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THE UNUSUAL PRESENTATION OF EXCESSIVE SPURS LIKE OSTEOCHONDROMAS IN AN ADOLESCENT WITH HEREDITARY MULTIPLE OSTEOCHONDROMAS

Orthopaedics

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ABSTRACT

Hereditary Multiple Osteochondromatosis (HMO) is characterised by the presence of multiple osteochondromas, mainly affecting the long bones. Osteochondromas are bony projections that are either sessile or pendulated in form. Isolated osteochondromas in the form of spurs have been described as occurring in the knee and the foot. Reports of thorn-like osteochondromas affecting the ribs are extremely rare. We describe the unique case of an adolescent with multiple osteochondromas affecting the long bones. While in the lower limbs they were noted to be sessile and pendulated, they had the form of excessively elongated spurs in the radius. They created pain and limitation of his activities. The surgical removal of these spurs enabled us to restore activity in the patient's dominant (right) arm.

KEYWORDS

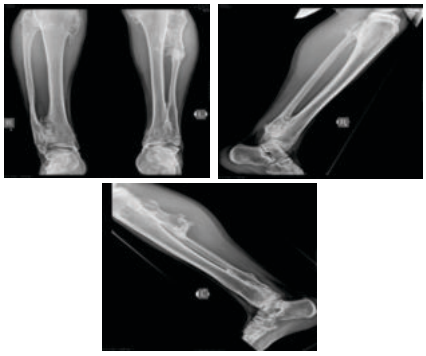
Hereditary Multiple Osteochondromatosis, spurs like osteochondroma, exostosis, forearm, HMO.

INTRODUCTION

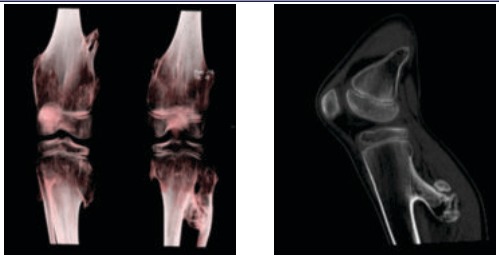
Hereditary Multiple Osteochondroma (HMO) is a genetic condition indicated by the presence of multiple osteochondromas. Osteochondromas are bony projections that are covered from hyaline cartilage. They have a marrow cavity that is in continuity with the medullary canal of the host bone. The most commonly affected areas are the long bones, femur, tibia, humerus and forearm bones (radius and ulna). Osteochondromas increase in size during the patient's developing years and cease growing when the epiphyseal plate is consolidated. [1-4] Two types of osteochondromas are described: sessile and pendulated. The sessile type has a broad base on the bone, while the second is growing from a pendulum, described as a cauliflower shape, with calcifications in the osteochondroma. Spurs that resemble bony projections have been observed mainly in adolescent athletes as isolated bone formations, causing irritation and pain due to tendon or nerve irritation. [5-7] Spurs of osteochondromas in HMO have been reported as affecting the ribs and are extremely rare. [8-10] We present the unique case of a patient with HMO affecting all the long bones displaying the typical sessile and pendulous types of osteochondromas. In the right forearm, he developed elongated spurs of osteochondromas, causing discomfort in daily activities. The spurs were surgically removed, following which he returned in his daily activities. These kind of elongated spurs have not been previously reported.

Case Report

A 14-year-old adolescent was under our supervision for the past 4 years for multiple osteochondromas that were affecting femur as well as the tibia and fibula. His father had also multiple osteochondromas—he is an active engineer who has never been operated for the osteochondromas. The adolescent patient had multiple large osteochondromas mainly affecting the knee and ankle joint. The osteochondromas displayed a typical sessile shape, while in the upper part of the fibula a pendulated shape was observed. **Figures 1-5**



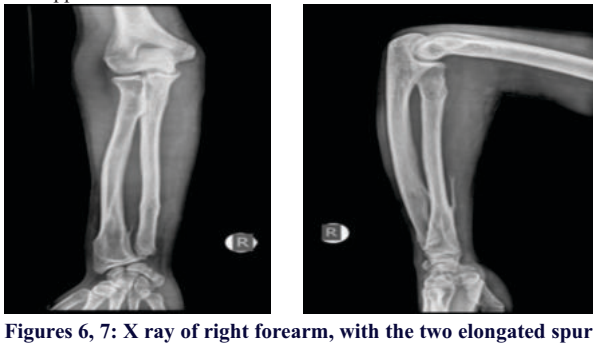
Figures 1-3: X-ray AP and lateral of tibia and fibula, with multiple sessile and pendulum type osteochondromas



Firures 4,5 CT scan of the affected knees, with multiple osteochondromas. The pendulated type of osteochondroma is displayed in figure 5.

The axis of the lower limbs was not affected. There was no leg length discrepancy. The large osteochondromas of the ankle joint had created deformities of the shape of the fibula and tibia. The boy complained of pain and discomfort of his right hand. He had been experiencing discomfort during his daily activities for the past 6 months, including writing, as he was right handed. His feeling was that there was a needle in the forearm, when he was moving his wrist and his fingers. On examination of the forearm, ulnar deviation of the wrist was found. His elbow has full flexion and extension. The wrist joint also displayed full range of movement.

He resisted examination to perform pronation and supination movements as he found them painful. On careful palpation, the bony projections were identified in the forearm. An x-ray evaluation of the forearm showed two enormous, elongated spur-type osteochondromas growing in the typical direction away from the epiphyseal plate. They had arisen from the radius. The ulna displayed reduced length, affecting the distal part of the bone. The proximal part of the radius and ulna appeared normal.



Figures 6, 7: X ray of right forearm, with the two elongated spur like osteochondromas. There is shortening of the distal part of the ulna.

We surgically removed the spurs with meticulous preparation in between the FCR and BR. The small base of the spur was in continuation with the medulla of the radius. Specimens were sent for pathology and the presence of mature bone was confirmed, as is the case with osteochondromas.

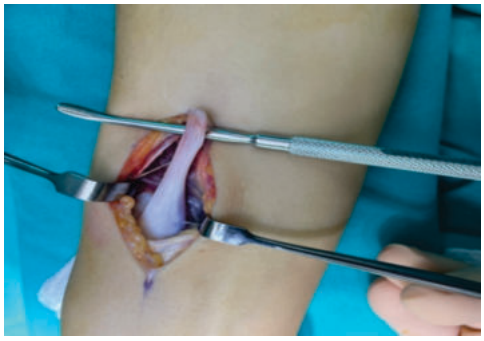


Figure 8 : Intra operative figure of the palmarly displayed elongated osteochondroma

The boy had an uneventful recovery and returned to his activities in 3 weeks. He is under our supervision for the osteochondromas in the lower limbs.

DISCUSSION

Osteochondromas usually appear either as either pendulated or sessile, with a broad base. They are found mainly in long bones. In the forearm, they cause the shortening of the ulna, deformity and bowing of the radius, and radial head dislocation. Early evaluation and recognition of deformities is essential for children whose forearm bones have been affected by HMO.[1,3,4]

Masada et al. described three types of deformities in the forearm for HMO (the second with two subtypes) based on the site of the osteochondromas and the presence of radial head dislocation. Our patient may be characterised as type I due to the shortening of the ulna. Otherwise with the radius not being bowed and the location of the osteochondromas in the distal part of the radius may be as the type III classification.[11]

Treatment methods for forearm osteochondromas remain controversial. Early removal of osteochondromas, mainly in the distal part of the ulna, is considered beneficial and may prevent the ulnar shortening and radius bowing. Unfortunately, this treatment is not always sufficient, and deformity has the tendency to recur. In established ulna shortening with radial head dislocation, Ilizarov procedures with ulna lengthening and relocation of the radial head are demanding, but display promising results. We have treated children with type IIb Masada with the Ilizarov technique with very good results. Yan and Nan have recently proposed the osteotomy of the ulna to be placed proximally in the ulna, where the interosseous connection is weak and there is higher proportion of cancellous bone. [12-14]

Spurs like osteochondromas are described in children as occurring around the knee joint and in the feet of young footballers. They may present with irritation of adjacent muscles, tendons, nerves or vessels. There is a danger of fracture mainly during strenuous athletic activities. [5,6,7,15]

Osteochondromas have been reported as early signs of HMO when found in the ribs. Nursat et al. reported the case of a 2-year-old boy who presented with bony overgrowths on the chest and recurrent respiratory infections. The diagnosis of osteochondromas affecting the ribs and the scapula was confirmed with CT scan. The positive family history of HMO was helpful to confirm the right diagnosis. There are few case reports on the presence of osteochondromas in the ribs as a cause of chest pain or pneumothorax. [16-18]

Thorn-like exostoses in the ribs, causing thoracic hemorrhage are extremely rare. [10] Dhamarshaktu reported thorn-like exostosis in the knee of a 23-year-old patient as a different type of osteochondroma. [19] Atypical osteochondromas affecting the hand have been reported.[20]

Our patient presents a unique case of elongated spur-type osteochondroma among the typical osteochondromas in the limbs.

The Orthopedico Rizzoli has proposed a classification of HMO in three classes based on the presence of deformities and functional limitations of the affected sites. In their large series of patients, there is no mention of spur-type of osteochondroma. [21] Our patient had a deformity of the forearm with ulnar shortening, but the functional limitation was due to the presence of two elongated spur type osteochondromas that were acting like thorns. Surgical treatment provided permanent relief from his discomfort.

CONCLUSION

Multiple osteochondromas appeared displaying elongated spurs-type exostosis. Despite the fact that the affected forearm had only a minor deformity, the osteochondromas acted as thorns, creating discomfort for the adolescent patient. Surgical removal of the osteochondromas restored the function of the forearm.

REFERENCES

1 Bovée JV. Multiple osteochondromas. Orphanet J Rare Dis. 2008 Feb 13;3:3. doi: 10.1186/1750-1172-3-3.PMID: 18271966 Review

2 Mordenti M, Shih F, Boarini M, Pedrini E, Gnoli M, Antonioli D, Tremosini M, Sangiorgi L. The natural history of multiple osteochondromas in a large Italian cohort of pediatric patients. Bone. 2020 Oct;139:115499. doi: 10.1016/j.bone.2020.115499. Epub 2020 Jun 24. PMID: 32592948.

3 Tepelenis K, Papathanakos G, Kitsouli A, Troupis T, Barbouti A, Vlachos K, Kanavaros P, Kitsoulis P. Osteochondromas: An Updated Review of Epidemiology, Pathogenesis, Clinical Presentation, Radiological Features and Treatment Options. In Vivo. 2021 Mar-Apr;35(2):681-691. doi: 10.21873/invivo.12308.PMID: 33622860 . Review

4 Pacifici M. Hereditary Multiple Exostoses: New Insights into Pathogenesis, Clinical Complications, and Potential Treatments. Curr Osteoporos Rep. 2017 Jun;15(3):142-152. doi: 10.1007/s11914-017-0355-2.PMID: 28466453

5 Turati M, Bigoni M, Omeljaniuk RJ, Griffet J, Zatti G, Courvoisier A.n Pediatric navicular dorsal osteochondroma: A rare case of navicular-cuneiform impingement. J Pediatr Orthop B 2019;28:602-6.

6 Fraser RK, Nattrass GR, Chow CW, Cole WG: Pes anserinus syndrome due to solitary tibial spurs and osteochondromas. J Pediatr Orthop. 1996, 16:247–8. 10.1097/00004694-199603000-00022

7 Tiwari V, Sampath Kumar V, Poudel R R, et al. (July 05, 2017) Pes Anserinus Bursitis due to Tibial Spurs in Children. Cureus 9(7): e1427. DOI 10.7759/cureus.1427

8 Simansky DA, Paley M, Werczberger A, Bar Ziv Y, Yellin A. Exostosis of a rib causing laceration of the diaphragm: diagnosis and management. Ann Thorax Surg. 1997;63:856e857.

9 Reynolds JR, Morgan E. Hemothorax caused by a solitary costal exostosis. Thorax. 1990;45:68e69.

10 Cheng PG, Chen CC, Wu SK, Hsu SM, Wang MN. Thorn-like osteochondroma presenting as hemothorax in an adult. Formos J Surg 2012;45:97-9.

11 Masada K, Tsuyuguchi Y, Kawai H, Kawabata H, Noguchi K, Ono K. Operations for forearm deformity caused by multiple osteochondromas. J Bone Joint Surg Br. 1989, 71: 24–9.

12 Farr S, van der Zwan AL, Kommol E; EPOS Upper Limb Study Group and Collaborators. Reliability of the Masada and Jo classifications for multiple hereditary exostoses in the forearm. J Hand Surg Eur Vol. 2021 Mar;46(3):318-320. doi: 10.1177/1753193420954356. Epub 2020 Sep 13.PMID: 32924768

13 Yan G, Nan G. Modified osteotomy for treatment of forearm deformities (Masada IIb) in hereditary multiple osteochondromas: a retrospective review. BMC Musculoskelet Disord. 2021 Nov 10;22(1):943. doi:10.1186/s12891-021-04829-x. PMID: 34758801

14 Li Y, Wang Z, Chen M, Cai H. Gradual ulnar lengthening in Masada type I/IIb deformity in patients with hereditary multiple osteochondromas: a retrospective study with a mean follow-up of 4.2 years. J Orthop Surg Res. 2020 Dec 9;15(1):594. doi:10.1186/s13018-020-02137-z. PMID: 33298090.

15 Ugai K, Sato S, Matsumoto K, Matsubara T, Mizuno K, Hirohata K: A clinicopathologic study of bony spurs on the pes anserinus. Clin Orthop Relat Res. 1988, 231:130–4. 10.1097/00003086-198806000-00018

16 Nusrat K, Mahmood S, Khan M, Khan A. An Unusual Presentation of Bony Outgrowths on the Chest: A Case from Pakistan. Cureus. 2020 Mar 18;12(3):e7309. doi: 10.7759/cureus.7309.PMID: 32313750

17 Mazza D, Fabbri M, Calderaro C, et al.: Chest pain caused by multiple exostoses of the ribs: a case report and a review of literature. World J Orthop. 2017, 8:436-440. 10.5312/wjo.v8.i5.436

18 Maeda K, Watanabe T, Sato K, et al.: Two cases of asymptomatic rib exostosis treated by prophylactic surgical excision. J Pediatr Surg Case Rep. 2017, 20:24-28.

19 Dharmshaktu GS, Pangtey T, Bhandari SS. Thorn-like exostoses: A rare presentation of hereditary multiple exostoses. J Musculoskelet Surg Res 2018;2:137-8.

20 Mukhtar IA, Alqasim E, Abdulkareem I, Prabhu J. Atypical presentation of hereditary multiple osteochondromas: Thirteen years of follow-up and analysis. J Musculoskelet Surg Res 2018;2:73-6.

21 Mordenti M, Gnoli M, Boarini M, Trisolino G, Evangelista A, Pedrini E, Corsini S,Tremosini M, Staals EL, Antonioli D, Stilli S, Donati DM, Sangiorgi L. The Rizzoli Multiple Osteochondromas Classification revised: describing the phenotype to improve clinical practice. Am J Med Genet A. 2021 Nov;185(11):3466-3475. doi: 10.1002/ajmg.a.62470. Epub 2021 Sep 3. PMID:34477285