Case Report



Congenital Pseudoarthrosis of Clavicle: A Case Report and Review of Literature

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Abstract

Congenital pseudarthrosis of the clavicle (CPC) is considered one of the rare conditions that regularly diagnosed during the first years if life and resulted from the failure of the union process of the ossification nuclei of the clavicle. Surgical management is considered the first line treatment of this condition however, associated with some complications. In this case study, we reported a case of 2- year- old- boy presented with Congenital pseudarthrosis of the clavicle that was treated with conservative management.

Keywords: Congenital pseudarthrosis, CPC, Pseudoarthrosis of Clavicle, case report, conservative management

Background

Congenital pseudarthrosis of the clavicle (CPC) is considered one of the rare conditions that regularly diagnosed during the first years if life and resulted from the failure of the union process of the ossification nuclei of the clavicle [1-3]. This condition was first reported and accurately described by Fitzwilliams in 1910 and reported as a case series of cleido-cranial dysostosis as a monostotic variant ^[4]. In general., CPC is more common among females than males and mostly are unilateral [5,6]. Moreover, CPC is more common in the right clavicle over the contralateral and when occurred at the left side, some authors reported a potential association with dextrocardia [3]. The etiology of CPC remains unknown however, the pathogenesis may be related with the embryology of the clavicle [7-9]. According to the literature published there is one well documented family with several members affected by CPC [8] however, a clear genetic pattern has not yet been associated with this condition ^[2,3,10]. However, CPC is present at birth, most of the cases has been reported between the first months and 5 years of life of the child ^[6]. Mostly, patients are presented with painless swelling over the middle third of the clavicle which tends to increase with growth. Diagnosis of CPC includes obstetric fracture which tends to rapidly heal with an exuberant callus, neurofibromatosis, cleido-cranial dysostosis and post- traumatic nonunion^[11]. In general, most of patients may remain asymptomatic during the entire life and the shoulder's range of motion remain normal with any pain [9,12]. Generally, the asymptomatic pseudarthrosis of the clavicle can be responsible for different

aesthetic issues as well as functional symptoms that indicating the need for surgical repair ^[13,14]. However, surgery is still debated and includes excision of the non-union with or without bone grafting and stabilization ^[14,15]. In this case, we reported a case of 2- year- old-boy presented with Congenital pseudarthrosis of the clavicle that was treated with conservative management.

Case report

A 2-year-old Saudi boy presented after his parents noticed a protruding right skin tinting o the right clavicle (**Figure 1**). The patient was asymptomatic of the swelling with no pain. There was no history of trauma and no definitive timeline as to when the lump was first noticed. The family history showed no instance of congenital or acquired musculoskeletal disorders or any other comorbidity. No prenatal history or having complications during birth. On physical examination a pointed 2×2 -cm non-tender mass with a normal overlying skin was present in the mid-third of the right clavicle with full range of motion. The mass appeared to flatten with abduction of the right shoulder. No other abnormities were noted.

Radiographs of the chest (**Figure 2**) showed a pseudarthrosis in the middle of the right clavicle. Both segments pointed slightly upward and forward and each ended in a bulbous mass. The two masses abutted each other with the sternal end above the acromial end. Conservative and operative management options were discussed with the parents. Conservative treatment was started and continued until having full range of motion and no pain.



Figure 1: A 2- years old boy presented with protruding right skin tinting



Figure 2: The radiological of the chest showing the pseudarthrosis in the middle of the right clavicle

Discussion

CPC is a rare condition which reported in over 200 cases in the literature review ^[16]. CPC is largely documented as being a girlsided condition with right sided cases representing 95 % of cases and left sided cases were associated with dextrocardia or situs inversus ^[16]. In this case, we reported 2- years old boy with symptoms of CPC. Most of the authors believe that CPC is conditions that caused by extrinsic pressure that exerted on the budding clavicle by the adjacent subclavian artery ^[17]. Radiological characteristics of CPC include clear separation in the middle portion with the medial fragment that may be positioned above the lateral fragment ^[17]. CPC is different from the obstetric fracture and post-traumatic nonunion ^[18] as the obstetric fracture of the clavicle should be suspected when there is a history of difficult delivery, pseudoparalysis of the arm with no voluntary limb movement and pain on passive movement ^[9]. In this case, no pain was reported by the patients with no history of difficult delivery, therefore, obstetric fracture of the clavicle was excluded from the diagnosis.

The indications for surgery are mainly taken due to the aesthetic impairment as well as due to progressive pain, functional limitation, late-starting thoracic outlet syndrome or because of combination of these factors ^[9,13]. The timing of the surgery is different among studies. Many authors recommended that surgery should be delayed until the patient is between 3-6 years of age where at this age, resection of the focus of the pseudoarthrosis is indicated with or without associated bone grafting and fixation ^[19-21]. Moreover, some authors suggested that conservative treatment could be adequate when patients do not have disturbed function of the affected side and complications of the operative surgery are

outweighing the potential benefits ^[10,22]. They argued that CPC is a benign condition that should be treated as such. In our case, no pain or functional impairments were reported by parents or upon physical examinations and after discussing the options of the treatment, parents with physicians chosen the conservative management with positive outcomes. Another reported study showed a successful conservative management of a 45-year-old man who was left with a mild cosmetic swelling in right midclavicular region and no functional impairment ^[23]. This shows that conservative management may be appropriate in some patients if there are no concerning features, and the cosmetic burden can be accepted.

Conclusion

CPC is a rare condition that is reported in few cases mostly among female children thus, our case is considered rarer and should be reported. Our case reported the successful application of conservative management of CPC without using any surgical intervention.

Ethics approval and consent to participate

Not applicable

Conflicts of Interest

None

Funding Statement

None

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