Pseudarthrosis Of The Clavicle: To Treat Or Not To Treat

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Citation

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Abstract

Congenital pseudarthrosis of the clavicle is a rare benign malformation of uncertain aetiopathogenesis. It is often unilateral and asymptomatic in presentation. The diagnosis is confirmed by radiology. For many authors the surgical indications are the presence of symptoms, functional impairment or cosmetic deformities.1 Treatment however, is still controversial as there are surgeons who advocate a conservative treatment of this condition2,3On the other hand, there are several surgical procedures described for treatment of this malformation. However, there is no single ideal procedure described. A case report is presented and the literature reviewed focusing the indications for treatment and the surgical techniques utilized.

A six-year-old male child was brought to the Orthopaedic outpatient department by his mother who reported noticing bump to the anterior aspect of her son's right shoulder three weeks prior to presentation. The mother stated that she only examined the area after her son complained of some trivial trauma while at school. She also reported that the bump remained unchanged and there was normal function in the right upper limb. She stated that there was no history of birth trauma and his other siblings displayed no similar bony deformities. The examination of this child revealed a playful active child in no painful distress with no dysmorphism. There was no alteration in the shape of the shoulder girdles and asymmetry was not detected in his upper trunk. There was no evidence of trauma of the overlying soft tissues and the skin was intact and normal in appearance. An obvious bony deformity of his right clavicle at the junction of the medial two-thirds and the lateral one-third was noted (figure 1). There was discontinuity in this area with the medial fragment being superior and anterior to the lateral fragment. Limited, painless mobility was present between the fragments. Normal function was present throughout the right upper limb.

Plain radiographs of the clavicle were obtained. These revealed bony separation of the clavicle at the junction of the medial two-thirds and the lateral one-third. The end of the medial fragment was noted to be tapered whereas the end of the lateral fragment was a bit enlarged and there was no evidence of callus formation (figure 2).

A diagnosed of pseudarthrosis of the right clavicle was made. The diagnosis and treatment options were discussed with the parents who decided on surgical treatment of the pseudarthrosis for cosmetic reasons. The child was admitted and pre-operative evaluation was performed. At surgery, the periosteal sleeve around the fragments was preserved (figures 3 & 4). The pseudarthrosis was excised and the fragments were stabilized with a plate (figure 5). Once the pseudarthrosis was excised, there was splintering of the medial end in attempting screw fixation so, there was little room for further fixation other than that which we were able to achieve. Bone grafting was not performed. The postoperative radiographs revealed good fixation (figures 6 & 7).

His recovery period was uneventful and he was followed up in the outpatient department with serial radiographs. Histological examination revealed fragments of clavicle covered with islands of hyaline cartilage and no evidence of callus formation. Four weeks post-operative radiographs revealed evidence of healing however there was also evidence of loosening of the screws from the lateral fragment. Biochemical markers, Erythrocyte sedimentation rate and C-reactive protein were normal. The clavicle went on to subsequent union clinically and radiographically. The implants were removed at a later date.

DISCUSSION

Congenital pseudarthrosis of the clavicle (CPC) is a rare

malformation of unresolved etiology and pathogenesis. To date, about 200 cases have been reported in the literature, the largest series being described by Gibson and Carroll.[1,4] This condition was initially described by Fitzwilliams et al in 1910 as a distinct variant of cranio-cleidodysostosis and post-traumatic pseudarthrosis.[5] Ogata et al studied the early development and ossification of the human clavicle and found that the clavicle is formed by two membranous primary ossification centers appearing by 6 weeks and fusing approximately 1 week later.[6] In a histopathological study of five patients diagnosed with congenital pseudarthrosis of the clavicle by Brouchet-Gomez et al, it was concluded that the findings confirm that the pseudarthrosis was due to the failure of coalescence of these two primary ossification centres of the clavicle.[7] Gibson et al however, supported the concept of a single centre of ossification of the clavicle. They thought that if the clavicle ossified from two centres that would be abnormal in itself, and while this may occur in cases of pseudarthrosis of the clavicle, it then required an explanation.[4] Familial occurrence of this anomaly has been documented but the pattern of genetic transmission remains obscure.[8,9]

The condition is most commonly unilateral, occurring on the right side in the majority of cases. A possible defect of fusion during intrauterine life was explained by Lloyd-Roberts et al in 1970 according to a simple anatomical consideration, namely that the subclavian artery, which runs posterior and inferior to the mid-portion of the clavicle, has a higher origin and course on the right side and could therefore influence ossification of the clavicle owing to its pulsations. This simple yet interesting pathogenic theory seems to be supported by the fact that in about 90% of cases the pathology affects the right side.[10] However bilateral and left sided cases have been documented in the literature.[11,12] The few cases affecting the left side are usually associated with dextrocardia, situs inversus or cervical ribs.[4] Again this simple theory put forward by Lloyd-Roberts et al explains the few left-sided cases in subjects with dextrocardia in which case the left subclavian artery has a higher origin and course.[10] This clavicular defect is usually not due to non-union of a birth fracture of normal bone. Owen et al stated in their experience that all neonatal clavicular fractures united readily leaving no disability.[8] The index case was in keeping with the majority of cases with the right-sided pathology.

The deformity is only rarely identified at birth or during breast-feeding in the first months of life. It generally presents as a painless subcutaneous swelling over the middle third of the clavicle, which tends to increase in size as the child grows. It may be an incidental finding or identified after some account of trivial trauma when the site is examined. However, it is usually painless and does not limit the range of motion of the upper limb. In this context the problem can be considered in terms of cosmesis. Radiological appearances are characteristic: a lack of bone continuity in the middle third of the clavicle without evidence of reactive bone. Absence of callus distinguishes the condition from traumatic causes.[13] The index case presented after trivial trauma however, there was no pain and no history of loss of function highly suggesting that the bony defect detected was due to some preexisting congenital abnormality and unlikely due to trauma. Pain is usually a common feature in post-traumatic pseudarthrosis.[2] This was reinforced on plain radiographs where no callus and smooth, rounded edges were seen.

This condition can be managed conservatively or surgically. Certain authors opt for treatment abstention. Wall et al reported on five cases where patients had no symptoms referable to the defect. He concluded that the aggressive approach to an asymptomatic lesion was not only unnecessary but symptoms in this condition were not sufficient to warrant the risks of surgical intervention.[2] Kite et al were of the opinion that a deformity without complaints was acceptable when one considered the surgical scars at the operative and bone graft sites, surgical risks and the possibility of failure of union.[14] More recently, Shalom et al considered the risks and failures of surgery, cosmetic appearance and complications and found it preferable not to perform any surgery if the problem was merely cosmetic.[3] Toledo and MacEwen, on the other hand, advocated a conservative treatment after being confronted with a motor and sensory paralysis of the brachial plexus following an osteosynthesis with a Steinmann pin, identified six hours post operatively.[15]

The indication for surgery has been cosmesis for some authors.[8,16-18] Grogan et al and Shoenecker et al performed surgery upon request from the patients or the parents, for cosmesis reasons and to relieve discomfort experienced by the patient.[18,19] Some patients however do have minor complaints, and either they or their parents wish to get rid of the lump or to obtain a stronger shoulder. Gibson et al stated that only in these circumstances should treatment be offered.[4] Hirata et al performed surgery on all children in order to avoid possible consequences on shoulder development.[20] During adulthood, pain, functional

impairment and compression syndromes of the neurovascular structures may develop.[21,22] These are seen as indications for surgical intervention. Ettl et al not only advocated surgery but also advised against conservative management since they felt that the lump over the clavicle increased with further growth resulting in instability of the shoulder girdle and increased risk of neurovascular complications. The indications for surgical intervention include the following: pain, cosmetic issues and shoulder girdle functional impairment. The parents of the index case wanted surgery despite the lack of symptoms.

The most commonly described surgical technique involves resection of the pseudarthrosis along with iliac crest bone grafting and internal fixation with a small fragment reconstruction plate.[20,23] Current literature favours surgical treatment with or without internal fixation or bone grafting. Grogan et al simply performed a resection of the pseudarthrosis without internal fixation and grafting. The authors carefully dissected the sclerotic bone ends and preserved the periosteal sleeve in order to maintain continuity and approximation of bone ends. They reported solid union in all their patients after fourteen weeks.[18] Owen et al in their series of twenty operative patients they advocated that fixation with metal of any type should be avoided as the clavicle seems to be peculiarly intolerant of metal. Removal of metal was necessary in four of their cases, resulting in delayed union in three patients and necessitating repeat grafting in one patient.[8]

Internal fixation was also achieved with K-wires and Steinmann pins in the past.[15,24] Carpenter et al used a small intramedullary pin to successfully stabilize after resection and bone grafting.[25] Although Toledo et al advocated conservative management, this was retrospectively after having used K-wires for fixation in their series.[15] These methods of fixation were rarely performed after many complications including bending, breaking, migration, infection and non-unions were reported.[15,26] Beslikas et al resected the pseudoarthrosis and stabilized the clavicular segments with an external fixator for two months until union.[27] Clinical results were excellent at the 7-year follow-up. These authors were of the opinion that surgical treatment of congenital pseudarthrosis of the clavicle in children using an external fixator provided a better cosmetic outcome with smaller postoperative scars and avoided a second surgical procedure to remove the implants.[27] Ettl et al used a contoured reconstruction plate with iliac crest bone grafting and achieved excellent fixation and solid bony fusion. The clinical results were deemed excellent with a stable shoulder girdle and no functional impairment.[23] The authors advocated surgical treatment before the school age thus allowing remodelling of the clavicle and avoidance of complications such as thoracic outlet obstruction.[23] There is paucity of information in the literature with respect to comparative outcomes of resection with bone grafting versus without bone grafting. In a series of seventeen surgical candidates, Cadhilac et al reported a higher incidence of non-unions in patients who had received resection of their pseudarthroses without bone grafting.[28] The index case was assessed after resection of the pseudarthrosis and the bone edge approximation in addition to the preservation of the periosteal sleeve were deemed adequate for bone union without grafting. The internal fixation that was utilized was inadequate as there were only two screws inserted at the medial end of the reconstruction plate. The appropriate number of cortices, which should have been used to achieve a strong construct, would have been six cortices.

The use of a locking reconstruction plate may have given a more rigid construct.

To avoid the complications of autologous iliac crest bone grafting, Elliot et al used a bovine cancellous xenograft with internal fixation.[29] Both cases resulted in treatment failures, which were associated with significant osteolysis and failure of incorporation of the graft material. This required removal of the loose metalwork and debridement of the failed graft material.[29] Although it was a level IV case series, the authors cautioned against the use of that particular xenograft in the surgical management of this condition.

The timing of surgery is still controversial. Among the authors that do in fact advocate surgery, there is still variation in the ages of these surgical candidates. Hirata encourages early intervention in order to avoid potential complications during shoulder development.[20] This may be a good concept for surgery, however all patients do not develop shoulder dysfunction and there is no way to tell which patients would develop complications. Therefore, many patients would potentially be operated on unnecessarily. Gibson and Carroll advocated delaying surgery until pre-school years (4-5 years) when the operation would be technically easier and to avoid complications such as iatrogenic splintering of bone.[4] The index patient was operated on at age six and there was still some amount of splintering of the bone upon internal fixation with a reconstruction plate, a complication that should be less likely

in older children. In a series of seventeen patients, Cadilhac et al performed surgery at a mean age of six years and four months.[28] Owing to the fact that many of these patients are asymptomatic, their presentations are often times incidental. Therefore, the age at which surgery is performed is usually not influenced by choice but by the age of presentation.

CONCLUSION

Congenital pseudarthrosis of the clavicle is a rare entity and as such the populations studied with this condition are quite small. The aetiopathogenesis is still not well understood and so the prevention or even the management is still controversial. Conservative management has been advocated, but most authors now recommend surgical intervention for of pain, cosmesis and functional impairment. In addition, the timing of intervention and the postoperative care have not been agreed upon. In the literature review, the most common surgical procedure involves resection of the pseudarthrosis, iliac bone grafting and internal fixation. Although there was no consensus, a popular suggestion was early treatment in all cases, even though in the initial stage CPC appears to be a 'simple' cosmetic defect. The onset of symptoms and functional impairment are believed to develop as the child grows with increased clavicular swelling over time. The index case underwent one of the more popular surgeries for cosmetic reasons. Although he presented at six years there were still a challenge with fixation, a real concern of authors who advocate delayed surgery. Fortunately, there was union with good functional outcome and no further complications.

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