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Research Article

Congenital Pseudoarthrosis of the Clavicle: Treatment Options Using Alternative Implants

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Abstract

Congenital pseudoarthrosis of the clavicle is a rare condition. It is diagnosed at an early age by a defect in the supraclavicular fossa and the absence of a central zone portion of the clavicle in the X-ray image. Origins of the condition are not well understood nor are the best age for, and need for treatment, since it is asymptomatic in many cases. If the clinical presentation is neurovascular compression or shoulder dysfunction, reconstruction of the clavicle with a plate and bone graft from the iliac crest seems to be the most commonly accepted option.

Our case corresponds to a girl aged 9 years with an established diagnosis and a dysfunctional clinical history of the shoulder, as well as a progressively worsening esthetic defect due to the progression of the malformation. The patient was treated using a 2.7 mm mandibular reconstruction plate shaped to resemble an adult clavicle plate with an iliac crest graft. Evolution after treatment was favorable.

Currently, mandibular reconstruction plates are broadly available for treatment in orthopedic and traumatology surgery departments, mainly in pediatric surgery, since they provide the same advantages as adult reconstruction plates but with lower profiles. Their main advantage lies in the availability of support materials for three-dimensional modeling systems allowing for the plate to be adapted to the particular anatomical site, which in this case would be the clavicle.

Introduction

Congenital pseudoarthrosis of the clavicle is a rare condition with unknown etiology, and it is more common on the right side. It was initially described by Fitzwilliams in 1910 [1]. No more than 200 cases have been described in the literature, with the cases reported in Owen's work [2] being the most extensive (33 cases), followed by Cadhilac [3] with 25 cases. The condition is caused by a supraclavicular defect detected in early childhood with a deformity associated with the clavicle. The clinical appearance is variable. Some cases are asymptomatic or produce a clinical history ranging from discomfort when performing certain movements and postures to thoracic outlet syndrome.

The cause is not well-established. Two possible theories have been suggested: a defect in the fusion of the two clavicle ossification centers [4] or pressure caused by the subclavian artery leading to a lack of ossification in the clavicle [5], which would explain why the condition is more common on the right side. Histologically, the cause is a defect in the bone union to the hyaline cartilage ends which could be connected instead by a fibrous bridge [6].

Treatment is controversial. In some cases, where there is no clinical history, the deformity is not very noticeable, conservative treatment using radiological control of the deformity during the growth period is chosen. In those cases where clinical symptoms are overt, surgical treatment resecting the hyaline bone ends is required in order to connect the clavicle [3,6], and this is usually achieved with an iliac crest graft. The deformity may progress as the patient grows [3] as muscle strength increases and there is increased proximal and medial traction at the proximal end of the clavicle caused by the SCM, and increased external and inferior traction at the distal end caused by the action of the anterior deltoids. This could lead to shoulder instability and vascular complications [6,7].

Clinical Case

Nine year old girl with no medical history of interest or other congenital malformations associated. Patient presents with insidious clinical symptoms and discomfort at the right shoulder, mainly during antepulsion, in daily activities like writing, cycling, doing exercise at school or carrying weights. A very prominent proximal end of the clavicle can be palpated, as well as a gap between the two ends of the clavicle. An initial diagnosis was made two years earlier when her parents consulted

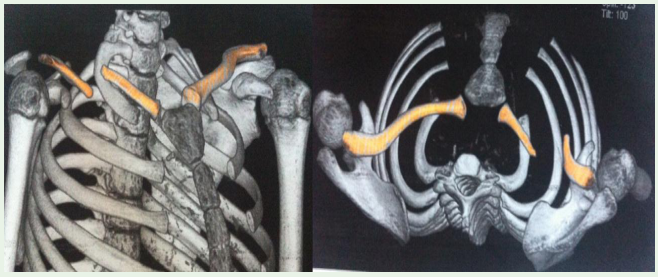

Figure 1: CT image showing the deformity of the clavicle due to muscular pulling with a defect of 2.5 cm.

the doctor concerned about asymmetry of supraclavicular gap. Additional testing with a CT scan (Figure 1) was performed because of the progressive deformity and the onset of clinical symptoms. The defect was determined to be caused by a clavicle shortening of 2.5 cm.

Surgical treatment consisting of placing a mandibular reconstruction plate (Matrix Mandible, Synthes, Oberdorf, Switzerland) with an iliac crest cortical graft, harvested from the internal table in order to minimize pain for the patient, was decided upon. The patient was situated in beach chair position and an infraclavicular incision was made in order to minimize unpleasant scars as much as possible, stable primary closure distant from the plate and upper positioning the plate in order to prevent any movement of the drill and screws towards the posterior surface, especially to the distal end, where the subclavian artery is located. During the surgery, atrophic borders of the clavicle ends covered with hyaline cartilage and joined by a fibrous tract, which was afterwards confirmed by an anatomopathological study, were removed. The medullary canal was perforated at both ends of the clavicle and the plate, made to resemble an adult plate, (Figure 2), was put in, thus eliminating the 2.5 cm defect.

Patient evolution was favorable at all times. The patient wore a sling for a month and, subsequently, active mobilization of the shoulder was initiated with no motor changes after 5 months. During that time, good consolidation and integration of the graft, as well as good evolution of the scar, were observed (Figure 3). Unfortunately, there is no further long-term information available since the patient and her family moved away and we don´t have images of complete consolidation.

Figure 2: Image of the 2.7 mm clavicle plate shaped to resemble an adult clavicle plate.

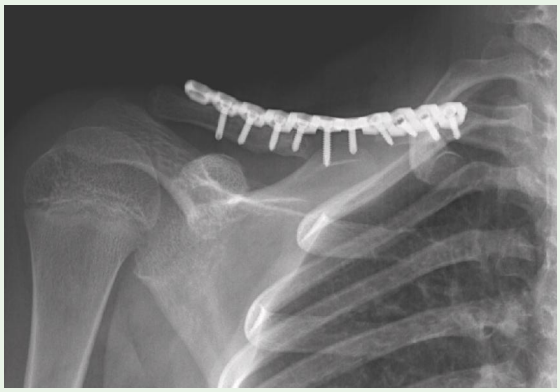

Figure 3: X-Ray image showing the results 3 months after the surgery. Adequate congruence of the plate to the original bone and the implant, where two screws were placed in order to prevent displacement and rotation, can be observed.

Discussion

Congenital pseudoarthrosis of the clavicle is a rare condition, with around 200 cases described in the literature. Diagnosis is made through clinical examination when a painless mass is observed, which increases as the patient grows, and X-ray imaging, which confirms the absence of clavicle continuity, mainly in the middle third. These findings are considered as pathognomonic by Ettl [6]. Differential diagnosis is performed to rule out cleidocranial disostosis or neurofibromatosis, and in those cases with no associated lesions, with clavicle fracture in a newborn in which a callus or a hypertrophic pseudoarthrosis is present [6].

Treatment shall be conservative while no pain or aesthetically significant deformity is present. Recommendations for surgical treatment differ depending on the author, but we based our decision on Kohler [8], who performs surgery in order to prevent possible consequences as the shoulder develops, especially if there already exists an onset of functional clinical symptoms, since complications may take place even at an adult age [7].

The age for treatment is also controversial: Schnall [9] recommends interventions in older children when the alteration is considered serious; Ettl [6] recommends surgery before school age. Cadilhac [3] suggests operating at around 2 years of age, but not later than 5 years of age. Persiani [10] performs surgeries on children between 4 and 7 years old; and Owen [2] recommends operating on children between 3 and 5 years old.

Nowadays, there are a variety of fixation systems in use for reconstructing the clavicle in infantile clavicle pseudoarthrosis. Needles have been used, but the proximity of the brachial plexus medially may cause complications [11] and not be sufficient to stabilize the wide gap between bone ends [9]. This could also lead to infections if the distal end was left outside the skin [10]. Likewise, plates may present other problems: short plates could break over time [10], systems designed for adults are too bulky in certain cases, causing protrusion of the plate because of screws that are too big causing possible osteosynthesis failures [10] as well as cutaneous discomfort. Plates with small fragments are not very resistant to

mechanical stress of the clavicle and they would only be indicated for surgeries at very early ages. Schnall [9] uses 2.7 mm Sherman-type plates for reconstruction in order to adapt the plate to the clavicle's contour, Pourtaheri [12] uses LCDC plates [Synthes, Oberdorf, Switzerland] for the same reasons. In this regard, the use of 2.8 mm mandibular reconstruction plates offer several advantages: they are long enough so that, the distance between the holes is enough so as to attach the graft with one or two screws, and the screws can be fixed to the plate making the construct more resistant. In addition, plates with narrower profiles can be used for smaller children and, above all, they can be three-dimensionally modelled in order to adapt the plates to the anatomical conformation of the clavicle just like adult plates are designed.

Unfortunately, follow-up of our patient's long-term evolution has not been possible. However, the absence of early complications or discomfort due to the osteosynthesis system 6 months after the surgery, convince us that the evolution has been completely favorable.

Conclusion

In conclusion, clavicle pseudoarthrosis with clinical symptoms and shoulder dysfunction should be treated with plate-screw system and iliac bone graft in children's of every age, even in 9 years old as in our report.

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