

Congenital Pseudoarthrosis of Clavicle-Rare Case

Dr Shrish Pratap Singh (DMRD)¹, Dr Archit Gupta²

D-352 Railway Colony Overbridge Road, Jhansi, Uttar Pradesh-284003

²MD Radiodiagnosis, Dr B.Y.L Nair Ch. Hospital, Mumbai
Room No-803-H building, Nair Hospital, Mumbai Central, Mumbai

Abstract: *Congenital pseudoarthrosis of clavicle (CPC) is a rare malformation. It usually presents with a swelling in the midportion of the clavicle. The diagnosis can be confirmed by X-ray. Surgery is indicated in symptomatic patients or patients with major deformity. We present a case of the right side CPC in a 14-year-old female patient. Usual presentation of these patients is cosmetic deformity. Congenital pseudoarthrosis of clavicle should be included in the differential diagnosis of clavicle abnormalities in pediatric age group.*

1. Introduction

Congenital pseudoarthrosis of the clavicle (CPC) was first diagnosed in 1910 in a 8-year-old girl with a defect in her right clavicle but without any other skeletal manifestation. Patients most commonly present as a painless mass seen over the right clavicle[1]. Radiographic analysis reveals a separation in the mid-third of the clavicle with both fragments characteristically having round, smooth edges[1]. According to Hans,[2] hormonal or environmental disruption results in CPC. The pathological findings are described as a true pseudoarthrosis with cartilage covering the bone ends. Synovial fluid is present in the capsule encasing the pseudoarthrosis. Etiology is unknown but most suitable hypothesis that the subclavian artery may compress the developing right clavicle, which might explain right sided predominance and the occurrence of the left-sided lesion in patients with dextrocardia. Bilateral involvement is rare.[3,4]

The differential diagnosis of CPC includes birth fracture, nonunion of a fractured clavicle, and neurofibromatosis. The pseudoarthrosis is usually not painful, and shoulder range of motion is normal. Radiographs of the pseudoarthrosis reveal an osseous separation with enlarged, rounded bone ends, and a distinctive absence of fracture callus.

2. Case Report

We present this case of a 14 year old female who presented to us with a chief complaint of a painless mass over the right clavicle [Figure 1]. This deformity was present since birth but not undergone any treatment. There was no history of birth trauma.

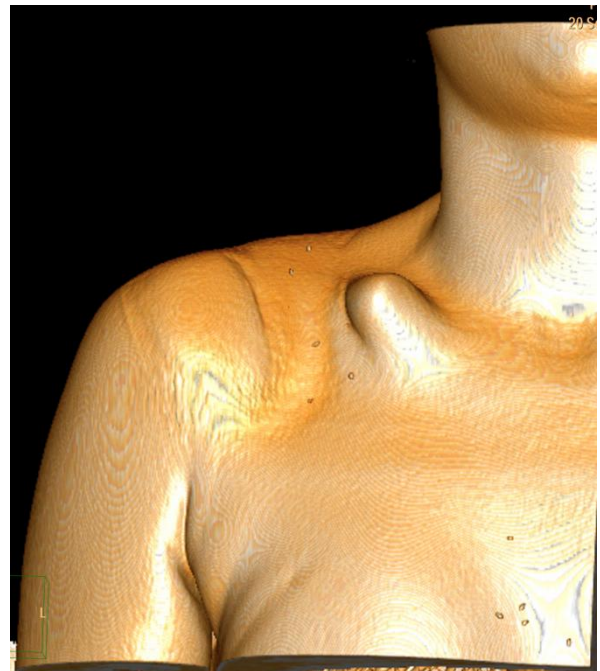


Figure 1: Showing external swelling in right clavicular region on VRT image

On initial clinical examination, there was deformity present on the middle one-third of the right clavicle. It was nontender with full range of motion of the right shoulder and the distal neurovascular status was normal. There was no evidence of any other deformity. On radiological examination, pseudoarthrosis of the right middle third of clavicle was seen [Figure 2].



CXR PA VIEW demonstrates pseudoarthrosis in the mid of right clavicle. Rest CXR is normal. Further CT Right clavicle was done which typically demonstrates separation of right clavicle into two with both fragments characteristically having round, smooth edges.(Figure 3,4 and 5)

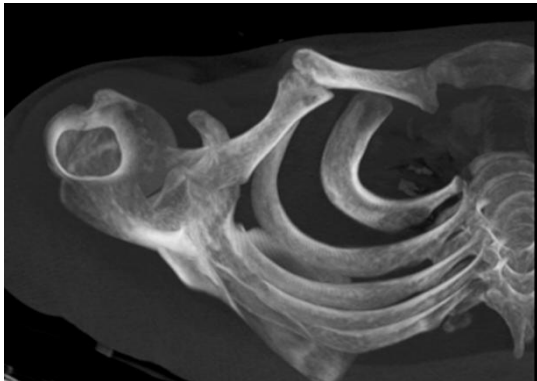


Figure 3: MIP images superior view

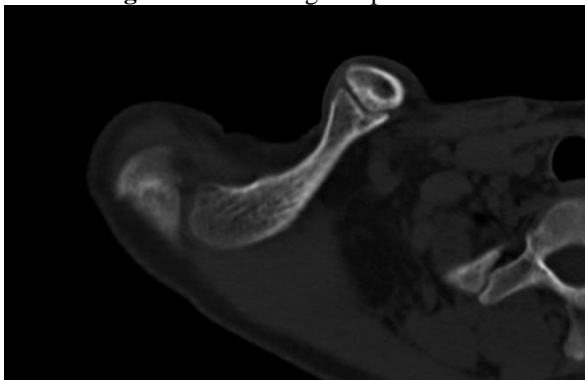


Figure 4: Axial CT images



Figure 5: VRT images of right clavicle

3. Discussion

CPC is a rare congenital disorder with right sided predominance. CPC may sometimes cause limitations of shoulder girdle function. Usually no pain or symptoms of neurovascular compression is present. Most common complaint is Cosmetic disfigurement. The degree of severity of deformity was directly related to amount of motion at pseudoarthrosis. To achieve union, bone grafting is necessary. The optimal age for grafting as being in the 2-4-year old period.[5] In girls over 8 years of age, trimming of the mass was the only treatment recommended. Jinkins[6] also proposed early stabilization as the treatment of choice.

Treatment of CPC varies depending on the severity of the deformity, age, and sex of child. The patients with no alteration in shoulder girdle biomechanics, and minimal cosmetic deformity, should be observed periodically. If increased cosmetic deformities develop and alteration of shoulder girdle function occurs, surgical stabilization should be performed before the age of 8 years. The recommended surgery would be resection of the pseudoarthrosis and stabilization by an autogenous iliac bone graft and K-wire or screw fixation. The hardware should be removed after the fusion is complete. The surgical fusion is usually successful especially between ages 2-4 years.[5,7,8]

4. Conclusion

CPC is a rare anomaly that usually occurs in the right clavicle and presents as a painless and cosmetically unappealing mass. The greater the motion at pseudoarthrosis of clavicle, more will be cosmetic deformity. Patients with mild deformity are usually observed, however surgical resection and stabilization is required in those with a severe deformity.

References

- [1] Wall JJ. Congenital pseudoarthrosis of the clavicle. *J Bone Joint Surg Am* 1970;52:1003-9.
- [2] Hans G. The Genesis of Skeletal Abnormalities, 2nd International Conference on Congenital Malformations. The International Medical Congress Ltd.; 1964. p. 219-23.
- [3] Lloyd-Roberts GC, Apley AG, Owen R. Reflections upon the aetiology of congenital pseudoarthrosis of the clavicle. With a note on cranio-cleidodysostosis. *J Bone Joint Surg Br* 1975;57:24-9.
- [4] Fairbank HA. Cranio-cleido-dysostosis. *J Bone Joint Surg Br* 1949;31B:608-17.
- [5] Alldred AJ. Congenital pseudoarthrosis of the clavicle. *J Bone Joint Surg Br* 1963;45-B:312-9.
- [6] Jinkins WJ Jr. Congenital pseudoarthrosis of the clavicle. *Clin Orthop Relat Res* 1969;62:183-6.
- [7] Toledo LC, MacEwen GD. Severe complication of surgical treatment of congenital pseudoarthrosis of the clavicle. *Clin Orthop* 1979;139:64.
- [8] Grogan DP, Love SM, Guidera KJ, Ogden JA. Operative treatment of congenital pseudoarthrosis of the clavicle. *J Pediatr Orthop* 1991;11:176-80.