

Case Report

Congenital pseudarthrosis of the clavicle in an adult: case report and literature review

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ABSTRACT

A 27 years old male, presented with a bump over the middle third of his right clavicle and a foreshortened shoulder girdle causing a severe deformity. Radiographs revealed a Congenital pseudarthrosis of the clavicle (CPC). A resection of the pseudarthrosis was carried and internal fixation with a reconstruction plate was done using iliac graft. One year after he was asymptomatic, without any functional impairment and very satisfied with the final cosmetic result. There is no consensus about optimal treatment. We present a rare case of surgical in an adult patient, and demonstrate why treatment must be individualized and surgical associated complications must be discussed.

Keywords: Pseudoarthrosis, Clavicle, Childhood, Congenital, Adult

INTRODUCTION

Congenital pseudarthrosis of the clavicle (CPC) is a rare condition first described by Fitzwilliams in 1910.¹ It is characterized by a definitive bone defect usually in the middle third of the clavicle.² It is more frequent in female patients, and the right side is the most commonly affected.²⁻⁴ When found on the left side, it is usually associated with dextrocardia, situs inversus or a left cervical rib.^{3,4} Unilateral involvement occurs in approximately 90% of cases.³

It is usually discovered in the first months of life, and is typically asymptomatic.² This entity may be present in familiar syndromes simultaneously such as cleidocranial dysplasia or neurofibromatosis.³ The aim of this report is to describe a case of CPC in an adult, and patient and do a brief discussion based on literature.

CASE REPORT

A 27 years old male was referred to our orthopaedic clinic because of a clavicular deformity. He had no previous history of trauma, including no obstetric trauma. There was no evidence of other congenital anomalies in his family. Presents for a long time with a bump over the middle third of his right clavicle and a foreshortened shoulder girdle causing a severe deformity (Figure 1).



Figure 1: Anterior view of patient's clavicle deformity before surgery.

He had no discomfort or pain at palpation of the clavicular area, had a functional shoulder range of motion (ROM), 120° abduction, 180° of forwarding elevation, 180° rotation, and less strength in abduction compared with the left side. The radiographs revealed a clavicular pseudoarthrosis presented by no callous formation and sclerotic borders (Figure 2).



Figure 2: Radiograph demonstrating right CPC.

We proposed surgical treatment to improve shoulder aesthetic and strength taking into account shoulder and donor bone graft site scar, and the risk of other surgical consequences. Despite that, the deformity was the biggest concern of the patient, so he accepted. Iliac crest graft, with 2.5 cm of length and 1cm of thickness, was used to restore bone length. We proceeded to direct anterior-superior approach, Judet decortication, iliac crest graft interposition, alignment of the fragments, and fixation with an anatomical clavicular plate (Figure 3).

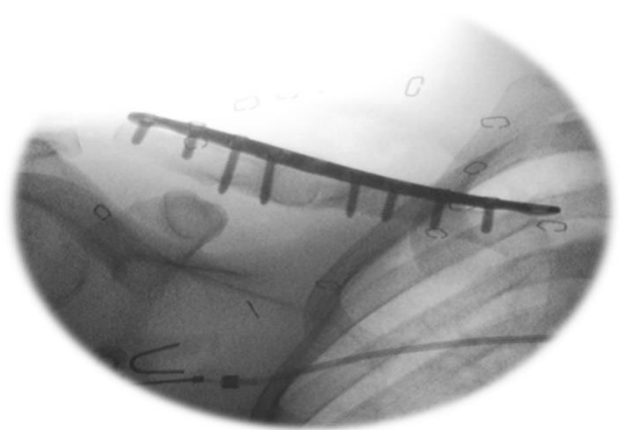


Figure 3: Intraoperative radiograph after CPC resection, graft interposition and fixation with an anatomical clavicular plate.

Histology of the tissues collected revealed a false joint with ends covered by hyaline cartilage. He evolved favorably, with no pain, showing callus formation at 6 weeks (Figure 4) of evolution and complete radiographic healing ensued after 12 weeks (Figure 5).

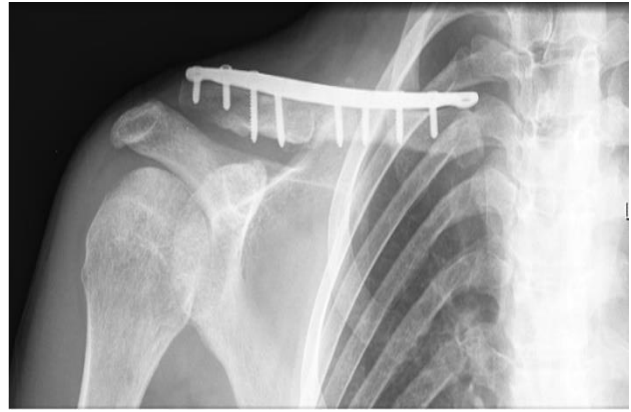


Figure 4: Postoperative radiograph at 6 weeks.

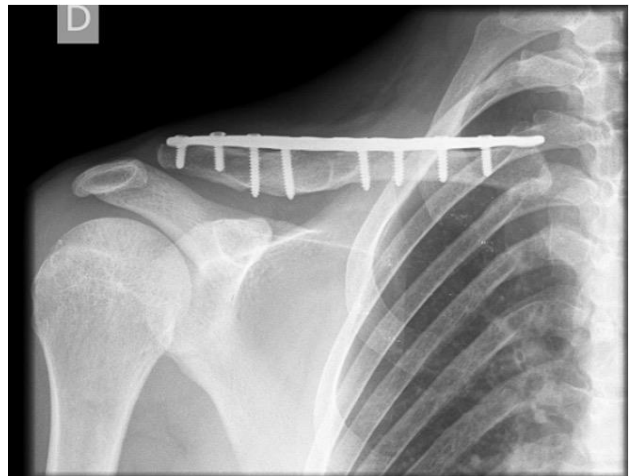


Figure 5: Radiographic healing ensued after 12 weeks.

At this stage, he started with physical rehabilitation to restore total ROM. One year after he presented with excellent clinical and radiological result, completely asymptomatic, without functional impairment (>120° abduction, 180° of forwarding elevation, 180° rotation and symmetrical strength) and was very satisfied with the final cosmetic result (Figure 6).



Figure 6: Deformity correction result.

DISCUSSION

The pathogenesis of CPC remains unclear; however, the embryonic theory is the most commonly accepted. The clavicle is the first fetal bone to undergo primary ossification (diaphyseal membranous ossification).^{3,5} It has 2 primary points of ossification that normally start to fuse at 7th week.³ In 1975, Lloyd-Roberts et al⁶ described failed union due to subclavian artery pressure that runs posteriorly and inferiorly to the midportion of the clavicle. The pulsatility during the formation of the clavicle can disturb the normal fusion and is now postulated by several investigators as the cause of the malunion.⁴ This would justify right-sided prevalence as the subclavian artery is normally in closer relation with the clavicle on the right, because of its higher position compared with the left side, and would explain the left-sided in the dextrocardia or situs inversus viscerum cases.^{4,6} In two different embryological studies, one found a single ossification center⁷ and the other showed that the junction of the two centers of ossification is situated between the lateral and middle third of the clavicle, not corresponding to the site of CPC (middle part).⁸

The diagnosis of CPC usually occurs during the first months of life, usually noticed after minor trauma or when the deformity increases with further growth.^{2,9} It is identified by a subcutaneous painless bump or swelling over the middle third of the clavicle. The esthetic aspect is more important when girls are affected, and become more accentuated when the patient raises the upper limb.¹¹ The skin over the bump may become thin and atrophic over the years.¹¹ ROM of the shoulder is usually normal and not painful, and some mobility can be found between two clavicular segments.^{4,5} Sometimes it can be painful or cause mild discomfort and some ROM limitations. Rarely, at the growth period, thoracic outlet syndrome may occur with neurovascular compromise.^{10,12,14}

Its correct diagnosis can be challenging. There is some differential diagnosis we have to keep in mind. We should suspect of obstetric fracture when there is a history of a difficult delivery or arm pseudoparalysis.^{5,9} The radiograph is usually sufficient to make a diagnosis, however, if there are suboptimal radiographical projections, computer tomography (CT-scan) with 3-dimensional (3D) reconstruction can be suitable demonstrating underlying anatomical details, excluding other differentials including trauma, infective and neoplastic causes, and provide essential information for operative planning.¹³

CPC and post-traumatic deformity have distinct characteristics: In clavicle fracture the borders are sharp, the fracture gap is narrow, there is a varying callus on follow-up imaging and no associated hypoplastic changes; On the other hand, CPC typically presents sclerotic borders, wide gap, there is no callus formation and has varying degrees of clavicular hypoplasia.⁴

A familiar history and multiple skeletal abnormalities suggest Cleidocranial Dysostosis, whereas, café-au-lait spots, soft tissue tumors and pseudoarthrosis affecting long lower bones suggest Neurofibromatosis type I diagnosis.⁴ Other implied syndromes include Ehlers-Danlos, Al-Awadi/Ras-Rothschild (aplasia/hypoplasia of the pelvis/lower limbs), Kabuki (short stature, musculoskeletal and cutaneous abnormalities, moderate mental retardation and facial dysmorphism) and Prader-Willi.¹¹

On histologic examination, it typically presents with hyaline cartilaginous caps on both ends of pseudoarthrosis with no signs of callus formation.⁹ The two portions are connected by a fibrous bridge that is contiguous with the periosteum, and a synovial membrane develops.^{5,6}

Because of its clinical innocence, surgical treatment remains controversial. Both surgical and nonsurgical approaches to management have been recommended.

Unacceptable appearance, painful deformity or progressive pain, functional limitations, and even parental desire may be causes of surgery, which contend improvement in upper extremity function, pain resolution and deformity correction.^{10,11,14}

Surgical complications reported, about which families should be advised, include: fixation implant failure, chronic pain and limb weakness, infections, aberrant scars on the shoulder, donor site morbidity and delayed or non-union requiring reoperation. Brachial plexus neuropraxia or vascular injury has also been reported.^{2,5,14,16}

There is no consensus about the timing of treatment or optimal method. Studer et al advice an early surgical treatment and recommend the best time of operation at the age of 5-7 years.¹⁵ However, most surgeons agree that the ideal time is between 3 years and 6 years.^{11,16} At that age, pseudoarthrosis resection is indicated, with or without associated bone grafting and/or fixation.¹¹ It is known that the older the patient, the largest size of the gap and longer time to heal.^{2,3,7,15} Type of fixation can be done using external fixation, plates and screws, screws alone, Kirschner wires or Steinmann intramedullary pins.^{11,14,15} The most commonly used technique is the excision of the non-union, bone grafting an internal fixation, however, the best type of fixation remains under debate.² Pseudoarthrosis resection only, without grafting or fixation is preferable in early childhood.¹⁶ Inclusive, Grogan et al suggested that internal fixation was not necessary for children aged <3 years.¹⁹ In his study, all children healed without internal fixation. In children with >8 years, grafts are strongly recommended, however, before that age, it can be optional.^{11,14} The most used graft is the iliac crest, but there are also other used rafts such as the tibia, ribs, and vascularized grafts.¹¹ In a review of six cases (ages ranging from 18 months to 4 years) K-wire fixation and bone grafting were preferred such as in a comparative study in children aged 4-7.5 years.^{17,21}

Nevertheless, other studies reported considerable complication rates including pin-tract infections, hardware breakage and migration, and delayed unions or nonunions.^{10,18,20} Chandran et al, concluded in a comparative study that excision of the pseudarthrosis, autogenous iliac bone grafting and stabilization with plates achieve more quickly and a higher union rate than stabilization with threaded pins.²⁰ Dzupa et al reported occurrences of clavicular fractures 20 months after removal of the synthesis semitubular plate.²²

We chose to use bone graft and internal fixation as it is commonly described, nevertheless, there are few case reports in the literature documenting CPC in late adolescence and adulthood. One case report was described in 1994 in a 45 years old patient, which was successfully treated conservatively, showing that conservative management may be appropriate if there are no functional impairment and cosmetic acceptance.²³

In our case, although he had a slight decrease in strength, the main cause for surgical treatment was the unaesthetic deformity, which was probably the cause of so late first evaluation and treatment. It is important to bear in mind that patients will exchange a protuberance for a scar, which could become hypertrophic, form keloid or remain painful.

CONCLUSION

There are few studies with extended follow-up and most of them present small sample sizes, do not allow for many statistical comparisons or subgroup analyses, so there is no optimal treatment or recommendation. Concluding, treatment must be individualized and possible complications must be discussed with the patient or family.

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