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Hereditary Multiple Exostoses (HME) with Peroneal Nerve Compresion: A Case Report

Onarisa Ayu[1,2], Muhammad Iqbal[1,2]

[1]Division of Orthopedic & Traumatology, Department of Surgery, Faculty of Medicine, Syiah Kuala University, Banda Aceh, Indonesia
[2]Dr. Zainoel Abidin Hospital, Banda Aceh, Indonesia
Email: iiqbalmd@gmail.com

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Abstract

Introduction: Hereditary multiple exostosis (HME) is a hereditary disorder characterized by multiple osteochondromas. Clinical symptoms can result from compression of adjacent structures such as peripheral nerves. In Indonesia, HME with nerve compression cases have rarely reported. **Presentation of Case:** An eleven-year-old female with complaining of left knee joint pain and progressive masses in left lower leg since 6 years ago. This complains followed by numbness and difficulty to dorso flexion motion on left ankle joint since four months ago. Physical examination showed of the bony masses was detected at the left lateral upper third lower leg with measuring about six into eight centimeters. Range of motion of left ankle joint patient had difficult to dorso flexion. X-ray imaging viewed demonstrates multiple exostosis appearance involving distal femoral, proximal fibula, proximal tibia and distal fibula bone. MR Imaging revealed cartilage cap of head fibula is thin less 1.5 cm and the axially specimen showed peroneal nerve compression. The patient underwent left head fibula wide resection. Intraoperative findings peripheral nerve peroneal compression and was decompression. Medical rehabilitation for physiotherapy was advised. The results of the follow-up after 2 years, no pain feels and the patient was able to dorso flexion of left ankle joint and no additional bumps in other areas of the body. These lesions may arise from any bone which was pre-formed in the cartilage. Nerve compression syndromes are the neurological complex symptom caused by the mechanical or dynamic compression of a specific single segment. MRI was excellent demonstration of blood vessels compromise and represents choices with peripheral nerves structures and to measuring cartilage cap thickness for criterion of osteochondromas differentiation and exostotic grade. Complete resection was importance of the cartilaginous cap to prevent recurrence. The decompressing the peroneal nerve that pressured by the masses and vascular problems

occured. **Conclusion:** Hereditary multiple exostosis is an inherited disorder characterized by multiple osteochondromas. It is important to monitor all cases of HME especially if the patient complains of pain or growth of an osteochondroma. The surgical excision, with complete resection of the cartilaginous cap of the tumor, is important in preventing recurrence.

Keywords

Osteochondroma, Hereditary Multiple Exostosis (HME), Peroneal Nerve Compression

1. Introduction

Osteochondroma, the most commonly seen bone tumor in children, developmental lesions is truth neoplasm and constitutes 20% - 50% of all benign bones tumour [1]. This is a cartilage-capped exostosis found primarily in the fastest juxta-epiphyses region the tips of long bones that grow [1]. Hereditary Multiple Exostosis (HME), also known as diaphyseal aclasia and multiple osteochondromatosis, characterized by development of multiple osteochondromas (exostosis) and often associated with progressive characteristics bone deformity [2]. This disorder shows autosomal dominant inheritance pattern with approximately two thirds of affected individuals have positive families history [3].

True prevalence of HME is unknown because many patients with asymptomatic lesions have never been diagnosed, hence the estimated prevalence is between 1: 50,000 to 1: 100,000 in the Western population. Previously it was thought to be male dominance, however HME is now appearing to affect both gender in the same way [4].

Clinical symptoms can result from mechanical irritation or compression of adjacent structures such as soft tissues, peripheral nerves, blood vessels and internal organs [5]. In Indonesia, HME with nerve compression cases have rarely been reported. This not a common feature in cases reported in the literature. The purpose of this paper is to report the management of heredity multiple exostoses case with peroneal nerve compression [5].

2. Case Presentation

An eleven-year-old Acehnese female was referred to the Orthopaedic outpatient clinic of Dr. Zainoel Abidin General Hospital with complaints of left knee joint pain and progressive masses in left lower leg since 6 years ago. This complaint is followed by numbness and difficulty to dorsiflex the left ankle joint since four months ago.

Hemodynamic is within normal limit. Physical examination showed of the bony masses was detected at the left lateral upper third lower leg with measuring about six into eight centimeters. Range of motion of knee joint was normal and

lead to pain while moved. Range of motion of left ankle joint patient had difficult to dorso flexion and showed "drop foot".

The left lower leg X-ray Antero-posterior/Lateral view demonstrated multiple exostosis appearance involving distal femoral, proximal fibula, proximal tibia and distal fibula bone (Figure 1). A Magnetic Resonance Imaging (MRI) revealed cartilage cap of head fibula is thin less 1.5 cm and the axially specimen showed peroneal nerve compression (Figure 2). Based on clinical and radiographic we suggest pre-operatively diagnostic is Hereditary Multiple Exostoses with peroneal nerve compression.

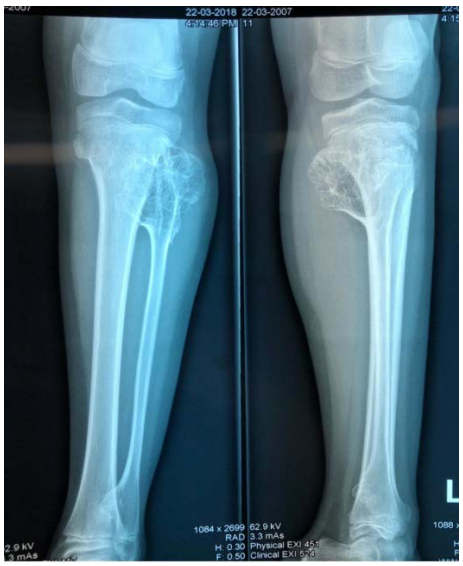

Figure 1. The preoperative radiograph showed exostosis masses at head fibula, lower femur, upper tibia and lower fibula bone.

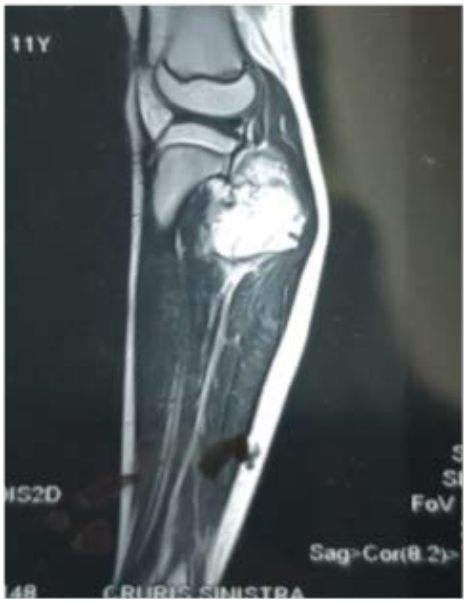

Figure 2. Magnetic resonance imaging revealed cartilage cap of head fibula is thin less 1.5 cm and the axially specimen showed peroneal nerve compression.

Resection of some of the exostoses especially those on fibula proximal was advised. The then patient underwent left upper fibula bone wide resection (Figure 3 and Figure 4). Intraoperative findings peripheral nerve peroneal compression and decompression was performed. Postoperatively, the patient was advised to medical rehabilitation for physiotherapy in outpatient care service. Histopathological examination reported features suggestive of exostosis osteochondroma (Figure 5). The results of the follow-up after 2 years was pain-free, the patient was able to dorsiflex the left ankle joint and no additional bumps in other areas of the body (Figure 6). Informed consent was obtained from the patient's family to report the case.

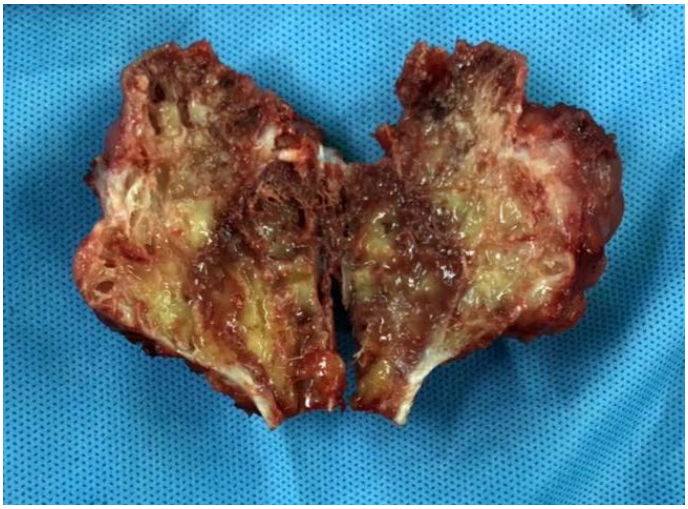

Figure 3. Resection mass mature areas of bone formation contain yellow marrow architecture and areas of calcified cartilage show high attenuation.

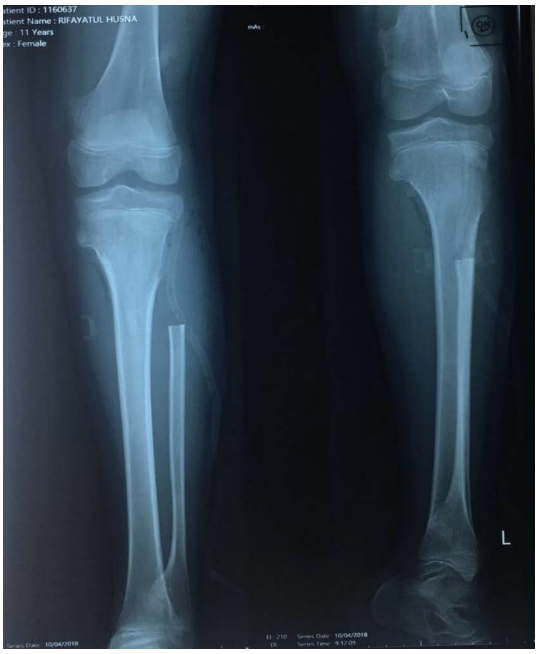

Figure 4. The postoperative radiograph showed osteotomy of head fibula.

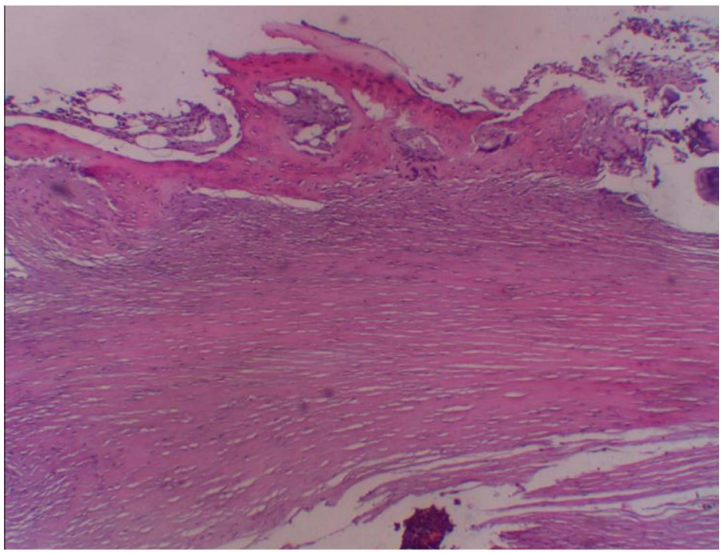

Figure 5. Histopathological examination showing cartilage caps of the osteochondroma on 100× magnification.

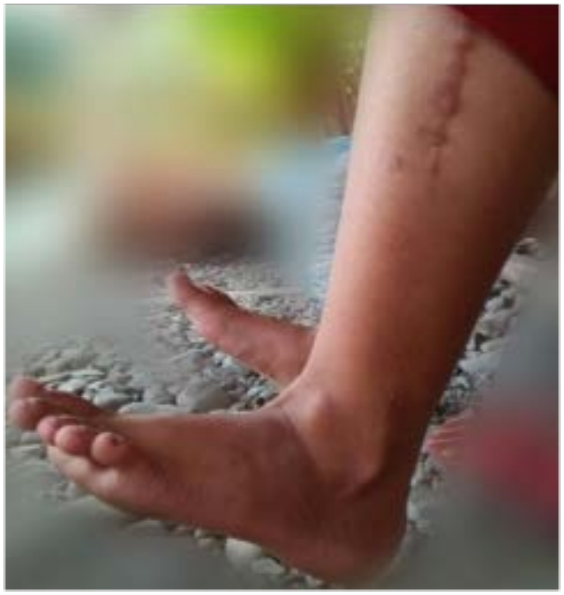

Figure 6. The patient was able to dorsiflex the left ankle joint.

3. Discussion

Hereditary Multiple Exostosis (HME) is a hereditary disorder characterized by multiple osteochondromas. These osteochondromas consist of multiple projections of bone (exostoses) or pedicle of the normal bone covered with proliferating cartilage cells [1]. The lesions are most commonly found in the metaphyses of long bones but may appear on the diaphysis of long bones, the flat bones and the vertebrae. These lesions may arise from any bone which was pre-formed in cartilage [6]. Most of the patients seek treatment for cosmetic reasons. Pain presents secondary to peripheral nerve compression through of osteochondromas or malignancy [5].

Patients rarely presents with local symptoms due to pressure upon surrounding nerves and blood vessels [7]. Nerve compression syndrome are the neurological complex symptom caused by the mechanical or dynamic compression of a specific single segment nerve at specific sites as it passes through a narrow fibro osseous tunnel or an opening in a fibrous or muscular structure [5]. The most common locations are the knees, ankles, shoulders, and wrists [6].

In our case with upper third of fibula HME, the patient had difficulty when moving. Patients also complains of pain and numbness in the lower leg which is a symptom of peroneal nerve compression by the masses, which also makes the left leg difficult to moved. In addition to the compression of the peroneal nerve, vascular disorders in the area can also occur [5]. So that the period in the knee joint is an indication of surgery to decompression of the nerves and blood vessels around masses [8].

Radiographic imaging as a diagnostic tools show cortex and medulla swelling continuous with that of parent bone. In this patient, exostosis appearance in the left upper is third of fibula and lower third of femur [9]. MRI was excellent modality to identify compromised blood vessels, changes of the peripheral nerves structures and measuring cartilage cap thickness for staging of osteochondromas differentiation and exostotic grade [10]. Histopathological examination confirmed the imaging findings. Diagnosis of HME is based on clinical examination and radiographic evaluation, whereas osteochondroma treatments are individual, depending on the presentation [8].

Surgical treatment of osteochondromas consists of simple removal; Mirra reiterated the importance of complete resection of the cartilaginous cap to prevent recurrence [11]. This patient was treatment with surgical wide resection to remove the upper part of the fibula bone that has formed exsostosis tissue, and then decompressing the peroneal nerve that is pressured by the masses and vascular problems occur in this area [8] [11]. So the risk for recurrency can be avoided. This patient showed improvement after resection of the lesion and at two years follow up there is no evidence of recurrence of lesion.

4. Conclusion

Hereditary multiple exostosis is an inherited disorder characterized by multiple osteochondromas [1]. Clinical manifestations are asymptomatic before age two but most commonly presents before the age of 10 [2]. It is important to monitor all cases of HME especially if the patient complains of pain or growth of an osteochondroma [3]. The treatment is individualized, and the best approach of treatment in almost all symptomatic cases is marginal excision of the tumor. The surgical excision, with complete resection of the cartilaginous cap of the tumor, is important in preventing recurrence [11].

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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