Hereditary Multiple Exostoses Complicated By Common Peroneal Nerve Palsy

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Abstract
Hereditary multiple exostoses is an autosomal dominant skeletal disorder. It is characterized by multiple bony prominences and skeletal deformities. It can lead to a series of complications including deformity of the joint, fractures through the tumor pedicle, mechanical block of nearby joints, nerve compression and malignant change. In this case it has been described a rare case of a seventeen-year old male patient with a history of hereditary multiple exostoses presented with foot drop. The management and Physiatric approach are discussed.

Key words: Hereditary multiple exostoses, foot drop, common peroneal nerve palsy

Introduction
Hereditary multiple exostoses (HME) is an autosomal dominant disorder characterized by multiple bony tumors or hamartomas arising near joints. These are cartilage capped and behave in a benign way and are associated with skeletal deformities1. It is the most common skeletal dysplasia, with a frequency of about 1:18,0002. It develops in early childhood and becomes obvious with skeletal development2. The knees, hips, ankles, and shoulders are the most commonly affected sites and growth disturbances are often present, primarily in the forearms and legs3. Foot drop is not an uncommon complication which can occur due to compression neuropathy by the exostosis4.

Case Report
A seventeen year old male patient attended the outpatient department of Physical Medicine and Rehabilitation at Shaheed Suhrawardy Medical College Hospital (ShSMCH) with the complaints of difficulty in walking and lifting right foot for 1 month. The patient also complained of multiple swelling around knees, shoulder, ankle joints, also in ribs and right scapular region. Some of the swellings were painful. Those were small initially and the patient could do the daily activities. However the swellings had increased in size over the last few years and the patient was unable to fully extend the right knee and dorsiflex the right ankle. the patient underwent a surgical excision of a hard lump in the medial aspect of lower right thigh few months back. On physical examination there was a surgical scar mark at the supero-medial aspect of right knee (Fig-1).

Fig-1: Multiple swelling around both knees with a surgical scar at supero-medial aspect of right knee.

There were multiple hard lumps around the knee joints with mild valgus deformity of the right knee as well as right ankle joints. There was also difficulty in knee extension and dorsiflexion of right foot but the peripheral pulses were felt normally. Sensation was impaired at the dorsum of right foot. Neurological examination revealed weakness of the tibialis anterior, lateral peroneal, and

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extensor digitorum muscles. Routine biochemical investigations were done which were found normal. Plain X-ray films showed multiple bony exostoses arising from the proximal end of both tibia and fibula at the both upper and lower end of femur, and also humerus and scapula. The margins were smooth, internal trabeculations were observed. The cortices and marrow of the parent bone were in continuity with the growth (Fig-2,3).

Nerve conduction study was done which revealed conduction block at the site of neck of fibula & distal to that site. Electrophysiological studies confirmed denervation of the muscles supplied by the right common peroneal nerve. CT scan was done with 3D reconstruction which showed multiple bony outgrowth in metaphyseal region pointing away from the joint which was consistent with bony exostosis. CT scan delineated the extent of the lesion and also established unequivocally the continuity of cancellous portions of the lesion and the host bone. These characteristics distinguished this lesion from the occasionally similar-appearing bone masses of osteoma, juxtacortical osteosarcoma, soft tissue osteosarcoma, and juxtacortical myositis ossificans (Fig.4). MRI of right knee was done which showed multiple exostosis in proximal end of tibia and fibula and also in distal end of right femur. The cartilaginous cap showed a high signal intensity on T2-weighted sequences. A narrow band of low signal intensity surrounding the cap representing the perichondrium was also seen (Fig-5). Based on history, physical examination, and different imaging modalities a diagnosis of foot drop in a case of HME was confidently made. The patient was managed accordingly by the drugs, physical therapy and orthosis. To relieve the pain, oral analgesic was used. In physical therapy electrical stimulation was given in right leg.

Different exercises such as strengthening exercise, and range of motion exercise were prescribed. Wax bath was also applied to the right knee followed by stretching exercise to reduce the flexion deformity.

The patient was advised crutch, walking stick, and ankle foot orthosis subsequently as per requirement along with Gait training. The patient came to the department on wheel
Discussion
Hereditary multiple exostoses constitute an autosomal dominant condition with variable penetrance. In this disease, osteochondroma of many bones are caused by an anomaly of skeletal development. The disease occurs only about 5% to 10% as often as solitary osteochondroma and is more common in males. Approximately 66% of affected individuals have a positive family history. The specific genetic abnormalities have recently been detected, with three distinct loci on chromosomes 8, 11, and 19. The lesions are usually discovered at about 2 years of age. The knees, hips, ankles, and shoulders are the most commonly affected sites and growth disturbances are often present, primarily in the forearms and legs. Malignant degeneration is extremely rare. Osteochondromas can lead to a series of complications including fractures through the tumor pedicle, mechanical block of nearby joints, and nerve compression.

Conclusion
The common peroneal nerve palsy is not very uncommon in case of hereditary multiple exostoses. Though surgery may be required to repair the damaged muscles, tendons and nerves to reduce the severity of the impairment the physiatric intervention plays an important role in the neurological recovery.

References
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