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Case Report

Congenital pseudarthrosis of the clavicle: a case report and review of the literature

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Abstract: Congenital pseudarthrosis of the clavicle (CPC) is a rare deformity. We describe a case of CPC presenting in a two-year seven-month old girl with a nontender and immobile mass in the middle of the right clavicle. This mass was found by her parents at birth. Treatments given were surgical resection of the pseudarthrosis, autologous iliac bone grafting and internal fixation with Kirschner wire. The functional abilities of the affected shoulder remained normal and without any complications at two years post-operation. The diagnostic characteristics of radiography and treatment of CPC in children will be investigated in the present study.

Keywords: Pseudarthrosis, clavicle, congenital

Introduction

Congenital pseudarthrosis of the clavicle (CPC) is a rare deformity. The first case was described by Fitzwilliams in 1910 [1-17]. CPC is predisposed to the middle third portion of the right clavicle [4, 18-20] with 10% of CPC cases presenting as bilateral [14, 21], and only rarely involving the left clavicle [7, 12, 22]. Left-sided unilateral involvement is often associated with dextrocardia [9, 17, 22-24], or cervical ribs [1-6, 10, 11, 14, 21, 25]. A painless and growing mass can be found in the middle of the clavicle at birth, while the function of the affected shoulder remains normal [18].

Case report

The presenting individual with CPC was a two-year and seven-month old female child. She was the mother's first child. The prenatal ultrasonography examination was normal. There was no history of birth injury and anoxia. A nontender and immobile mass in the middle of the right clavicle was found by her parents at birth. The function of her affected shoulder and upper limb was normal. The plain radiograph at 4 days (**Figure 1A**) and at 38 days of age (**Figure 1B**) showed a break in the middle third of the affected clavicle. The rounded, enlarged and sclerotic fragments were shown on the radiograph. This patient was presented to our institution when she was two-years and seven-months old. The physical examination showed a 2 cm × 2 cm mass in the middle of the right clavicle. The mass was hard, immobile and nontender, and the function of the affected shoulder was normal. The plain radiograph showed a break in the middle third of the affected clavicle (**Figure 1C**). Computer Tomography (CT) scan also showed a break in the middle third of the affected clavicle. There was soft dense tissue between each end (**Figure 2**). Magnetic Resonance Imaging (MRI) showed that there were mixed signals between the two fragments of the broken clavicle. The medial part of the clavicle was smaller than the lateral part (**Figure 3**).

During surgery, it was found that there was some fibrous tissue between the pseudarthrosis without bony connection. An en-bloc of the pseudarthrosis was performed with a residual bony defect of about 1.0 cm, in which an autologous ilium graft was performed (**Figure 4**). The two portions of the clavicle and the autologous ilium were fixated with a 1.5 mm diameter Kirschner wire. Autologous iliac cancellous bone was filled at the pseudarthrosis. An intraoperative image intensifier was used (**Figure 5**).

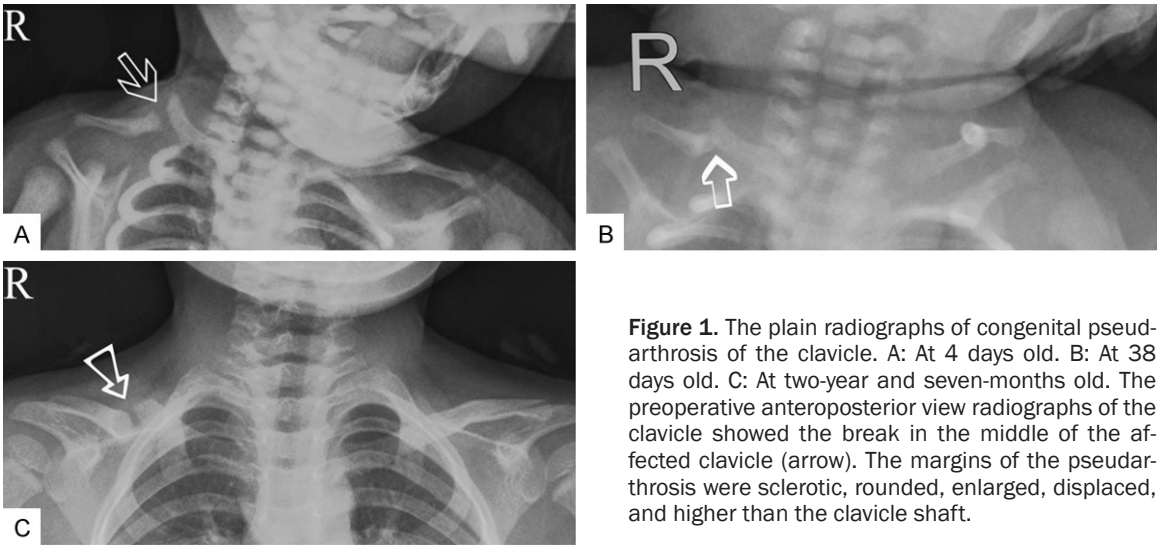

Figure 1. The plain radiographs of congenital pseudarthrosis of the clavicle. A: At 4 days old. B: At 38 days old. C: At two-year and seven-months old. The preoperative anteroposterior view radiographs of the clavicle showed the break in the middle of the affected clavicle (arrow). The margins of the pseudarthrosis were sclerotic, rounded, enlarged, displaced, and higher than the clavicle shaft.

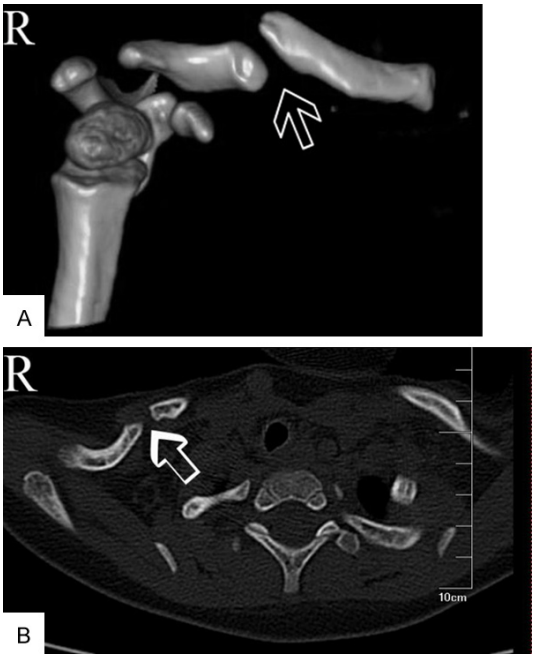

Figure 2. Preoperative appearance of the three dimensional Computed Tomography scan (CT) of the clavicle. A: There was a discontinuous and displaced clavicle (arrow). B: Horizontal scans showed a discontinuous and displaced clavicle with soft tissue interposition (arrow).

The plain radiograph showed bony healing at two years post surgery (**Figure 6**). The function of the affected shoulder remained normal and without any complications.

Discussion

The etiology of CPC remains unclear [3-5, 7, 9-12, 14, 16-19, 21-29]. It is believed that cer-

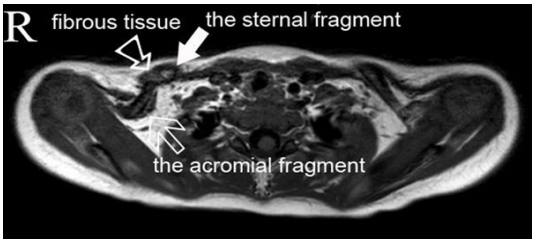

Figure 3. The preoperative Magnetic Resonance Imaging (MRI) horizontal scans of T1 weighted of the clavicle showed a discontinuous clavicle with fibrous tissue interposition (upper hollow arrow). The discontinuous clavicle including the sternal end (solid arrow) and the acromial end of congenital pseudarthrosis of the clavicle (lower hollow arrow).

vical ribs or vertically orientated upper ribs caused the compression of the subclavian artery between the clavicle and the first rib. The excessive arterial pulsations and pressure can result in the development of a pseudarthrosis. The development of the clavicle relies on medial and lateral separate ossification centers [2, 5, 7, 9, 10, 12, 14-18, 24, 25, 29]. At the seventh week of embryonic development, the first ossification center of the clavicle is formed. The non-fusion of two ossification centers of the clavicle may be another reason for CPC [1-12, 14-16, 19, 21, 22, 24-26, 28, 29]. However, based on the study by Ogata and Uhthoff [30], the junction of the two primary ossification centers was located between the lateral and middle third of the clavicle, which is not the typical location for CPC. Gibson [31] thought there was only one ossification center in the early stage of clavicle ossification. Environmental factors also have an important effect on the formation of

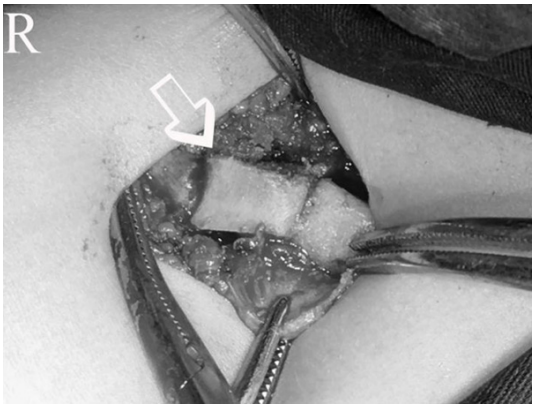

Figure 4. An autologous ilium grafting was performed and packed in the bony defect of pseudarthrosis in the surgery (arrow). The two residual parts of the clavicle and the autologous ilium were fixated with a Kirschner wire.

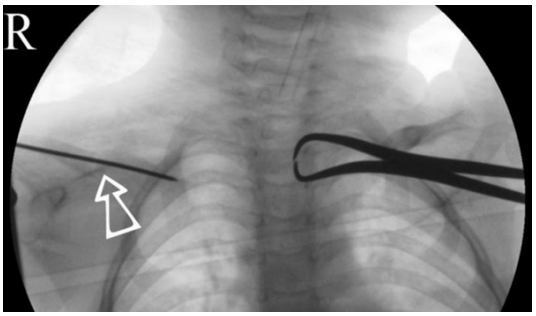

Figure 5. The intraoperative image intensifier showed the autologous ilium graft and two parts of the clavicle fixated by a Kirschner wire (arrow).

CPC [3]. The formation of CPC may be related to the RUNX2 (CBFAI) gene mutation [6].

The English literature on congenital pseudarthrosis of the clavicle from 1964 to 2017 is shown in **Table 1**.

The clinical manifestations of CPC are a hard, painless, and an immobile bony mass [5, 27]. The mass often grows as the patient ages. The affected scapula may be shortened and pendulous. The patient may feel pain in the pseudarthrosis location because of the activity and pressure, but the function of the affected shoulder is always normal [6, 9]. The skin around the pseudarthrosis becomes atrophic and thin. The swelling in the pseudarthrosis will be aggravated when the affected arm is raised [2, 5, 9]. The plain radiograph shows a break in the middle third of the affected clavicle, which is divided into two fragments. CT scan

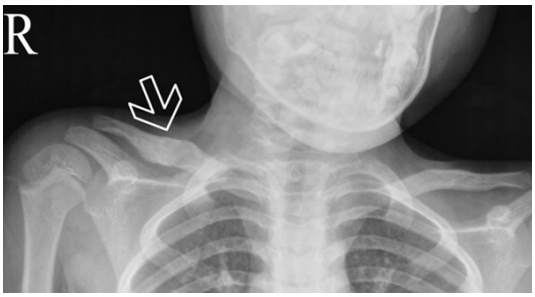

Figure 6. The plain radiograph of the clavicle at 24 months postoperative, showing bony union of the pseudarthrosis (arrow).

shows that there is a soft tissue density mass between both ends. In the histologic examination, there are hyaline cartilage caps in the proximal and distal pseudarthrosis. The cartilage caps are connected by fibrocartilage. A single or pair of chondrocytes may be found in the rich collagen fibers [11].

The differential diagnosis of CPC include Cleidocranial Dysplasia (CCD), trauma of the birth canal and neurofibromatosis [3, 7, 10-13, 15, 18, 22, 28, 29]. The formation of CPC is closely related to the cervical rib [18]. A portion of the clavicle or the entire clavicle is missing in the patients of CCD. There is no pseudarthrosis with retarded cranial ossification, supernumerary teeth, short stature [1, 5, 16, 22], scoliosis and pelvic defects [2]. The history of trauma and birth trauma of the patient should be ruled out when CPC is diagnosed, which will cause local tenderness of clavicle. The plain radiogragh of the clavicle would show the formation of a callus.

The function of the affected shoulder is normal in most patients with CPC, but the treatments for CPC are controversial. Conservative treatment should be considered when there are no cosmetic concerns, or when there are no clinical symptoms of the affected shoulder and upper extremity [2, 12, 18, 27, 31]. Surgery of CPC [2, 7-9, 12, 18, 22] can completely restore the physiological function of the shoulder joint [8], and prevent thoracic outlet syndrome and improve the cosmetic appearance.

The most common complication of CPC is the thoracic outlet syndrome. The cause of thoracic outlet syndrome is that the subclavian artery, the subclavian vein, and the brachial plexus are compressed at the thoracic outlet. Thoracic

Table 1. Literature of congenital pseudarthrosis of the clavicle from 1964 to 2017

Year	Author	No	Side	Surgery age	Radiograph	Treatment	FU (No)
2013	Watson [3]	1	Right	19 y	Right clavicular pseudarthrosis	RAP	Good
2011	Elliot [4]	2	Right	5 y	Congenital pseudarthrosis of the clavicle	RP and cancellous bovine bone grafting	Good (2)
2009	Currarino [5]	4	Right	4 y, 7 y, 8 y, 14 y	A pseudarthrosis in the middle of the right clavicle. Both globular segments were upward and forward	RP (1) RA (2) NT (1)	Good (3) No FU (1)
2017	Studer [7]	7	Right (6) B (1)	7.1 y (5-8)	Congenital pseudarthrosis of the clavicle	RAP	Good
2008	Persiani [10]	17	Right	6.08 y (4.3-7.6)	The bone ends were hypertrophic or thinned; the sternal fragment was always longer than the acromial fragment	RAP (5) RP (3) RAK (7)	Good (11) Fair (3) Poor (3)
2004	Gomez [11]	5	Right	6.5 y (3-15)	A discontinuity of the clavicle; two voluminous bony ends	RAK	Good (4) Recurrence (2)
2005	Ettl [12]	3	L (1) Right (2)	4 y, 6 y, 8 y	Enlarged clavicular ends at the pseudarthrosis site and no callus formation	RAP	Good (3)
1999	Sakkers [14]	1	L	13 y	The residual bony ends were smooth and segment was tapered and located superior to the lateral, bulbous segment	NT	No FU
1980	Quinlan [16]	4	Right (3) L (1)	1.5-6 y	Pseudarthrosis of the clavicle without evidence of reactive bone and callus	NT (1) RA (1) Rshin (1)	Good (4)
1979	Manashil [17]	3	Right	Newborn (2), 2 y (1)	Congenital pseudarthrosis of the clavicle without evidence of reactive bone and callus	NT	No FU
2009	Glotzbecker [18]	1	Right	4 y	Congenital pseudarthrosis of the clavicle	RS	Good (4)
2009	Brévaut [19]	1	Right	Newborn	The borders of the two fragments were sclerotic and no callus formation	NT	No callus formation
1991	Grogan [21]	8	Right	8 m-6 y	Congenital pseudarthrosis of the clavicle. The proximal of ends ossified	R	Good (8)
2008	O'Leary [22]	3	Right (2) B (1)	1 m, 18 y, 22 y	Congenital pseudarthrosis of the clavicle. The ends of clavicle were smooth	NT	No FU
2005	Sforza [23]	1	Right	33 y	Rounded enlargement with sclerotic and smooth margins, with no signs of reactive bone or callus formation in the pseudarthrosis site	RP and bone grafting	Good
1990	Russo [24]	1	B	Newborn	The sternal fragment was the larger, with no overlap	NT	No FU
1993	Morin [25]	1	Right	Newborn	The sternal segment was smooth, blunt and above the acromial segment	NT	No impairment
2016	Nieto Gil [26]	1	B	3 m	Bilateral pseudarthrosis of the clavicles	NT	No FU
2003	Karakurt [27]	1	B	22 y	Wide separation of bone fragments, no callus formation between the bone ends, no arthritic changes in the acromioclavicular joints	NT	No functional disability
2001	Lorente Molto [28]	6	B (1) Right (5)	2 m-10 y	The wide separation of bone fragments. The bone ends are bulbous	NT (1) RAK (5)	Good
1995	Hirata [29]	1	Right	5 y	A lack of bone continuity in the middle third of the clavicle. The bone ends are globulous	RAP	No FU
2012	Galanopoulos [2]	1	Right	9 y	Clear separation but no displacement of the clavicular diaphysis	RAS	Good
1988	Schnall [15]	6	Right	4-15.5 y	No callus formation in the pseudarthrosis. The bone ends are bulbous	RAP	Good (6)
2013	Sung [1]	1	Right	Newborn	The residual bony ends were smooth	Unstated	Good

2017	Di Gennaro [6]	27	Right (26) L (1)	7.0±3.2 y (0-47)	Gap distances were 9.5±5.6 mm (2-28) in the pseudarthrosis. The clavicular segments were horizontal or upward, with an elephant-foot appearance	R (19); A (15) Allofibula grafting (4) P (1); K (18)	Good (25) Internal rotation impairment (1)
2011	Chandran [8]	10	B (1) Unknown (9)	Unknown	Unknown	Group A: RAK. Group B: RAP (5)	Union (3) and nonunion (2) in group A Union (5) in group B
1999	Sales de Gauzy [13]	1	Right	15 y	Congenital pseudarthrosis of the clavicle	First sugery: RAK Second surgery: RAP	Good after the second surgery
1994	Shalom [32]	1	Right	45 y	The mobile pseudarthrosis without arthritic change	NT	Accepted disability and appearance
1999	Cataldo [20]	1	Right	7 m	Pseudarthrosis with the stump dysgenesis and the absence of a bony callus	NT	No FU
1964	Morrison [34]	1	Right	23 m	The right clavicle to be in two parts	RK	Union

Note: L means left side; B means bilateral sides; y means years old; m means month old; RAP means resection of the pseudarthrosis (R), and autologous iliac bone grafting (A) and internal fixation with plate (P). RAK means resection of the pseudarthrosis (R), and autologous iliac bone grafting (A), and internal fixation with Kirschner wire (K). RK means resection of the pseudarthrosis (R) and internal fixation with Kirschner wire (K). RP means resection of the pseudarthrosis (R) and internal fixation with plate (P). RA means resection of the pseudarthrosis (R) and autologous iliac bone grafting (A). RAS means resection of the pseudarthrosis (R), and autologous iliac bone grafting (A) and internal fixation with screw (S). RShin means resection of the pseudarthrosis (R) and autologous shin bone grafting (Shin). RS means resection of the pseudarthrosis (R) and internal fixation with screw (S). FU means follow-up. NT means no treatment.

outlet syndrome can result in symptoms affecting the arm and hand, including cold, fatigue, ischemia, necrosis, edema, paresthesia and thromboembolism, and a subclavian aneurysm [3]. The surgical procedures for CPC include resection of the pseudarthrosis, autologous iliac bone grafting and internal fixation [1-4, 8, 10, 12, 17-19, 23, 25, 26, 32]. The age for surgery of CPC patients is usually between 3 and 14 years, with the optimal age of surgery being between 5 and 7 years thus avoiding the shortening and asymmetric growth of the shoulder [7]. Bone grafts include autogenous bone, allograft bone and autologous stem cells [6], and autogenous ilium being the best option [6, 33]. Pain is the main complication of autogenous iliac bone grafting [4]. The choice of internal fixation devices are the Kirschner wire, the Steinmann pins, the threaded pins, the compression plate, with other external fixators possible [4, 8, 9, 18]. The compression plate could reach a stable fixation and quickly heal pseudarthrosis compared with the threaded pins [8]. Compared with the compression plate fixation, the threaded pin fixation has had more postoperative infections and nonunion of the pseudarthrosis. Postoperative complications include nonunion and delayed union of pseudarthrosis, local infection, sepsis, scaring of the incision site, and injury to the brachial plexus [2].

Conclusion

In summary, our study describes a case of congenital pseudarthrosis of the clavicle, which is a rare deformity. The etiology of CPC remains unclear. A painless growing mass can be found in the middle of the clavicle, with the function of the affected shoulder remaining normal. Surgery of CPC can completely restore the physiological function of the shoulder joint, and prevent thoracic outlet syndrome and improve the cosmetic appearance. The surgical procedures for CPC include resection of the pseudarthrosis, autologous iliac bone grafting and internal fixation.

Disclosure of conflict of interest

None.

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