

BILATERAL CONGENITAL PSEUDARTHROSIS OF THE CLAVICLE REPORT OF A CASE WITH CLINICAL, RADIOLOGICAL AND NEUROPHYSIOLOGICAL EVALUATION

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Congenital pseudarthrosis of the clavicle is a rare condition. Bilateral involvement is extremely rare : only seven cases have been reported in the literature. Although the anatomy of the thoracic outlet can be markedly altered by the hypermobility of the shoulder and although few cases with mild symptoms suggesting brachial plexus impairment have been reported, the neurological status of the brachial plexus in congenital pseudarthrosis of the clavicle has not been well assessed. We report a case of bilateral congenital pseudarthrosis of the clavicle in which clinical, neurophysiological and radiological evaluations were performed.

Keywords : congenital pseudarthrosis of the clavicle ; neurophysiological evaluation ; treatment.

Mots-clés : clavicule ; pseudarthrose congénitale ; évaluation neurophysiologique.

INTRODUCTION

Congenital pseudarthrosis of the clavicle is a rare condition, which was described for the first time by Fitzwilliam in 1910, in association with cleidocranial dysostosis, distinguishing it from birth fracture. Several authors later described the typical features of this condition (1, 4). Clinically, it presents as a prominence at the midportion of the clavicle without pain. Radiological examination shows a defect of osseous tissue in the middle clavicle and rounded sclerotic bone ends.

The lesion is usually unilateral and localized on the right side. Bilateral occurrence is extremely rare : to our knowledge, only seven cases have been reported in the literature (1, 2).

The marked hypermobility may cause a temporary narrowing of the thoracic outlet with consequent brachial plexus impairment and outlet syndrome (5). Some authors reported cases with symptoms that could be related to brachial plexus impairment (1, 4), but no complete neurological evaluation was reported, and neurophysiological studies were never performed.

We report clinical, radiological and neurophysiological evaluation in a case of bilateral congenital pseudarthrosis of the clavicle in a 5-year-old child, who was not treated surgically.

CASE REPORT

The patient was seen for a suspected clavicular fracture soon after birth and has been regularly followed over the past 5 years. Clinically, there were no tender bumps in the middle of either clavicle. Neither pain nor functional deficits were observed in the arms and shoulders, and there were no café au lait spots on his skin. The diagnosis of bilateral congenital clavicular pseudarthrosis was based on radiographic examinations (fig. 1). Other associated abnormalities were

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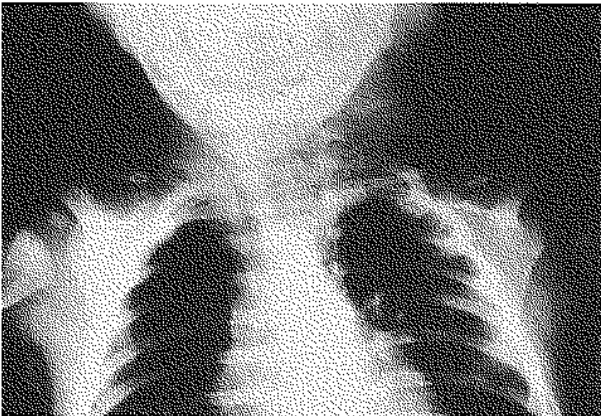


Fig. 1. — Radiographic appearance of bilateral clavicular pseudarthrosis shortly after birth.

excluded by radiographic examination of the spine and cranium and the patient underwent yearly radiographic and clinical evaluation.

At the time of the last observation, the child, who is now 5 years old, had no pain or discomfort at shoulder movements bilaterally. The range of motion of the shoulder joints was completely normal except for greater anteflexion (fig. 2). In view of this marked hypermobility of the shoulders, in order to exclude subclinical impairment of the brachial plexus, we performed motor and sensory nerve conduction studies of the brachial plexus and of the nerves of the upper limbs; no abnormal conduction findings were made.

Follow-up radiographs confirmed bilateral defects in the middle clavicles. The sternal part of the clavicles was above the acromial part, and no reactive bone was visible between the ends of the segments (fig. 3). Surgery was not recommended because of the complete absence of functional limitation, pain or cosmetic problems.

DISCUSSION

The etiology of congenital clavicular pseudarthrosis has not yet been well established, and several theories have attempted to explain this condition. Lloyd-Roberts (4) suggested anatomical anomalies of the subclavian artery, situated at a higher than normal level during intrauterine life,

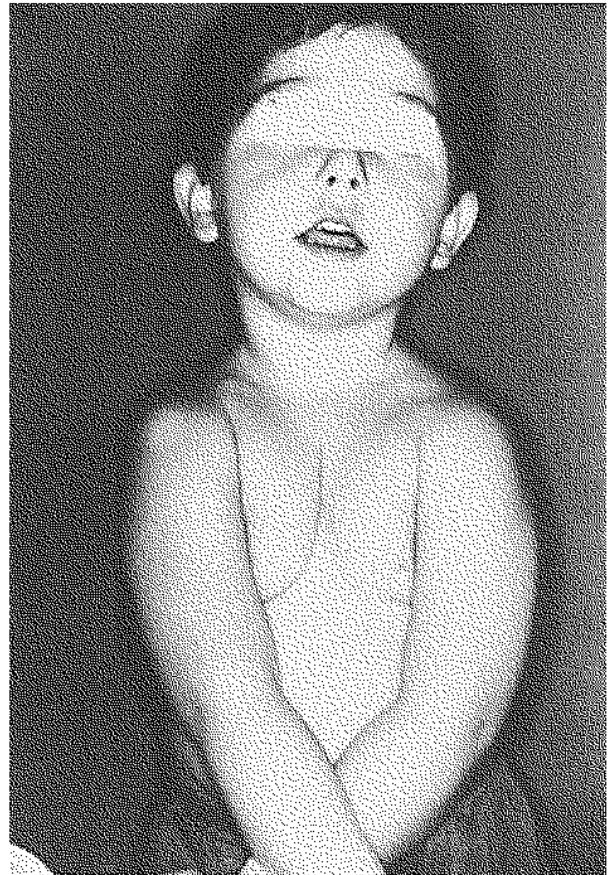


Fig. 2. — Hypermobility in anteflexion of the shoulders.

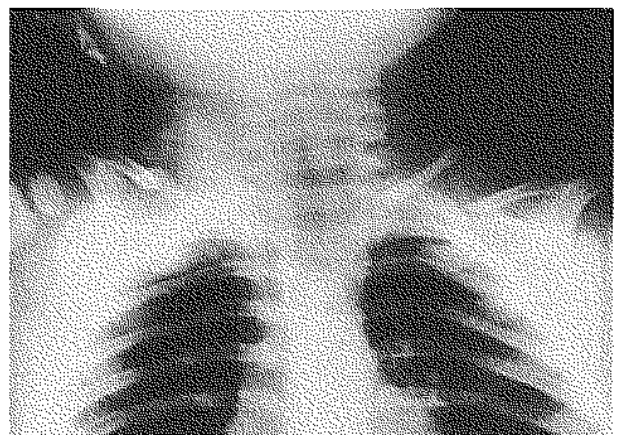


Fig. 3. — Radiographic control at the age of five years.

but the bilateral occurrence of the condition does not support this theory and suggests other possibilities. As reported by Hirata *et al.* (3) a lack of fusion of two ossification centers with a separation of the clavicles into two parts could be a possible etiology. Nevertheless Gibson and Carrol (1) observed only one single ossification center, and they stated that the etiology of this condition is still obscure.

Congenital pseudarthrosis of the clavicle should be differentiated from cleidocranial dysostosis, neurofibromatosis or from simple fracture. Bilateral pseudarthrosis especially may be confused with cleidocranial dysostosis. In the latter, all parts of both clavicles are deficient, and associated anomalies are seen in the skull, facial bones, pelvis, hands and feet. In congenital pseudarthrosis of the clavicle there are no other skeletal anomalies present. Unlike congenital pseudarthrosis of the tibia, there is no relationship between congenital pseudarthrosis of the clavicle and neurofibromatosis. Congenital pseudarthrosis can be differentiated from a simple fracture by the typical roetgenographic features. Unlike a fracture, spontaneous union does not occur, and there is no callus formation.

The bilateral form is extremely rare but must, nevertheless, be considered on clinical presentation of bilateral prominences without pain in the midportion of the clavicles.

Treatment of this condition has not been well standardized. Several authors have recommended the use of both surgical and conservative therapy (3, 5). No study reported neurological evaluation as a parameter for therapeutic approach.

Although the anatomy of the thoracic outlet can be markedly altered because of the hypermobility of the shoulder and although some cases with mild symptoms suggesting brachial plexus impairment have been reported, the neurological status of brachial plexus in congenital pseudarthrosis of the clavicle has not been well assessed.

In our case no clinical or subclinical neurological impairment was observed. We think that, especially in children who are unable to report slight neurological symptoms, the neurological status must be carefully assessed through extensive clinical evaluation in all cases (uni- or bilateral), and when

neurological impairment is suspected neurophysiological assessment must be performed.

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SAMENVATTING

R. PADUA, E. ROMANINI, C. CONTI, L. PADUA, F. SERRA. *Congenitale pseudo-artrose van de clavícula.*

Congenitale pseudo-artrose van de clavícula is een zeldzaam letsel. De bilaterale aantasting werd slechts zeven maal teruggevonden in de literatuur. Niettegenstaande dat het aspect van de thorax flink kan worden gestoord door de instabiliteit van de schouder en dat er enkele gevallen werden beschreven met plexus brachialis compressie, werd de neurologische status in geval van congenitale pseudo-artrose van de clavícula nooit in detail bestudeerd. De auteurs beschrijven een geval van congenitale pseudo-artrose van de clavícula met zijn klinische, neurofysiologische en radiologische evolutie. Er werd geen enkele behandeling voorgesteld gezien het ontbreken van een functioneel of esthetisch probleem.

RÉSUMÉ

R. PADUA, E. ROMANINI, C. CONTI, L. PADUA, F. SERRA. *Pseudarthrose congénitale bilatérale de la clavicle. Présentation d'un cas avec évaluation clinique, radiologique et neurophysiologique.*

La pseudarthrose congénitale de la clavicle est une lésion rare. L'atteinte bilatérale est plus rare encore : les auteurs n'en ont trouvé que 7 cas rapportés dans

la littérature. Bien que l'anatomie du défilé thoracique puisse être sensiblement altérée par l'instabilité de la ceinture scapulaire, et même si l'on a rapporté quelques cas de patients qui présentaient des signes de souffrance du plexus brachial, l'état neurologique du plexus brachial dans la pseudarthrose congénitale de la clavicule n'a jamais été étudié en détail. Les auteurs rapportent

un cas de pseudarthrose congénitale bilatérale de la clavicule qui a fait l'objet d'une évaluation clinique, neurophysiologique et radiologique.

L'étude neurophysiologique n'a révélé aucune anomalie. Aucun traitement n'a été proposé, en l'absence de déficit fonctionnel ou esthétique.