Congenital Clavicular Pseudoarthrosis

Global Pediatric Health
Volume 7: 1–3
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DOI: 10.1177/2333794X20974211
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Received May 27, 2020. Revised October 20, 2020. Accepted for publication October 26, 2020.

Highlights

What do we already know about this topic?

Congenital clavicular pseudoarthrosis is a rare entity with only 200 cases reported worldwide.

How does your research contribute to the field?

My research describes clinical and radiological features of congenital clavicular pseudoarthrosis which allow its disctinction from the more frequent fractures.

What are your research's implications toward theory, practice, or policy?

Congenital clavicular pseudoarthrosis being a pathology rarely reported in pediatric and radiological journals; we aim to familiarize practitioners with this entity's diagnostic criteria.

Introduction

Congenital pseudoarthrosis of the clavicle is an uncommon condition, with 200 cases reported worldwide so far.[1] It is defined as a bone defect of the middle third of the clavicle, more prevalent in girls and on the right side.[1] First noted at birth or in early childhood; it presents as a painless mid-clavicular bulge.[1] Radiographs demonstrate a mid-clavicular large bone gap with sclerotic ends and no callus.[2] We report the case of a 6 year old girl with a right mid-clavicular painless swelling whose clavicular radiograph and computed tomography revealed 2 overlapping clavicular segments with bulbous borders and no bridging in favor of a congenital pseudoarthrosis of the clavicle. Our aim is to highlight congenital pseudoarthrosis of the clavicle's specific clinical and radiological features that allow prompt diagnosis and distinction from the more common clavicular fractures.

Case Report

A 6 year-old girl with negative past medical and surgical history was referred to our radiology department for investigation of a right mid-clavicular painless swelling (Figure 1), first noted by parents in the neonatal period, but never explored. The deformity became more prominent as she grew. Pregnancy was uneventful. She was born to a non consanguineous couple, at full term, via spontaneous atraumatic vaginal delivery and had completely normal development. There were no similar familial cases. On examination, the swelling was non tender. Her right shoulder's ranges of motion and right upper extremity vascular and neurological examination were normal. No other abnormalities-musculoskeletal in particular-were noted. A radiograph revealed a defect in the right mid-clavicular portion (Figure 2). Computed tomography with 3 dimensional reconstructions showed the bone gap with bulbous, well corticated borders and no callous formation (Figure 3). No other abnormalities were noted. Clinical and radiological features were in favor of a right clavicular congenital pseudoarthrosis.

Discussion

First described by Fitzwilliams in 1910,[3] congenital pseudoarthrosis of the clavicle is an uncommon malformation with 200 cases reported worldwide.[1]

Etiopathogenesis is unclear. It is suggested to be due to failure of the clavicle's 2 ossification centers to coalesce around the seventh week of gestation.[1] It is more prevalent in girls and involves the right side in 80% of cases.[1] Some investigators stipulate that the subclavian artery pulsations cause the ossification centers failure to fuse. The right subclavian artery's higher position brings it closer to the

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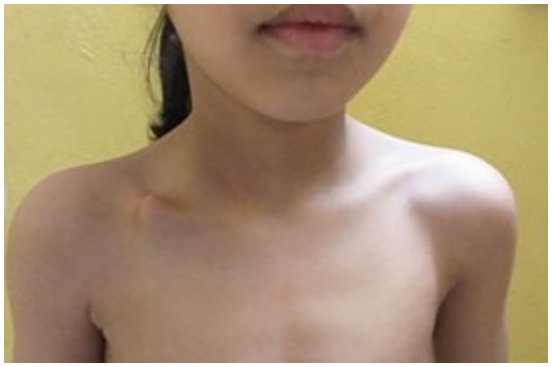

Figure 1. Clinical appearance of the patient. Note the right clavicular swelling with normal overlying skin.

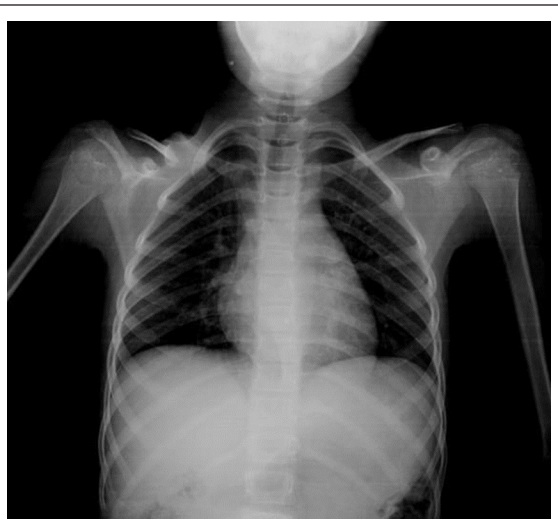

Figure 2. A plain frontal chest radiograph demonstrating the right mid-clavicular bone defect with no callous formation. The 2 clavicular segments overlap and the medial fragment is in a cephalad position.

clavicle in comparison to the left side, which may explain CPC's right sided predominance. Left CPC is rare and may associate with dextrocardia, situs inversus and cervical ribs.[4] Bilaterality is noted in 10% of cases. Familial cases are seldom encountered and suggest a plausible inheritance pattern.[1] Another unconfirmed etiological hypothesis is the fetus' abnormal intrauterine positions.[1,5]

Diagnosis is clinico-radiological. Clinically; a painless mid-clavicular swelling is noted at birth or in early childhood, and becomes more prominent as the child grows.[1] Usually, the affected shoulder's mobility range is normal.[4] Reviewing birth history carefully to rule out trauma is important notably in macrosomia, dystocia and forceps delivery settings.[5]

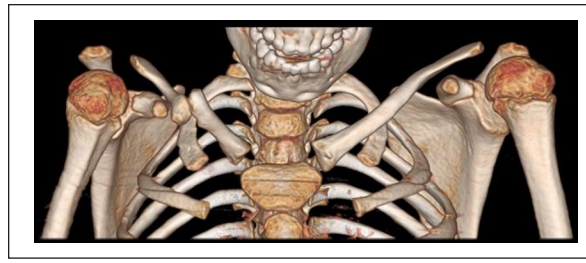

Figure 3. A 3 dimensional computed tomography reconstruction showing the right mid-clavicular bone gap with well corticated ends and no bridging. The medial clavicular half points upwards in regards to the lateral larger half.

Radiographic features are characteristic and allow differentiation from clavicular fractures. CPC appears as a mid-clavicular defect, wider than the one seen in fractures.[2] CPC bone borders may be rounded, elephant-foot shaped or tapered.[4] They are well corticated, whereas fracture's bone ends are sharp and non sclerotic. Classically, the clavicle's medial-sternal- half is atrophic and located anteriorly and superiorly in regards to the lateral-acromial-, larger half.[5] Unlike CPC; fractures do not associate with clavicular hypoplasia and demonstrate bone bridging and callous formation a few weeks into follow-up.[5] Computed tomography 3 dimensional reconstructions delineate morphological details and discern a callous when present.[2] They are of use in cases of suboptimal radiographic projections, for surgical planification and differential diagnosis including trauma, infection, neoplasms . . .[2]

Other than trauma, CPC's differential diagnosis include cleidocranial dysostosis; an uncommon autosomal dominant bone developmental abnormality resulting in both clavicles hypoplasia or absence, fontanels and sutures persistence and dental anomalies.[4] CPC association's with neurofibromatosis, Ehlers-Danlos, Prader-wili, Kakubi, Al-Awadi/Raas-Rothschild syndromes, brachial plexus injuries, elevated and verticalized upper ribs, cervical ribs, clavicular bifurcation as well as vertebral anomalies have been reported.[1]

Upon evolution, the clavicular deformity becomes more apparent and the skin underlying it may atrophy; hence the aesthetic issue.[4] To the best of our knowledge, spontaneous consolidation was only reported once, in an infant with bilateral involvement.[6] Progressive pain and thoracic outlet syndrome have been reported.[4]

Treatment is controversial. Surgery is indicated for aesthetic and/or functional (progressive pain, thoracic outlet syndrome) repercussions. Early treatment avoids complications, and surgery's best time is between 4 and 7 years.[7] Operative options consist of pseudoarthrosis curettage and fixation (external or internal), with or without bone grafting.[7,8]

Author Contributions

HR: contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy. SEH: contributed to conception; contributed to acquisition and interpretation; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy. NA: contributed to conception; contributed to analysis and interpretation; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy. LC: contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical Approval

Ethical approval is not required for de-identified single case reports based on institutional policies.

Informed Consent

Informed consent was obtained for publication.

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