

BILATERAL CONGENITAL PSEUDARTHROSIS OF CLAVICLE: A CASE REPORT

GIRISH GOPINATH¹, REJU VENGAKANDIYIL² AND MOHAMMED KUNJU RAFFIC³

¹Assistant Professor, Department of Orthopaedics, Sree Gokulam Medical College and Research Foundation

²Junior Resident, Department of Orthopaedics, Sree Gokulam Medical College and Research Foundation

³Professor and HOD, Department of Orthopaedics, Sree Gokulam Medical College and Research Foundation

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CORRESPONDENCE

drgg000@gmail.com

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ABSTRACT

Congenital pseudarthrosis of the clavicle is a rare condition. Bilateral presentation is even rarer, with less than ten reports in the literature. There are no definite treatment recommendations for want of sufficient long term follow up of untreated cases. We report a 27 year old man with bilateral congenital pseudarthrosis who has no pain or functional impairment and is actively involved in all day to day activities. The radiological appearance of the case is different from previous reports in that there is a hypertrophic segmental nonunion at the pseudarthrosis bilaterally. We propose that surgical intervention as a prophylaxis for future thoracic outlet compromise is not indicated.

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Congenital Pseudarthrosis of the clavicle is a rare disorder with only about 200 cases reported in the literature. Fitzwilliams(1910) described in detail three patients with pseudarthrosis of the clavicle, two of whom had other associations of cleidocranial dysostosis. The third patient in his description, with an isolated clavicular pseudarthrosis, is generally credited as the first report.^{1,2} Most cases are unilateral, and invariably right sided.^{3,4} Left sided involvement is commonly associated with dextrocardia,⁴⁻⁶ but there are reports of cases where the presentation was ipsilateral to the heart.⁷ The exact aetiology is unknown,⁸ but the pathogenesis is related to the embryologic development of the clavicle.^{5,9} The clavicle ossifies in membrane and is the first ossific mass in the developing embryo. It first appears as a mesenchymal bar at four weeks, followed by a precartilaginous mass at fifth week, and an ossific mass by seventh week.^{5,9,10} Pressure from the subclavian artery, which is normally slightly higher on the right

side, can hinder fusion of the two developing ends at the centre, and this is the proposed reason for increased incidence on the right side.¹¹ Occasionally, both subclavian arteries may be highly placed predisposing to a bilateral pseudarthrosis, as seen in association with cervical ribs or high, vertically oriented upper ribs.⁸ Some series report cases running in families, leading to a suggestion of possible autosomal recessive⁷/dominant⁸ inheritance. Congenital pseudarthrosis of the clavicle presents at birth or in the neonatal period with a painless prominence of the clavicle, slightly lateral to the midpoint.^{2,8,12,13} There is some abnormal mobility at the site. The deformity becomes more prominent with age¹³ and is more pronounced when the patient lifts the arm. The affected shoulder may be lower.¹⁴ Usually asymptomatic, some patients may have mild to moderate pain around the shoulder, with weakness of arm and limitation of abduction. There are no definite treatment recommendations for this condition.



Some authors suggest that although congenital pseudarthrosis of the clavicle is asymptomatic in childhood, surgical treatment can restore normal morphology and prevent functional or vascular problems in adolescence and adulthood. Union requires open reduction and bone grafting, the ideal time for which is between 3 to 5 years. The older the patient, more difficult is the grafting. There are however reports of successful union with resection of fibrous pseudarthroses and sclerotic bone ends, careful dissection and preservation of the periosteal sleeve to maintain continuity, and approximation of bone ends, without bone grafting or internal fixation. These were in patients less than 5 years of age.

We report a case of bilateral congenital pseudarthrosis of the clavicle in a 27 year old male who presented for other reasons to the orthopaedic department. Bilateral involvement is extremely rare, there being less than ten reports¹⁵⁻¹⁷ in the international literature, to the best of our knowledge. Most of these reports are in patients in their childhood, and for want of sufficient reports on the natural history of this condition, there is confusion on what treatment approach to be adopted. The patient was informed that information regarding his condition would be submitted for publication, and he provided consent.

CASE REPORT

A 27 year old male patient presented to the emergency with a history of a trivial assault to upper back and pain around the shoulders. No significant assault related injury was detected. His examination revealed bilateral prominences of his 'collar bone' which he claimed to have been present since his earliest memories of himself. There was increased mobility of the two ends. Shoulder movements were full and painless, and there was no weakness of the muscles around the shoulder girdle. There were no signs of thoracic outlet compromise. His mother said that the lump had been present since birth. There was no history of a difficult delivery or any birth injury. Clinical findings and plain radiographs of the right shoulder and clavicle confirmed the diagnosis of congenital pseudarthrosis of the clavicle. Radiographs showed a hypertrophic segmental non-union at the midpoint of the clavicle. To the best of our literature search, we could not find a previous description of a segmental congenital non-union of the clavicle. There was no similar abnormality in the patient's parents or siblings. There were no skin stigmata of neurofibromatosis and radiological evaluation of skull and spine were normal. The patient had always been comfortable and had not contemplated any treatment as he was neither concerned functionally nor cosmetically.



FIGURE 1



FIGURE 2



FIGURE 3



FIGURE 4

DISCUSSION

The differential diagnoses to be considered in pseudarthrosis clavicle are obstetric fracture, post-traumatic non-union, cleidocranial dysostosis, and neurofibromatosis.^{18,19} A history of a difficult delivery with neonatal pseudoparalysis and pain, and massive callus progressing to union later are suggestive of obstetric fracture. In a traumatic non-union, the bone ends seem attenuated.⁸ In Cleidocranial dysostosis, the clavicular defect is often bilateral with associated skull deformities (wide fontanelles and sutures, large skull, small facial bones); crowded teeth; pelvic ring deficiencies; scoliosis; and abnormal epiphyses of hands and feet.⁵ Neurofibromatosis is characterised by cafe-au-lait spots on the skin.⁴ When the diagnosis is in doubt, CT scan may be helpful to confirm the diagnosis.²⁰ Some syndromes associated with congenital pseudarthrosis of the clavicle are Ehlers-Danlos, Al-Awadi/Ras-Rothschild, Kabuki and Prader-Willi.⁷

When patients are asymptomatic and have no functional disability, some authors do not recommend any treatment.^{18,21,22} Surgical treatment should be considered for symptomatic patients with dysfunction of the arm;²¹ when patients are embarrassed by the appearance of the lump;^{9,23} and to prevent thoracic outlet syndrome in the future.^{24,25} The optimal age for surgery is controversial. Some recommend that surgery be done between 3 and 4 years of age. Others recommend that the primary excision of the pseudarthrosis be performed during infancy. Surgical treatment consists of excision of the pseudarthrosis and internal fixation of the bone ends, with or without bone graft, using intramedullary devices,²⁶ or plates and screws.¹⁹ External Fixation is another possible method of treatment.¹⁰ There is also a report of the use of Ilizarov fixator for treating a congenital pseudarthrosis.²⁷ The reported complications of surgery include

hypertrophic scar, non-union, infection and graft site morbidity.

Kite has proposed a classification system for congenital pseudarthrosis. Type I which is seen at birth with a space between the fragments, with a hypoplastic distal fragment, and a larger medial fragment, where surgery is not indicated; and Type II where the clavicle may be normal at birth, fractures with the slightest trauma and goes for non-union, much alike congenital pseudarthrosis of the tibia, where surgical treatment may be indicated.²⁸

The inadequate recording of natural history of this condition is the reason why surgery is often attempted, mainly for a fear of any future neurovascular/functional deficit.²

This patient is reported for mainly three reasons.

1. Because of the rarity of this condition presenting bilaterally.
2. There are not many reports of pseudarthrosis clavicle followed up to adulthood without any active surgical intervention, as a guide to suggest that the natural history is compatible with normal function. This patient is a young man who has had no functional deficits despite the lumps on his clavicles, and has no evidence of any thoracic outlet compromise. He has full range of movements in all directions and has good power. Even though there is a cosmetic problem, he does not want to consider any surgical treatment, suggesting that he is comfortable. It is therefore likely that if congenital pseudarthrosis is allowed to remain, the patient can expect to have normal function, and surgery as a prophylaxis for future thoracic outlet compromise is not necessary. There is a previous report of an ENT surgeon with unilateral untreated pseudarthrosis clavicle actively involved in sports and also performing fine surgeries at the age of 45.²⁹
3. The typical radiologic appearance is of a non-union at the centre, slightly lateral to the midpoint. In this case, the location of the non-union is typical, but the appearance is different in that it is hypertrophic, and also segmental in configuration. This type of a segmental non-union is also not previously reported.

SUMMARY

Bilateral congenital pseudarthrosis is very rare, and except for cosmetic problem, it is compatible with normal function and neurovascular status. The radiologic appearance of a segmental configuration at the pseudarthrosis is not previously reported to the best of our knowledge.

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