INTRODUCTION
Hereditary multiple exostoses (HME) is an autosomal dominant skeletal disorder characterized by the presence of multiple osseous prominences. It can occur sporadically or within families (22 – 56%). Two genes, EXT1 and EXT2 located respectively at 8q24 and 11p11-p12, have been isolated to cause HME. It can cause gross deformity of limbs and growth disturbance which is quite a common complication. Malignant transformation to chondrosarcoma can also occur. Neurological presentations are rare and usually happen due to direct compression of a peripheral nerve or nerve root or less often the spinal cord. This case is possibly the first case of HME described from Bangladesh, presented with dorsal cord compression. Decompression was done and the complaints of myelopathy were improved.

CASE REPORT
A 16 years boy presented with the complaint of multiple bony swelling on different sites for the last 4 years. The swelling first appeared over right forearm and successively involved proximal tibia, distal femur, arms and forearms, right shoulder, chest and metacarpal bones (proximal phalanx of right index and middle finger). The swelling were fixed, hard, painless and continuous with underlying bone. Some of them were increasing in size and some halted at one point. The last swelling appeared over right index finger 6 months back. He complained of gradual bowing of his right arm over 2 years and recently became inactive due to restricted movement. He also developed urinary incontinence for the last one month. He had no significant past history of trauma and nothing contributory regarding his family.

On examination, he appeared of short stature (height 154 cm) according to his age. There were multiple bony projection, arising commonly from the metaphysis of long bones. Complications of this disease are growth disturbance and malignant transformation to chondrosarcoma. Rarely, the disease may cause peripheral nerve or nerve root and spinal cord compression.

HME is a rare disorder and cord compression is the rarest complication of the disease which influenced the authors to report it.

Hereditary Multiple Exostoses Causing Cord Compression
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ABSTRACT
Hereditary multiple exostoses (HME) is an autosomal dominant skeletal disorder characterized by the presence of multiple osseous prominences with cartilage caps, arising commonly from the metaphysis of long bones. Complications of this disease are growth disturbance and malignant transformation to chondrosarcoma. Rarely, the disease may cause peripheral nerve or nerve root and spinal cord compression.

HME is a rare disorder and cord compression is the rarest complication of the disease which influenced the authors to report it.

Key words: Hereditary Multiple Exostoses (HME). Exostoses. Cord compression.

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Received December 28, 2010; accepted April 16, 2012.
department for decompression. MRI showed vertebral compression between thoracic 9th and 10th and 11th and 12th vertebra level. After surgery, complaints of myelo-pathy were significantly improved.

The patient was counselled about the hereditary nature of disease and of regular follow-up to detect malignant transformation and other complications earlier.

**DISCUSSION**

The disease has a series of synonyms like ‘multiple hereditary osteochondromata’, ‘exostotic dysplasia’ etc. Prevalence is estimated at 1:50,000 persons within the general population and seems to be higher in males (1.5:1) probably because females tend to have a milder phenotype and are, therefore, easily overlooked. The disease may occur sporadically or within the previously affected family. Approximately 22 – 56% of the patients with HME can have a positive family history based on EXT 1 or EXT 2 mutation. This boy might be a sporadic case or the first case of his generation. Two genes, EXT1 and EXT2 located respectively at 8q24 and 11p11-p12, have been isolated to cause HME. Additional linkage to chromosome 19p has been found, suggesting the existence of an EXT3-gene too.

Exostoses can involve any bone with predilection to long bones of lower extremities. It rarely involves face, tarsal and carpal bone, vertebrae and sternum. In this case involvement of carpal bones were present. These osteochondromas increase in size in the first decade of life, ceasing to grow when the growth plates close at puberty. Majority of them are asymptomatic and covered by thickened periosteum. The marrow cavity is continuous with the cavity of the underlying bone. A variety of deformities can be found like shortening of the ulna with secondary bowing of radius; (39 – 60%), limb length discrepancy (10 – 50%), varus or valgus angulations of the knee (8-33%), disproportionate short stature (37 – 44%) etc. all of which were present in this case. Important differential diagnoses of HME is metachondromatosis (MC), another autosomal dominant disease. Here, osteochondromas characteristically occur in the hands and feet, predominantly the digits and toes, and point toward the adjacent growth plate, while in HME the osteochondromas are mainly located in the long or tubular bones and point away from the epiphysis which happened in this case. Moreover, MC do not cause any shortening or deformity of limbs and the size of the osteochondroma regress spontaneously both clinically and radiologically around the age of puberty. Dysplasia epiphysealis hemimelica (DEH) is another important differential but here the lesion predominantly affects the lower extremity and restricted on one side of the body which disfavours the condition in this case. Significant complication of HME is malignant transformation (0.5 – 5%) leading to development of secondary peripheral chondrosarcoma (94% cases). The suspicion is indicated by growth of the tumour after puberty, the presence of pain, or a thickness over one cm of the cartilaginous cap in adults. Compression over adjacent spinal cord is another rare complication which happened in this case.

HME do not hamper life expectancy. Diagnosis at early age, proper explanation of the problem and keeping registry of affected family is very important. Symptomatic problem relief, genetic counselling about future family planning and regular follow-up to avoid complications is the mainstay of management.

**REFERENCES**


