

Growth Differentiation of Scapular Exostosis In Relation To Its Position

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Abstract: Osteochondroma or exostosis is a benign tumor arising from the bone involving the long bones most commonly, pelvis, ribs and the scapula at rare instances. It can run in families called as multiple hereditary exostosis (MHE) which is different from solitary exostosis in terms of malignant transformation. MHE has a higher chance of malignant transformation. In our study 7 cases of solitary scapular exostosis were studied and followed up. The size of the tumor in relation to the forces acting on it were assessed. Pain was also a feature in this study without malignant transformation, but suspicion of malignancy needs needle biopsy and later complete excision as like any malignant tumour.

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I. Introduction

Exostosis or osteochondroma is a benign tumor arising from the bone usually noticed before physal closure and growth stops with cessation of growth. Malignant transformation is suspected if growth continues after physal closure or presence of pain. Other causes of pain are when there is formation of bursae around the tumor, compressing on neurovascular structures and fracture of the tumor at the pedicle. Scapular exostosis are rare and can occur on the dorsal and ventral aspect. Excision and biopsy of the tumor is the treatment of choice if malignancy is not suspected.

II. Aim

To assess the growth differentiation of scapular exostosis in relation to its position and structures surrounding it.

III. Materials And Methods

This is a prospective study done during the period 2015 to 2018. It is a prospective and a retrospective study in which 7 cases of scapular exostosis were studied pre operatively, intra operatively and postoperatively. All cases were clinically diagnosed to be solitary scapular exostosis and radiological impressions were also obtained. X-ray and magnetic resonance imaging was done and the size of the tumor was measured approximately. All patients were between the age group of 12 -21. 1 patient had complaints of pain all others had come for a cosmetic deformity. Radiology showed physal closure in all the patients. 6 patients underwent excision and biopsy, 1 patient with pain underwent biopsy and then excision.

IV. Results

Out of the 7 patients, 5 were dorsal exostosis and 2 were ventral exostosis.

DORSAL	5
VENTRAL	2

1 patient had complaints of pain during movements of the scapula.

CHIEF COMPLAINTS	COSMETIC DEFORMITY	PAIN
	6	1

V. Discussion

In our study, 7 cases of scapular exostosis were diagnosed clinically, radiologically and underwent excision biopsy. 1 case had pain during scapular movements for which patient underwent core needle biopsy followed by excision. All 7 cases were histopathologically proved to be osteochondroma.

Sizes of all the excised tumors were compared and it was found that ventral tumors are smaller than dorsal. Even though it was not significant, a special mention should be made here as ventral tumors are pressured by the muscles and bones surrounding it, the growth of the tumor can be potentially reduced. Intra operative observation shows that ventral exostosis are more rounded with smooth surface where as, dorsal tumors are bigger in size and have an irregular surface.

One patient had pain which was suspected malignancy for which biopsy was done followed by excision, the pain was due to friction between the tumor and surrounding structures. All patients were radiologically and clinically followed up for one year and none of the patients had any signs of recurrence. Patient with pain also became symptom free after excision.



Figure 1



Figure 2

A case of ventral exostosis (figures 1 and 2) which is smaller, well rounded and has a smooth surface when compared to dorsal exostosis (figures 3,4 and 5).



Figure 3

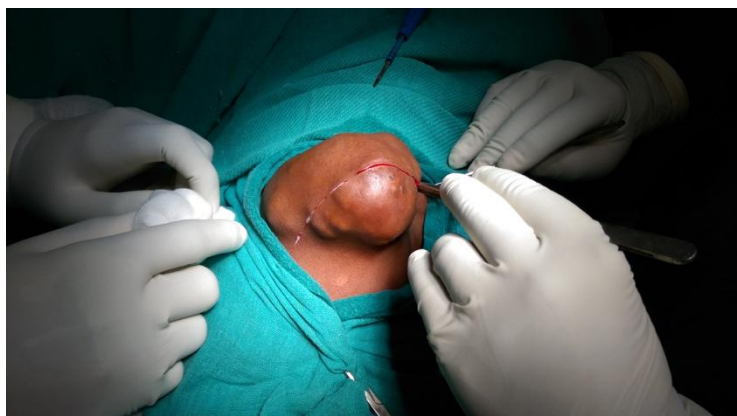


Figure 4



Figure 5

VI. Conclusion

1. Excision biopsy is the treatment of choice for exostosis of scapula unless there is a suspicion of malignant transformation.
2. Biopsy followed by excision is better option of management even for the slightest doubts of malignant transformation.
3. **Prabhakar's theory of histogenesis:**Based on the observation that ventral exostosis is small, more organized and with a smooth surface, whereas the dorsal exostosis is irregular, large and mushroom like. I would like to postulate that osteochondroma even though is a true neoplasm, recent studied revealed that, it is due to mutation in the gene encoding exostosin 1 (EXT1), it still behaves like a physis, which under pressure and loading produces small rounded organized growth. Whereas an unloaded osteochondroma produces bizarre growth. The tumor may represent a perverted activity of the periosteum, which reverts to its role as perichondrium.

This theory is supported by Virchow in 1891 and Plate defect theory by Keith in 1920.

References

- [1]. Campanacci M. Bone and soft tissue tumors. Bologna: Springer-Verlag Wien GmbH; 1990.
- [2]. Schmale GA, Conrad EU, III, Raskind WH. The natural history of hereditary multiple exostoses. *J Bone Joint Surg Am.* 1994;76A:986e92. [[PubMed](#)]
- [3]. Ryckx A, Somers JF, Allaert L. Hereditary multiple exostosis. *Acta Orthop Belg.* 2013;79(6):597–607.[[PubMed](#)]
- [4]. Shapiro F, Simon S, Glimcher HJ. Hereditary multiple exostoses. Anthropometric, roentgenographic, and clinical aspects. *J Bone Joint Surg Am.* 1979;61A:815e24. [[PubMed](#)]
- [5]. Wicklund CL, Pauli RM, Johnston D, Hecht JT. Natural history study of hereditary multiple exostoses. *Am J Med Genet.* 1995;55:43e6. [[PubMed](#)]
- [6]. Porter DE, Lonie L, Fraser M, et al. Severity of disease and risk of malignant change in hereditary multiple exostoses. A genotype-phenotype study. *J Bone Joint Surg Br.* 2004;86B:1041e6. [[PubMed](#)]
- [7]. Stickens D, Clines G, Burbee D, et al. The EXT2 multiple hereditary exostoses gene defines a family of putative tumor suppressor genes. *Nat Genet.* 1996;14:25–32. [[PubMed](#)]
- [8]. Jennes I, Pedrini E, Zuntini M, et al. Multiple osteochondromas: mutation update and description of the multiple osteochondromas mutation database (MOdb) *Hum Mutat.* 2009;30(12):1620–1627. [[PubMed](#)]

- [9]. Szuhaik K, Jennes I, de Jong D, et al. Tiling resolution array-CGH shows that somatic mosaic deletion of the EXT gene is causative in EXT gene mutation negative multiple osteochondromas patients. *Hum Mutat.* 2011;32(2):E2036–E2049. [[PubMed](#)]
- [10]. Goud AL, Lange J, de Scholtes VAB, et al. Pain, physical and social functioning and quality of life in individuals with hereditary multiple exostoses in the Netherlands. A national cohort study. *J Bone Joint Surg Am.* 2012;94A:1013e20. [[PubMed](#)]
- [11]. Malini K, Gudi Narayan S, Kutty AVM, et al. Mutational Analysis of Exostosin 1 and 2 Genes in Multiple Osteochondroma. *Indian J Pediatr.* 2015;82(7):649–650. [[PubMed](#)]
- [12]. Kang Qing-lin, Xu Jia, Zhang Zeng, et al. Mutation Screening for the EXT1 and EXT2 Genes in Chinese Patients with Multiple Osteochondromas. *Arch Med Res.* 2013;44(7):542–8. [[PubMed](#)]
- [13]. Jones KB, Piombo V, Searby C, et al. A mouse model of osteochondromagenesis from clonal inactivation of Ext1 in chondrocytes. *Proc Natl Acad Sci USA.* 2010;107(5):2054–2059. [[PMC free article](#)][[PubMed](#)]
- [14]. Reijnders CM, Waaijer CJ, Hamilton A, et al. No haploinsufficiency but loss of heterozygosity for EXT in multiple osteochondromas. *Am J Pathol.* 2010;177(4):1946–1957. [[PMC free article](#)] [[PubMed](#)]
- [15]. Hecht JT, Hogue D, Strong LC, et al. Hereditary multiple exostosis and chondrosarcoma: linkage to chromosome 2 and loss of heterozygosity for EXT-linked markers on chromosomes 2 and 8. *Am J Hum Genet.* 1995;56:1125–1131. [[PMC free article](#)] [[PubMed](#)]
- [16]. Lind T, Tufaro F, McCormick C, et al. The putative tumor suppressors EXT1 and EXT2 are glycosyltransferases required for the biosynthesis of heparan sulfate. *J Biol Chem.* 1998;273:26265–26268. [[PubMed](#)]
- [17]. McCormick C, Leduc Y, Martindale D, et al. The putative tumour suppressor EXT1 alters the expression of cell-surface heparan sulfate. *Nat Genet.* 1998;19:158–161. [[PubMed](#)]
- [18]. Busse-Wicher M, Wicher KB, Kusche-Gullberg M. The exostosin family: proteins with many functions. *Matrix Biol.* 2014;35:25–33. [[PubMed](#)]
- [19]. Zak BM, Schuksz M, Koyama E, et al. Compound heterozygous loss of Ext1 and Ext2 is sufficient for formation of multiple exostoses in mouse ribs and long bones. *Bone.* 2011;48(5):979–987. [[PMC free article](#)] [[PubMed](#)]
- [20]. Jones KB, Pacifici M, Hilton MJ. Multiple hereditary exostoses (MHE): elucidating the pathogenesis of a rare skeletal disorder through interdisciplinary research. *Connect Tissue Res.* 2014;55(2):80–8. [[PubMed](#)]
- [21]. Stickens D, Brown D, Evans GA. EXT genes are differentially expressed in bone and cartilage during mouse embryogenesis. *Dev Dyn.* 2000;218:452–464. [[PubMed](#)]
- [22]. Benoist-Lassel C, de Margerie E, Gibbs L, et al. Defective chondrocyte proliferation and differentiation in osteochondromas of MHE patients. *Bone.* 2006;39:17–26. [[PubMed](#)]
- [23]. Stickens D, Zak BM, Rougier N, et al. Mice deficient in Ext2 lack heparan sulfate and develop exostoses. *Development.* 2005;132:5055–5068. [[PMC free article](#)] [[PubMed](#)]
- [24]. Pedrini E, Jennes I, Tremosini M, et al. Genotype-phenotype correlation study in 529 patients with multiple hereditary exostoses: identification of “protective” and “risk” factors. *J Bone Joint Surg Am.* 2011;2011(93):2294e302. [[PubMed](#)]
- [25]. Clement ND, Porter DE. Hereditary multiple exostoses: anatomical distribution and burden of exostoses is dependent upon genotype and gender. *Scott Med J.* 2014;59(1):35–44. [[PubMed](#)]
- [26]. Hennekam RC. Hereditary multiple exostoses. *J Med Genet.* 1991;28:262e266. [[PMC free article](#)][[PubMed](#)]
- [27]. Mordenti M, Ferrari E, Pedrini E, et al. Validation of a new multiple osteochondromas classification through Switching Neural Networks. *Am J Med Genet.* 2013;161A:556–560. [[PubMed](#)]
- [28]. Darilek S, Wicklund C, Novy D, et al. Hereditary multiple exostosis and pain. *J PediatrOrthop.* 2005;2005(25):369e76. [[PubMed](#)]
- [29]. Ali S, Kaplan S, Kaufman T, et al. Madelung deformity and Madelung-type deformities: a review of the clinical and radiological characteristics. *PediatrRadiol.* 2015;45:1856–1863. [[PubMed](#)]
- [30]. Masada K, Tsuyuguchi Y, Kawai H, et al. Operations for forearm deformity caused by multiple osteochondromas. *J Bone Joint Surg Br.* 1989;71B:24e9. [[PubMed](#)]
- [31]. Woodside CJ, Ganey T, Gaston RG. Multiple osteochondroma of the hand: initial and long-term follow-up study. *HAND.* 2015;10:616–620. [[PMC free article](#)] [[PubMed](#)]
- [32]. Wang YZ, Park KW, Oh CS, et al. Developmental pattern of the hip in patients with hereditary multiple exostoses. *BMC MusculoskeletDisord.* 2015;16:54. [[PMC free article](#)] [[PubMed](#)]
- [33]. Vaishya R, Swami S, Vijay V, et al. Bilateral total hip arthroplasty in a young man with hereditary multiple exostoses. *BMJ Case Rep.* 2015 doi: 10.1136/bcr-2014-207853. [[PMC free article](#)] [[PubMed](#)][[CrossRef](#)]
- [34]. Clement ND, Porter DE. Can deformity of the knee and longitudinal growth of the leg be predicted in patients with hereditary multiple exostoses? A cross-sectional study. *The Knee.* 2014;21:299–303. [[PubMed](#)]

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