Bilateral cubitus varus due to ulnar exostoses- a case report

Peer review status:
No

Corresponding Author:
Dr. Mohit K Jindal,
Senior Resident, ESI PGIMSR Delhi, 2150/2 Ballabgarh (haryana), 121004 - India

Submitting Author:
Dr. Mohit K Jindal,
Senior Resident, ESI PGIMSR Delhi, 2150/2 Ballabgarh (haryana), 121004 - India

Article ID: WMC005019
Article Type: Case Report
Article URL: http://www.webmedcentral.com/article_view/5019
Subject Categories: ORTHOPAEDICS
Keywords: bilateral cubitus varus ; manus varus; negative ulnar variance; Multiple hereditary exostoses

How to cite the article: Jindal MK. Bilateral cubitus varus due to ulnar exostoses- a case report. WebmedCentral ORTHOPAEDICS 2015;6(11):WMC005019

Copyright: This is an open-access article distributed under the terms of the Creative Commons Attribution License (CC-BY), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Source(s) of Funding:
institutional funded

Competing Interests:
Nil
Bilateral cubitus varus due to ulnar exostoses- a case report

Author(s): Jindal MK

Abstract

Osteochondroma or exostosis is most common primary benign bony tumor comprising of more than one third of the total occurrences. Osteochondromas are considered as an aberration in the normal physiological growth plate and originate from the metaphysis of long bone with more than third (35-46%) of cases affecting the bone around the knee (Lower end femur> Upper end Tibia), 10% cases involve the small bones of the hand and 5% involve the pelvis and flat bones like scapula (4-6%) are least involved. These tumors usually affect the growing skeleton and cease to increase in size after skeletal maturity. These are usually painless and do not cause any symptoms or functional impairment. Hereditary multiple exostosis is an autosomal dominant disorder manifested by the presence of multiple osteochondromas. Although exostoses are benign lesions, they are often associated with characteristic progressive skeletal deformities and may cause clinical symptoms. The most common deformities include short stature, limb-length discrepancies, valgus deformities of the knee and ankle, asymmetry of the pectoral and pelvic girdles, bowing of the radius with ulnar deviation of the wrist, and subluxation of the radiocapitellar joint.

CASE REPORT

A 8 year old girl came to our out patient department with bilateral cubitus varus deformity with associated manus varus bilaterally since 3 years duration. A skiagram of bilateral elbow joints and wrist revealed bilateral ulnar osteochondromas with associated ulnar shortening and radial bowing. Other skiagrams were done to reveal any other bony exostoses. There was no associated family history. The condition caused no functional impairment to the patient clinically and the patient was explained regarding the condition and kept for regular 6 monthly follow up to look for any increase in deformity or development of newer deformities.

DISCUSSION AND CONCLUSION We concluded that surgical management in a case of bilateral elbow and hand deformities due to multiple exostoses is not always required and should be delayed till the patient attains the age of at least 12 years as it will prevent post surgical recurrence of deformities and newer deformities which develop can be tackled simultaneously.

Introduction

Osteochondroma is the most common primary benign bone tumor usually occurring near the end of long bone usually solitary (90%), may be multiple in the form of hereditary multiple exostosis (HME) in about 10% of cases. Hereditary multiple exostoses is a condition in which people develop multiple benign exostoses. The number of exostoses and the bones on which they are located vary greatly among affected individuals. The exostoses are not present at birth, but approximately 96 percent of affected people develop multiple exostoses by the time they are 12 years old. Exostoses typically form at the end of long bones and on flat bones such as the hip and shoulder blade. Multiple exostoses can disrupt bone growth and can cause growth disturbances of the arms, hands, and legs, leading to short stature. Often these problems with bone growth do not affect the right and left limb equally, resulting in uneven limb lengths (limb length discrepancy). Bowing of the forearm or ankle and abnormal development of the hip joints (hip dysplasia) caused by exostoses can lead to difficulty walking and general discomfort. Multiple exostoses may also result in pain, limited range of joint movement, and pressure on nerves, blood vessels, the spinal cord, and tissues surrounding the exostoses.

Case Report(s)

A 8 year old girl came to our out patient department with bilateral cubitus varus deformity with associated manus varus bilaterally since 3 years duration. There was no antecedent history of trauma or growth disturbances suggestive of rickets. A skiagram of bilateral elbow joints and wrist revealed bilateral ulnar osteochondromas with associated ulnar shortening and radial bowing. Other skiagrams were done to reveal any other bony dysplasia. There was no restriction in supination and pronation. There was no associated family history suggestive of multiple hereditary exostoses. The condition was non symptomatic and caused no functional impairment to the patient. The patient was explained regarding the
condition and kept for regular 6 monthly follow up to look for any increase in deformity or development of newer deformities.

Discussion

Hereditary multiple osteochondromas is a rare disorder that affects bone growth. Bony tumors (exostoses or osteochondromas), covered with cartilage, typically appear in the growth zones (metaphyses) of the long bones adjacent to the areas where tendon and muscles attach to the bone. These growths vary in size and number among affected individuals, even within the same family. Some individuals will present with a few large “lumps” while others will show several small growths. The median age of diagnosis is three years and almost all affected individuals are diagnosed by 12 years of age.

In many cases, no treatment is required. If the exostoses are small, they may have little or no effect on the patient. However, in more severe cases, the growths may cause deformities of the forearm, knees, ankles, spine and/or pelvis. They may impose upon nerves, tendons and/or blood vessels, and interfere with movement or circulation, causing substantial pain as a result of pinched nerves or compressed tendons.

Bones that develop exostoses most often are the upper arm (humerus), forearm, knee and shoulder blades (scapulae). Bowing of the forearm and ankle are the problems that most often require surgical correction. Approximately 40 percent of affected individuals have mild short stature as a result of shortened and/or bowed legs.

Multiple hereditary exostoses affecting the forearm was described by Masada et al. These are Type I, Type IIa, Type IIb and Type III. Of these type I had a distal ulna exostosis with Distal Radio Ulnar Joint (DRUJ) disrupted; Type IIa had distal ulna and proximal radius exostosis and the Proximal Radio Ulnar Joint (PRUJ) and DRUJ disrupted; Type IIb had distal ulna exostosis and both the PRUJ and DRUJ are disrupted. Type III had distal radius exostosis only with both PRUJ and DRUJ intact.

Our case belonged to Type 1 as per the Masada classification and as such would require a surgical intervention at a later juncture and hence kept for 6 monthly follow up to monitor the increase in deformity as well as development of newer deformities which would develop till the age of 12 years of age.

Conclusion

We concluded that surgical management in a case of bilateral elbow and hand deformities due to multiple exostoses is not always required and should be delayed till the patient attains the age of at least 12 years as it will prevent post surgical recurrence of deformities and newer deformities which develop can be tackled simultaneously.

References

1. Illiterate patients: greater care and caution required in the Indian context. Med law Cases Doct, vol 5 issue page 180

Illustration

1. Clinical photograph showing Bilateral cubitus and manus varus bilaterally.
2. Skiagram of bilateral elbow and wrist showing bilateral ulnar exostoses resulting in cubitus and manus varus deformities bilaterally.
Illustrations

Illustration 1

Clinical photograph showing Bilateral cubitus and manus varus bilaterally.

Illustration 2

Skiagram of bilateral elbow and wrist showing bilateral ulnar exostoses resulting in cubitus and manus varus deformities bilaterally.