Case Report

Congenital pseudoarthrosis of the clavicle with bifurcation

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Abstract

Congenital pseudoarthrosis of clavicle is a rare clinical entity. It usually presents as a swelling in the clavicular region at birth or soon after birth. Fitzwilliam's original description of 60 subtypes of congenital pseudoarthrosis of clavicle have addressed several anatomical variants, e.g. association with cervical rib and abnormally vertical and elevated upper ribs. However, congenital pseudoarthrosis of clavicle associated with bifurcation is an atypical anatomic variant. To the best of our knowledge, this variant has never been mentioned in the literature. In the present report, we have described this subtype of symptomatic congenital pseudoarthrosis of the clavicle with bifurcation and its possible management.

Key words: Brachial plexus, clavicle, duplication, pseudoarthrosis
MeSH terms: Brachial plexus, clavicle, congenital anomalies

Introduction

Congenital pseudoarthrosis of the clavicle is a rare condition, following its first report in 1910, many individual cases and case series have been published.[1-6] It usually presents as a painless swelling in the clavicle region at birth or soon after birth. The swelling can occasionally be associated with weakness and inability to push the arm while crawling due to excessive movement at the pseudoarthrosis site. The clavicle is no longer thought to develop in membrane, for endochondral centers of ossification have been clearly identified.[2] There is some debate as to whether there is one primary center or two. Failure of two centers of ossification to unite is a possible factor of etiological importance. Fitzwilliam's original description on hereditary cranio-cleido dysostosis have addressed several morphological variants of clavicle e.g. association with cervical rib and abnormally vertical and elevated upper ribs.[1,2] However, congenital pseudoarthrosis of clavicle associated with bifurcation is an atypical anatomic variant. To the best of our knowledge, this variant has never been mentioned in English language literature. In the present report, we describe a symptomatic congenital pseudoarthrosis of the clavicle with bifurcation and its possible management. The relationship between bifurcation at the pseudoarthrosis site and brachial plexus compression symptoms have also been addressed.

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Case Report

A 21 year old female patient presented with right sided shoulder pain, aggravated by overhead activity and lifting heavy weights. It was associated with occasional paraesthesia of the ring and little fingers on the right side. On examination, a bony hard lump was palpable in the middle third of right clavicle with an indentation, which was present since birth. An abnormal mobility was present between the sternal and acromial ends of clavicle. Neurological examination of bilateral upper limbs was normal except mild hypoesthesia in C_8 distribution over hand and forearm on the right side. Adson's test was negative. Bilateral pulses were equally palpable. No history suggestive of birth trauma was present.

Plain radiographs revealed two areas of pseudoarthrosis in the middle of clavicle. The clavicle was bifid in its middle and separated from the sternal fragment through false joints. The edges of false joints were rounded. The inferior part was broad and more rounded than its superior counterpart [Figure 1a]. Magnetic resonance imaging scan showed

compression of nerve roots inferiorly at the pseudoarthrosis site [Figure 1b].

The patient underwent surgery in the supine position on a radiolucent table with a sheet bolus under the shoulder. Through an 8 cm direct incision over the abnormal bony lump of the clavicle, the pseudoarthrosis was exposed [Figure 2a]. Fibrous tissue was resected from the pseudoarthrosis and frank abnormal mobility could be elicited, in both the superior and the inferior parts. Inferior part of the bifid clavicle was osteotomized and resected in an attempt to decompress the brachial plexus. The edges of the superior part were freshened and the canal was opened using a 3.5 mm screw tap. It was stabilized with a 3.5 reconstruction plate [Figure 2b]. The apparent defect size was 3 cm following resection and was reconstructed with a strut graft of the same length from the contra lateral iliac crest [Figure 2c]. Subcuticular Skin closure was done over negative suction drain. The upper limb was immobilized in a cuff and collar sling.

Wound healing was uneventful. Custom made abduction orthosis was given after surgery and gradual shoulder mobilization was started after 3 weeks. Time taken for complete incorporation of the bone graft was 10 weeks. The patient was relieved of her symptoms after surgery. Nerve conduction velocity was conducted at 30 months and showed significant improvement.

Discussion

It was thought that the clavicle develops from endochondral ossification with one center of ossification.[2] Researchers suggest that environmental factors and rather than genetic factors influence the development of congenital pseudoarthrosis of clavicle.[2] It affects mainly girls and is nearly always on the right side.[2] Theories of development of congenital pseudoarthrosis of clavicle include development of clavicle from two ossification centers in the embryonic stage.[2-3] It is believed that certain anatomical abnormalities such as cervical ribs or elevated upper ribs may cause compression of the subclavican artery between the clavicle and the first rib. The exaggerated arterial pulsations and the resultant pressure influences the development of pseudoarthrosis. Various other theories based on anatomical variants of pseudoarthrosis of clavicle have been defined in the English language literature [Table 1]. Alldred

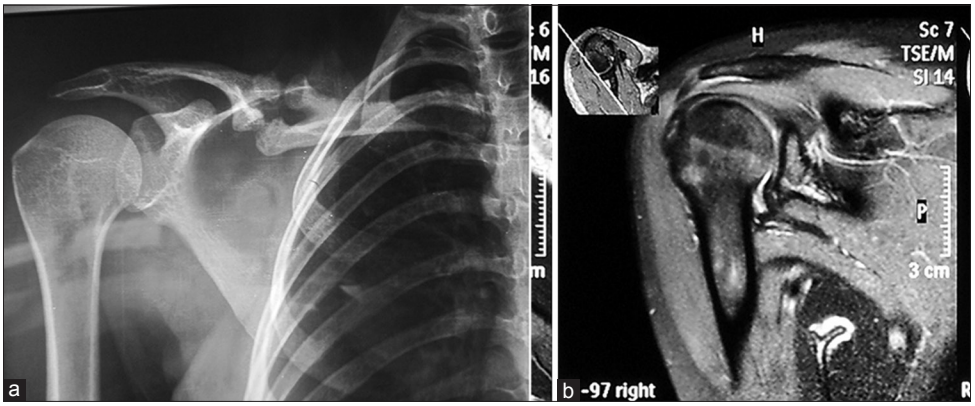

Figure 1: (a) Radiograph (R) clavicle anteroposterior view showing congenital pseudoarthrosis with bifurcation, (b) Magnetic resonance imaging scan showing compression of nerve roots at pseudoarthrosis site

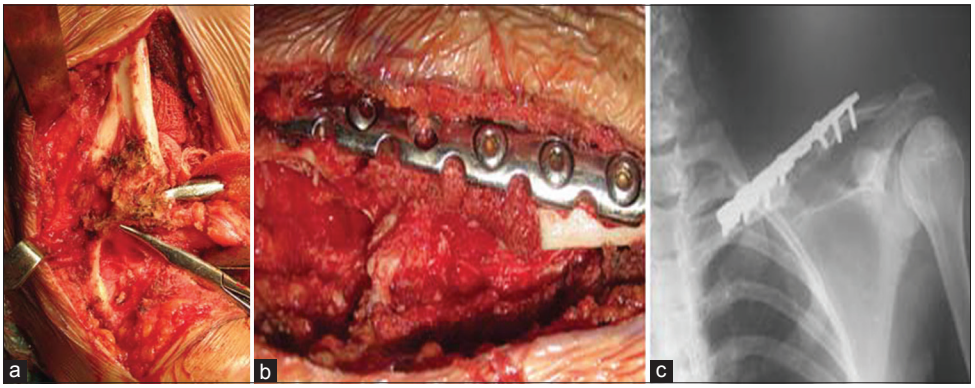

Figure 2: (a) Peroperative clinical photograph showing exposure of pseudoarthrosis region showing superior and inferior parts for bifurcated clavicle (b) Peroperative clinical photograph showing fixation of clavicle with reconstruction plate following resection of the inferior part of pseudoarthrosis with bone grafting (c) Followup Radiograph showing consolidation at autograft clavicle interface

Table 1: Literature review for morphological variants of clavicle

Authors	Year of study	Anatomical variant (CPC)
Golthamer	1957	Supernumerary bone over the clavicle
Oestreich	1981	Lateral clavicle hook variant
Ogden	1990	Neo-clavicle within periosteal sleeve
Dwivedi *et al.*	2011	Os subclaviculare variant
Present study	2013	CPC with bifurcation variant

CPC=Congenital pseudoarthrosis of clavicle

have suggested two centers of ossification for clavicle; failure of fusion between two centers has been cited as a possible etiology of congenital pseudoarthrosis.[3] Golthamer described bifid clavicle as a supernumerary bone just below clavicle.[4] Twigg and Rosenbaum in their study have reported a bifid clavicle and concluded that it was an anatomical variant.[5] Oestreich have suggested developmental origin for partial bifurcation of clavicle. They described a lateral clavicle hook variant.[6] Ogden have stated a traumatic etiology for bifurcation clavicle. The physis of bone is encased in thick periosteal sleeve. Following injury, a neo-clavicle is formed within this periosteal sleeve, separate from the displaced fracture fragments.[7] Pedersen and Frich have reported a similar bifurcation of clavicle following traumatic clavicle epiphysiolysis in a 15-year-old boy.[8] Dwivedi *et al*. have reported an os subclaviculare variant on computed tomography scan. They showed a well defined cortical bone below the clavicle, overlying the coracoid process. The lateral end of bone was rounded and the medial end was articulating with the coracoid process without any continuity with the clavicle.[9] The present case attempts to highlight the clavicular bifurcation in a background of congenital pseudoarthrosis.

Congenital pseudoarthrosis of the clavicle is an uncommon cause of brachial plexus compression.[10-12] Bargar *et al*. have reported a case of 28-year-old women with late presentation of thoracic outlet syndrome secondary to pseudoarthrosis of the clavicle. They performed the surgical resection of pseudoarthrosis and mid 3 cm of the clavicle with complete relief of symptoms following surgery.[10] Krishnan *et al*. have described eight cases of space occupying pseudoarthrotic clavicle nonunion associated with progressive brachial plexus compression. Though these cases were posttraumatic and not congenital, it highlights the significance of brachial plexus decompression. They had performed the resection of clavicle pseudoarthrosis, external neurolysis of brachial plexus and reconstruction of defect with free fibular flap secured with plates. All patients became free of pain and showed motor improvement following the surgery.[11] Young *et al*. described a case of 20-year-old woman with congenital pseudoarthrosis of the clavicle. She presented with features of thoracic outlet syndrome with both neurogenic and vascular symptoms. Surgical resection of pseudoarthrosis mass and internal fixation of the clavicle along with bone grafting was done.[12] The present case had sensory symptoms in C_8 dermatome before surgery, which could be due to pressure irritation of the lower trunk of brachial plexus by the inferior part of the bifurcated clavicle at the pseudoarthrosis site. The inferior part of pseudoarthrosis was resected in order to relieve this pressure. Patient was relieved of her neurological symptoms after surgery.

Fitzwilliam's in 1910 studied 60 cases for congenital pseudoarthrosis of the clavicle and defined its variants. None of the case in their series had bifid pseudoarthrosis lesion.[1] The present case adds a rare subtype of congenital pseudoarthrosis of clavicle with bifurcation to current literature.

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