

BILATERAL MEDIAL END OF CLAVICLE ENCHONDROMAS. AN UNUSUAL PRESENTATION.

Zeeshan Khan, Ummara Siddique Umer, Shahjehan Alam, Syed Ghulam Ghaus

Orthopaedic Oncology & Radiology Department, Rehman Medical Institute, Peshawar, Pakistan.

PJR October - December 2018; 28(4): 326-328

ABSTRACT

We report a case of 17 years old male who presented with a sternoclavicular swelling. X ray revealed large lytic lesions in bilateral medial clavicle ends. MRI showed heterogeneous large exophytic mass in left clavicle and a smaller lesion in right clavicle. Perioperative and histopathological findings were consistent with enchondroma. Clinical, radiographic and pathological investigations were necessary to establish the diagnosis. Main stay of surgical treatment for symptomatic lesions is detailed intralesional curettage with or without bone grafting. To our knowledge, this cartilaginous tumor isolated in clavicle has never been previously reported in this location and with bilateral presentation.

Introduction

The clavicle is a rare site for primary bone tumors and little literature is available regarding their characteristics and long term outcomes. Relatively few clavicular neoplasms are localized to the medial one-third of the bone.¹⁻⁴ This area is particularly challenging because of the peculiar anatomy of the clavicle and its close association with the important neurovascular structures.

Enchondromas are benign cartilaginous neoplasms that are usually solitary lesions in intramedullary bone and are mostly present in long bones.⁵ The clavicle shares its oncological characteristics with flat bones and not with other long bones, so the chances of enchondroma being present in clavicle are very little.² The enchondroma lesions replace normal bone with mineralized or un mineralized hyaline cartilage, thereby generating a lytic pattern on radiographs or, more commonly, a lytic area containing rings and arcs of chondroid calcifications. We report a rare case of bilateral Enchondromas arising from the medial end of clavicles of a 17 yr old male. We could only find

one reported case in the English literature of periosteal chondroma and to our knowledge, this is the first reported case of isolated bilateral clavicle enchondromas.

Case Report

A 17 year old male patient presented to the orthopedic oncology clinic with a painless swelling just above the left clavicle for the last 18 months with no history of trauma or any associated features. The patient was otherwise fit and well with no significant past medical or family history. Physical examination revealed a hard, non-tender swelling over the left sternoclavicular joint with normal overlying skin. Plain chest X-ray revealed large ovoid lytic lesions in bilateral medial clavicle ends giving appearance of large punched out defects, which were more pronounced on left side (Fig. 1).

Cross sectional imaging with MRI images showed a

Correspondence : Dr. Ummara Siddique
Orthopaedic Oncology & Radiology Department
Rehman Medical Institute,
Peshawar, Pakistan.
Email: ummara_81@hotmail.com

Submitted 30 October 2018, Accepted 9 November 2018

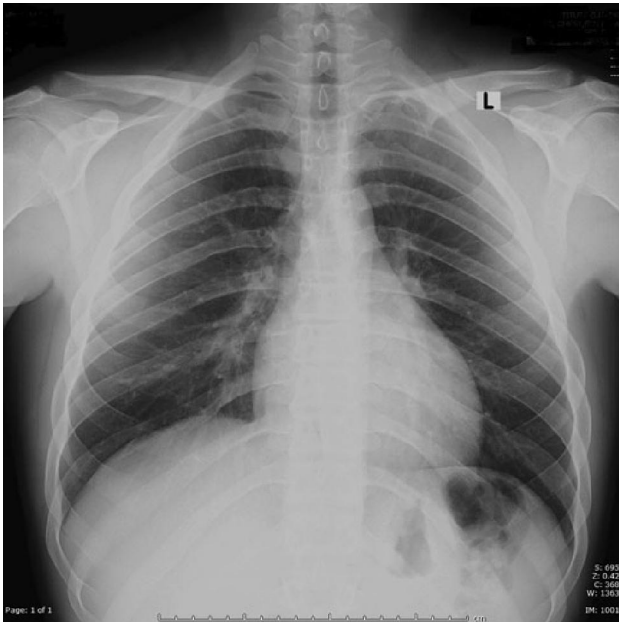


Figure 1: Plain radiograph chest showing lytic lesions in the medial ends of clavicle, more pronounced on the left side.

large 3.0x4.7x4.3cm heterogenous mass in medial sternal end of left clavicle involving the articular surface and bulging into the sternoclavicular joint space (Fig. 2). There were heterogeneous hyperintensities on T2 weighted images with central fluid signals. It was hyper intense to muscles on T1 weighted images and showed heterogeneous post contrast



Figure 2: Axial T1 weighted image depicting the destructive lesion in clavicle.

enhancement. Part of the mass was bulging into the infraclavicular fossa with mass effect on left subclavian vessels. No definite vessel invasion or extension into pleural cavity was seen. Mild edema was seen in adjacent marrow of manubrium sterni. T1 weighted images revealed another smaller hypo intense lesion in medial end of right clavicle involving its inferior wall and sternal articular end. There was subtle post contrast enhancement. Considering bilateral clavicle involvement, major differentials on imaging were eosinophilic granulomas and brown tumour. Core needle biopsy of the left clavicle under image intensifier revealed cartilaginous material on gross appearance with histopathological examination confirming enchondroma with no evidence of malignancy (Fig. 3). Further review of cross sectional imaging did not reveal any endosteal scalloping. The patient and family were counselled about the diagnosis and is completely asymptomatic at 9 months with yearly follow up planned.



Figure 3: Gross appearance of specimen obtained from left clavicle at biopsy revealing pearly white cartilaginous material consistent with chondroid lesion.

Discussion

The presence of island of ectopic cartilaginous tissue surrounded by bone is called an “enchondroma.” It may be due to abnormal proliferation of chondrocytes as in Olliers disease (OD) or misdirected growth as in Metachondromatosis or failure of reabsorption of chondrocytes at enchondral ossification as in Spondyloenchondrodysplasia. The solitary enchondroma is

mostly detected incidentally as radiolucent lesion on plain radiograph and is mostly seen in long tubular bones.⁶ The lesions likely arise from cartilaginous nests that are displaced from the growth plate.⁵ Clavicle is a less known site for enchondromas.

More than one enchondromas is associated with different disorders. Genochondromatosis, an extremely rare autosomal dominant disorder and is characterized by bilateral symmetrical lesions, normal stature, and no skeletal deformities.⁷ Based on the involvement of clavicle, two types of Genochondromatosis have been described both having benign course.⁸⁻¹⁰ Type I has lesions at the metaphysis of the proximal humerus, distal femur and medial end of clavicle while type II has normal clavicle with involvement of metaphysis of the short tubular bones of the hand, wrist and feet.¹⁰⁻¹² Because of limited number of cases reported, genes and molecular defects responsible for Genochondromatosis have not been elucidated.⁹

Isolated enchondromas are managed symptomatically and if there size increases more than 5 centimeters, it is advisable that they should have cross sectional imaging to look for any endosteal scalloping, which can represent chondrosarcomas. Larger than 5 cm enchondromas are also called CLUMP (Cartilage lesion of unknown malignant potential) and they should be followed up radiologically on an annual basis.

Conflict of Interest: Authors declare that they have no conflict of interest.

References

1. Smith J, Yuppa F, Watson RC. Primary tumors and tumor-like lesions of the clavicle. *Skeletal Radiol* 1988; **17**: 235-46.
2. Dahlin DC, Unni KK. *Bone Tumors: General Aspects and Data on 8542 Cases*. Springfield, IL, Charles C Thomas, 1986, ed 4.
3. Barlow IW, Newman RJ. Primary bone tumours of the shoulder: An audit of the Leeds Regional Bone Tumour Registry. *J R Coll Surg Edinb* 1994; **39**: 51-4.
4. Sudhir Kapoor, Akshay Tiwari, and Saurabh Kapoor. Primary tumours and tumorous lesions of clavicle. *Orthop*. 2008; **32(6)**: 829-34.
5. Semenova LA, Bulycheva IV. Chondromas (enchondroma, periosteal chondroma, enchondromatosis). *Arkh Patol*. 2007; **69(5)**: 45-8.
6. Sareen A, D'souza MM, Reddy KB, Kanojia RK, Kumar A. Genochondromatosis type I: A clinico-radiological study of four family members. *Am J Med Genet* 2017; **167**: 2758-66.
7. Le Merrer M, Fressinger P, Maroteaux P. Genochondromatosis. *J Med Genet* 1991; **28**: 485-9.
8. Pansuriya TC, Kroon HM, Bovee JV. Enchondromatosis: Insights on the different subtypes. *Int J Clin Exp Pathol* 2010; **3**: 557-69.
9. Superti-Furga A, Spranger J, Nishimura G. Enchondromatosis revisited: new classification with molecular basis. *Am J Med Genet* 2012; **160C**: 154-64.
10. Lee YS, Dan J, Ryu KJ, Kim BK, Han S-H, Kim HJ. Case of Genochondromatosis Type I in an 8-year-old Boy. *Am J Med Genet* 2013; **161A**: 1513-6.
11. Kozlowski K, Jarrett J. Genochondromatosis II. *Pediatr Radiol* 1992; **22**: 593-5.
12. Isidor B, Guillard S, Hamel A, Le CC, David A. Genochondromatosis type II: Report of a new patient and further delineation of the phenotype. *Am J Med Genet* 2007; **143A**: 1919-21.
13. Pansuriya TC, van Eijk R, d'Adamo P, van Ruler MAJH, Kuijjer ML, Oosting J, Cleton-Jansen A-M, van Oosterwijk JG, Verbeke SLJ, Meijer D, van Wezel T, Nord KH. Somatic mosaic IDH1 and IDH2 mutations are associated with enchondroma and spindle cell hemangioma in Ollier disease and Maffucci syndrome. *Nature Genet* 2011; **43**: 1256-61.
14. Amary MF, Damato S, Halai D, Eskandarpour M, Berisha F, Bonar F et al. Ollier disease and Maffucci syndrome are caused by somatic mosaic mutations of IDH1 and IDH2. *Nature Genet* 2011; **43**: 1262-5.