ACTA
ORTHOPAEDICA
et
TRAUMATOLOGICA
TURCICA

Author's translation

Acta Orthop Traumatol Turc 2005;39(3):266-269

A case of congenital pseudarthrosis of the clavicle

Doğuştan klavikula psödoartrozu: Olgu sunumu

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Doğuştan klavikula psödoartrozu nedeni bilinmeyen nadir bir durumdur ve genellikle klaviküler bölgede ağrısız bir kitle şeklinde gözlenir. Bazen ağrı ve güçsüzlük şikayetleri oluşturabilmektedir. Sportif aktiviteler ve ağır günlük çalışma sırasında sağ omzunda ağrı ve güçsüzlük şikayetleriyle başvuran 21 yaşındaki erkek hastada doğuştan klavikula psödoartrozu saptandı. Cerrahi tedavide madial ve lateral fragmanların uçları sağlıklı kemiğe kadar eksize edildi; oluşan defekt iliyak kanattan alınan trikortikal otogreft ile onarıldı ve klavikula plak ve vidalarla tespit edildi. Hastanın yedinci aydaki son kontrolünde klinik olarak şikayetlerinin kaybolduğu, herhangi bir hareket kısıtlılığının olmadığı görüldü ve Constant skoru tam bulundu.

Anahtar sözcükler: Klavikula/patoloji; psödoartroz/doğuştan/cerrahi.

Congenital pseudarthrosis of the clavicle is a rare condition of unknown etiology. It usually presents as a painless mass in the clavicular region; however, it may sometimes be associated with pain and weakness in the shoulder. We presented a 21-year-old male patient who had pain and weakness in the right shoulder during sportive and daily vigorous activities due to congenital pseudarthrosis of the clavicle. The patient was successfully treated by excision of the ends of the medial and lateral fragments, reconstruction of the defect with a tricortical iliac crest autograft, and internal fixation of the clavicle with a plate and bone screws. After seven months postoperatively, his complaints disappeared, with a full range of motion and Constant score.

Key words: Clavicle/pathology; pseudarthrosis/congenital/surgery.

Congenital pseudoarthrosis of the clavicle (CPC) is a rare condition of unknown etiology. Although approximately a hundred patients have been reported to date, some of these cases do not comply with the definition of CPC.[1] CPC manifests as a painless mass at the midline. Rare cases associated with pain have also been presented.[2] Treatment is pursued for aesthetic reasons in girls and for the development of thoracic outlet syndrome in adults.[1,2]

An adult patient who has undergone surgical treatment for CPC is presented in this article.

Case report

A 21-year-old patient presented to the outpatient clinic with a congenital mass in the right shoulder as well as pain following physical activity and weakness that had been persisting for two years. He had no history of birth trauma and no complaints except a growing mass until 19 years of age. He reported that the pain became apparent during sports and vigorous daily activities. Besides, there was no family history of skeletal system pathology. Physical examination revealed a mass at the right clavicular mid-

**This study was presented as poster in congress of 3rd Turkish Shoulder Elbow Surgery. (March 31 – April 3, 2004, Ankara, Turkey)*
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Received: 05.06.2004 **Accepted:** 25.01.2005

line bulging through the skin and pathological movement (Figure 1a). Palpation of the region revealed antero-superior displacement of the medial fragment and postero-inferior displacement of the lateral fragment. A full range of motion was observed in the shoulder. No café-au-lait like skin lesions were detected. The Constant score[3] was 91. A pseudoarthrosis appearance with the medial fragment of the middle part of the right clavicle showing a superior displacement extending over the lateral fragment and the lateral fragment showing an inferior displacement was observed in the radiographic examination (Figure 1b). The ends of the fragments were round and no callus tissue was present on the pseudoarthrosis line. The patient was diagnosed as suffering from congenital pseudoarthrosis of the clavicle according to history, physical examination and radiographic findings. Surgical treatment was considered since the patient was young and the complaints prevented sports activities. Surgical intervention was carried out while the patient lay in beach-chair position. An incision parallel to the clavicle at 1 cm distance was made starting from the right clavicle extending cranially. The clavicle was exposed by stripping subperiosteally. No fibrous union or callus tissue formation was observed at the pseudoarthrosis region (Figure 2a). The atrophic ends of the fragments were excised up to the healthy bone. Following the excision, a tricortical autograft obtained from the iliac wing, in accordance with the size and shape of the defect, was implanted between the fragments, and the clavicle was fixed with titanium plates and screws fit for its shape (Figure 2b). The patient started doing shoulder exercises with active assistance at week one postoperatively and he used a simple shoulder sling for three weeks. Fusion started from both ends of the graft at the second postoperative month; the last follow-up visit at seven months revealed both fragments of the clavicle to have fully welded into the graft and the clavicle had continuity (Figure 3a). Moreover, clinical complaints had disappeared with a full range of motion and Constant score was full.

Discussion

The clavicle is the bone ossifying earliest in humans at the fifth or sixth week of intrauterine period. It is the only osseous link between the shoulder belt and the trunk. The etiology of congenital pseudoarthrosis of the clavicle is unknown. One opinion is that CPC originates from inadequacy of fusion of the two ossification centers of the clavicle.[4] However, embryonic studies have shown that there was only one ossification center in the clavicle.[5] Llyod-Roberts et al,[6] suggest that CPC develops due to pressure exerted by the subclavian artery on the developing clavicle. The right subclavian artery lies under and near the clavicle. Although this opinion explains why CPC appears predominantly on the right and the left side when dextrocardia is present, it does not elucidate the etiologic mechanism of left sided CPC in patients with bilateral involvement and no dextrocardia. Congenital pseudoarthrosis of the clavicle usually appears on the right side. The frequency of bilateral involvement is around 10%.[1] Hereditary trait of this condition is not clearly defined. Despite the presence of familial cases, genetic transmission appears to be recessive.[7] Yet, the general opinion is that CPC is not a genetic disease.[1] The finding that our patient had a right sided involvement and that there was no family history, is in accordance with findings in literature.

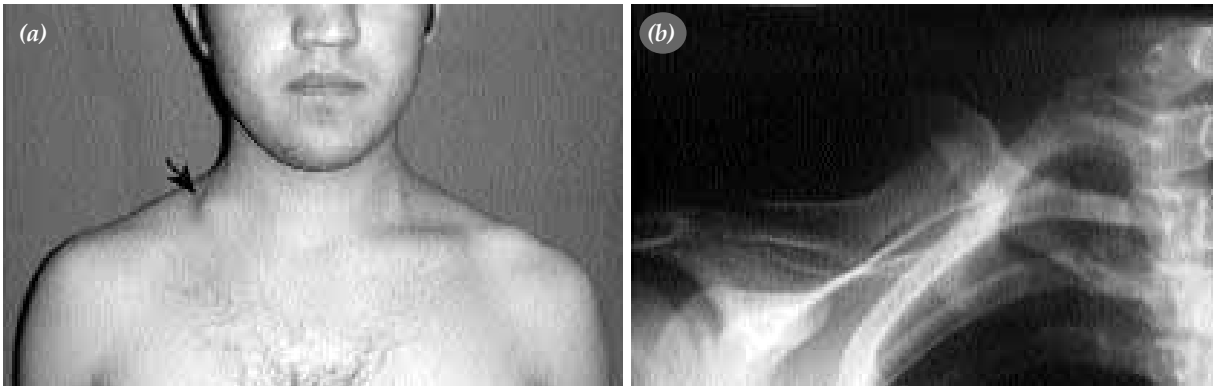

Figure 1. Preoperative **(a)** clinical and **(b)** radiographic appearance of the patient.

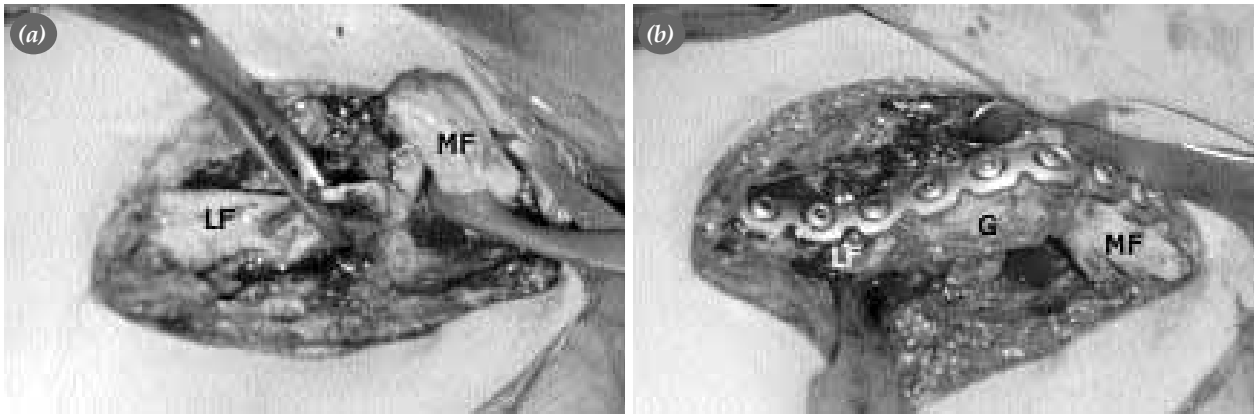

Figure 2. Images of **(a)** the excision of fragment ends during the operation and **(b)** the appearance of the tricortical iliac graft after fixation (LF: Lateral fragment; G: Graft; MF: Medial fragment)

Congenital pseudoarthrosis of the clavicle should be differentiated from cleidocranial dysostosis, neurofibromatosis and pseudoarthrosis due to trauma. The differential characteristics of CPC are lack of ossification problems in other bones of the body, lack of fibrous or osseous callus between the fragments and lack of café-au-lait like lesions on the skin.[1] Our case had no skeletal system pathology except pseudoarthrosis of the clavicle.

Spontaneous remission has not been reported in congenital pseudoarthrosis of the clavicle.[8] Treatment is surgical and is particularly undertaken in girls for aesthetic concerns.[8,9] Although the condition is considered asymptomatic, some cases may have complaints including pain and weakness and rarely thoracic outlet syndrome may also be present.[1,2,10] Our patient did not have findings of thoracic outlet syndrome. However, we decided to per-

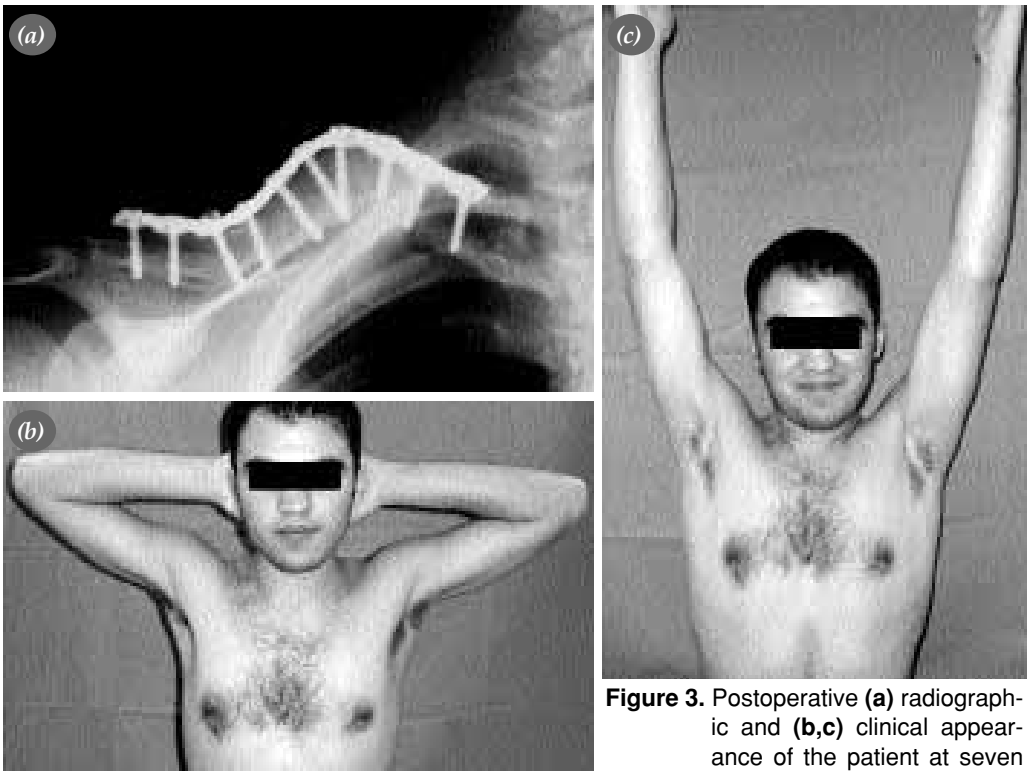

Figure 3. Postoperative **(a)** radiographic and **(b,c)** clinical appearance of the patient at seven months postoperatively.

form surgical intervention since he had pain and weakness severe enough to interfere with his daily activities.

Surgical treatment of congenital pseudoarthrosis of the clavicle includes resection of the atrophic fragments, autografting and internal fixation.[10] An apparent defect was detected to have formed in our patient following resection due to the atrophic nature of both fragments (particularly the medial fragment) in the pseudoarthrosis area. In addition, a straight clavicle could not be achieved due to inadequate mobilization of the sternoclavicular joint and the growth of the medial fragment anterosuperiorly. Thus, we chose a flexible implant, which concurrently allowed a rigid fixation, and we ensured maximum contact and adequate fixation of the implant. Complications such as lack of fusion, infection, neuropraxia of the brachial plexus and fracture of the clavicle after removal of the fixative material are very rare.[11-13] In the final control visit of our patient, fusion had been achieved, Constant score was full and daily activities were performed to the full.

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