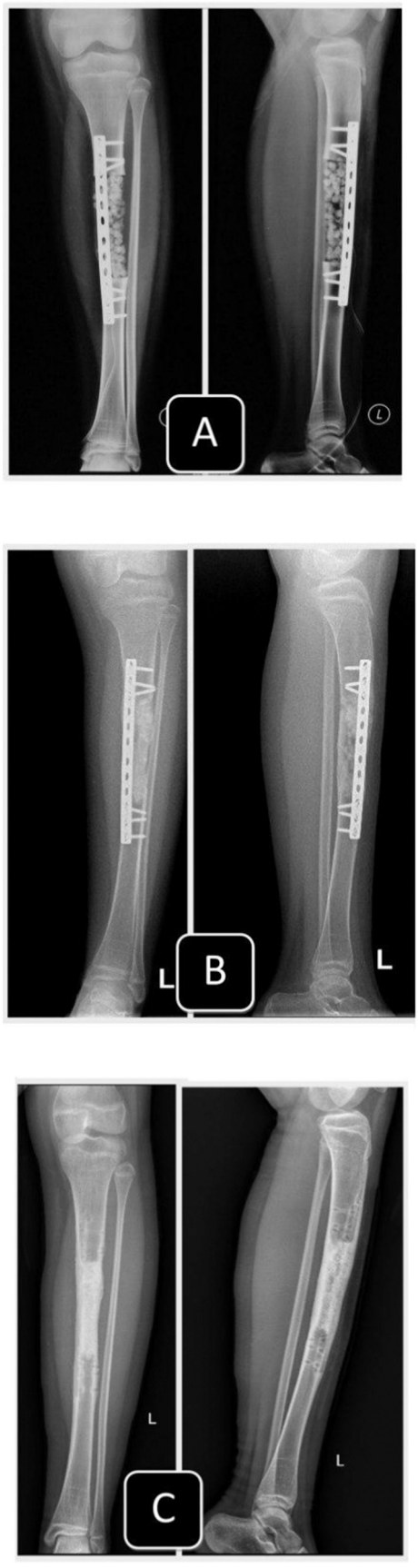

Fig. 5. A. Radiographs post-operative.
B. Follow-up at 20 weeks, callus formation seen.
C. Follow-up at 48 weeks, hard callus formed with optimal stability of tibia.

structural environment for hosting cells to facilitate bone healing process. Hydroxyapatite (HA) is bone major anorganic component which nucleates from specific zones on the collagen fibers deposited by osteoblasts. It has high osteo-conductive potential [12].

As an osteo-inductive agent, biomaterials actively induce new bone formation. Their chemical composition such as calcium phosphate, alumina ceramic, titanium or glass ceramics were classified as osteo-inductive [15,16].

Osteo-genesis of biomaterials actively supports de novo bone formation through the recruitment of osteoblasts. Its stiffness supports osteo-genic differentiation. It provides voids for cell attachment and differentiation and provides stress-relaxation biomechanics for further bone formation. The coupling factors (i.e. growth factors) of the biomaterial attract osteogenic progenitor cells [12,13].

Mechanical stability is obliged to ensure the bone healing. As for this case, an internal fixation using plate and screw was performed to maintain stability of the final construction. Rigid external stimulators can increase cell differentiation and callus formation in the initial period of fracture healing [15].

The authors decided to preserve the periosteum as an important factor to ensure of bone healing during the tumor resection. The periosteum is very critical for callus formation during bone healing process. Moreover, activation of periosteum-derived progenitor cells induces robust chondrogenesis and osteogenesis [16].

However, some disadvantage for this technique include the longer period of time for non-weight-bearing period and higher surgery cost. But, the satisfactory effects to the clinical and radiological outcomes of these techniques outweigh the disadvantages.

4. Conclusion

The chosen treatment for osteofibrous dysplasia has to be the simplest, the safest, and possess the highest rate of bone union. Periosteum preservation followed by reconstruction using biomaterial substitutes combination between demineralized bone matrix and bone substitute “hydroxyapatite” and “calcium sulphate” and internal fixation using plate and screw, can be recommended to overcome the massive bone defect after tumor resection in osteofibrous dysplasia patient.

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Consent

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CRediT authorship contribution statement

AFK – Conceptualization, Methodology, Writing - Reviewing and Editing.

DSR – Writing- Original draft preparation, conceptualization, and Data curation.

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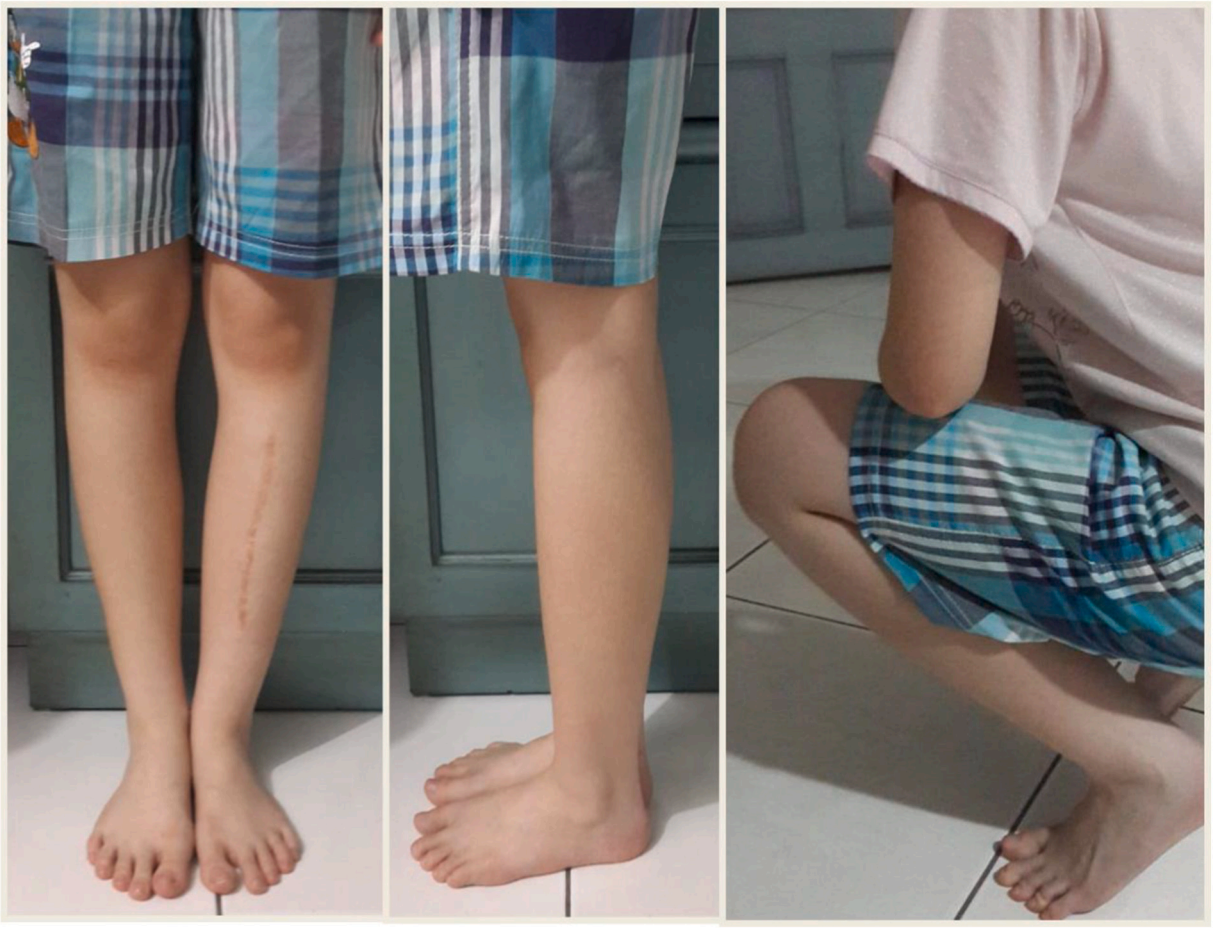
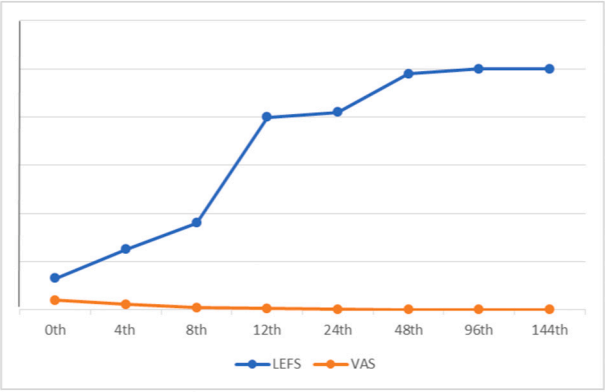


Fig. 6. Clinical condition at 3 years after surgery. The patient could perform highly intensity exercise; full weigh bearing and squatting, scoring 100% of LEFS.



Graph 1. The value of Visual Analog Scale (VAS) and Lower Extremity of Functional Score (LEFS) proposed in follow-up until 3 years (144 weeks) after surgery.

Declaration of competing interest

The authors declare no conflict of interest in this subject matter or material discussed in this case report.

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Ethical approval

Research studies involving patients require ethical approval. Please state whether approval or exemption has been given, name the relevant ethics committee and the state the reference number for their judgement. Please give a statement regarding ethnical approval that will be included in the publication of your article, if the study is exempt from ethnical approval in your institution please state this.

Ethical approval was not required in the treatment of the patient in this report.

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Achmad Fauzi Kamal contributes in the study concept or design, data collection, analysis and interpretation, oversight and leadership responsibility for the research activity planning and execution, including mentorship external to the core team. Didi Saputra Ramang contributes to the study concept or design, data collection and writing the paper.

Conflict of interest statement

All authors must disclose any financial and personal relationships with other people or organisations that could inappropriately influence (bias) their work. Examples of potential conflicts of interest include employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding.

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Guarantor

The Guarantor is the one or more people who accept full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

Prof. Achmad Fauzi Kamal, MD

Research Registration Number

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