

Ahmet Yiğit GÖKTAY
Mustafa SEÇİL
Erkin ÇETİNKOL
Diğç ÖZAKSOY

Congenital Pseudoarthrosis of the Clavicle with Multiple Vertebral Anomalies

Department of Radiology, Faculty of Medicine,
Dokuz Eylül University, İzmir - TURKEY

Received: April 18, 2001

Key Words: Clavicle - Congenital pseudoarthrosis - Vertebral anomalies

Congenital pseudoarthrosis of the clavicle is a rare anomaly which is considered to be a separate clinical entity and should be differentiated from birth fracture, cleidocranial dysostosis or neurofibromatosis [1-4]. Most patients with congenital pseudoarthrosis of the clavicle present in infancy or early childhood with a lump over the midportion of the clavicle. In this report, we present a case of congenital pseudoarthrosis of the clavicle with various vertebral anomalies in an adult patient.

Case Report

A 35-year-old woman, living in a rural area, was referred to our unit for radiographic evaluation of skeletal abnormalities. The patient had a history of back pain and a small lump over the right clavicle, which were present since her early childhood. The history excluded trauma and she had never received any medical care for the abnormalities. On physical examination, a non-tender mass was palpated over the right clavicle; a postural asymmetry and kyphosis were also noted. The radiological examination of the patient revealed that the mass was secondary to the pseudoarthrosis of the right clavicle (Figure 1). The clavicle showed an osseous separation with rounded well-corticated margins and large bone ends at the sites of the pseudoarthrosis. The cervical radiographs showed fusion of the bodies and the posterior elements at the levels of C2-3 and C3-4 vertebrae (Figure 2a). The dorsal and lumbar radiograms demonstrated roto-scoliosis and kyphosis with multiple vertebral abnormalities including butterfly vertebra and hemivertebra, various anomalies of the pedicles and transverse processes, and accompanying rib anomalies (Figure 2b). Cranial radiographs were normal.



Figure 1. Antero-posterior radiograph demonstrating the pseudoarthrosis of the clavicle.

Congenital pseudoarthrosis has been defined as a separate entity that differs from cleidocranial dysostosis, birth fracture and neurofibromatosis [1-4]. The etiology and pathogenesis of the entity is not well established. A familial incidence has been reported; however, most cases are sporadic [2]. The hypothesis of a non-union of the two ossification centers has been disputed as it has been demonstrated that the clavicle has a single ossification center [1,2]. A mechanical factor has been proposed to exist in the pathogenesis, where minor variations in the development of the subclavian artery have been postulated to be the reason, as it passes just below the clavicle particularly on the right side [2]. The left-sided involvement of the clavicle in congenital pseudoarthrosis is extremely rare and most cases are associated with dextrocardia resulting in the reversal of the relative positions of the subclavian arteries. Other cases supporting the mechanical factors involved the existence of well-formed or rudimentary cervical ribs with a fibrous



Figure 2. Radiographs showing vertebral multiple anomalies.
a. Lateral cervical radiograph showing the fusion of the bodies and the posterior elements of C2-3 and C3-4 vertebrae.
b. Antero-posterior radiographs demonstrating butterfly vertebra in T12, hemivertebra in L1, abnormally shaped lumbar vertebral lateral processes, and rudimentary 12th ribs.

connection to the first rib which results in the elevation of the first rib, and thus the subclavian artery theoretically plays a role in the development of the pseudoarthrosis [1-3]. This was not the case in our patient, but it may be proposed that the multiple vertebral anomalies and the resultant changes in the alignment possibly contribute to the development of pseudoarthrosis, as a mechanical factor.

Congenital pseudoarthrosis of the clavicle is usually diagnosed in early childhood with a palpable, non-tender mass over the clavicle, causing an asymmetric appearance. As in our case, the range of motion of the shoulder is commonly not affected and pain is an

infrequent symptom. Roentgenologically, the pseudoarthrosis is typically located in the middle third of the clavicle, close to its junction with the lateral third. The medial segment lies above the lateral segment, and the ends are enlarged and bulbous with no evidence of callus or reactive bone [1-4].

Congenital pseudoarthrosis of the clavicle is an anomaly confined to the clavicle and the coexistence of other skeletal abnormalities, apart from the cervical ribs, has not been previously reported. The case presented here is unique, being the first case of multiple vertebral anomalies accompanying congenital pseudoarthrosis of the clavicle.

Correspondence author:

Ahmet Yiğit GÖKTAY

Dokuz Eylül Üniversitesi, Tıp Fakültesi,

Radyodiagnostik Anabilim Dalı,

35340 İnciraltı, İzmir-TURKEY

References

1. Schnall SB, King JD, Marrero G. Congenital pseudoarthrosis of the clavicle: a review of the literature and surgical results of six cases. *J Pediatr Orthop* 8: 316-21. 1988.
2. Schoenecker PL, Johnson GE, Howard B, Capelli AM. Congenital pseudoarthrosis. *Orthop Review* 21: 855-60, 1992
3. Grogan DP, Love SM, Guidera KJ, Ogden JA. Operative treatment of congenital pseudoarthrosis of the clavicle. *J Pediatr Orthop* 11: 176-80, 1991
4. Manashil G, Laufer S. Congenital pseudoarthrosis of the clavicle. Report of three cases. *AJR* 132: 678-9, 1979