CASE REPORT

Venous Thoracic Outlet Syndrome Secondary to Congenital Pseudoarthrosis of the Clavicle. Presentation in the Fourth Decade of Life

P. Lozano*, M. Diaz, R. Riera and F. T. Gomez

Angiology and Vascular Surgery Department, Hospital Son Dureta, Palma de Mallorca, Spain

Key Words: Congenital pseudoarthrosis; Clavicle; Thoracic outlet.

Introduction

Congenital pseudoarthrosis of the clavicle is a rare disorder that should not be confused with the more common form of pseudoarthrosis that occurs secondary to the fractured clavicle. The lesion is usually unilateral and more often affects the right side. Although the condition is usually asymptomatic, it may raise cosmetic concerns, minor problems, and functional problems, rarely it can cause thoracic outlet syndrome. Here, a case leading to symptomatic subclavian vein compression is presented.

Case Report

A 48-year-old, right-handed woman presented with a 5-year history of worsening swelling, heaviness, paraesthesia (ulnar distribution) and weakness affecting her right upper limb. There was no history of trauma. On examination, the patient was obese and the margin of the clavicle could not be perceived, there was oedema leading to a 4 cm differential in arm circumference, also a network of dilated superficial veins around the shoulder and no blood pressure differential between the two arms. A chest X-ray (Fig. 1) showed lack of union at the middle third of the clavicle, with a significant reduction in the diameter of the sternal (medial) fragment, which was located at a higher position than the lateral fragment. Duplex ultrasonography excluded thrombosis of the subclavian and axillary veins. Venography, performed with the arm in neutral position, revealed extrinsic compression of the subclavian vein by the sternal segment of the clavicle (Fig. 2), which was exacerbated on abduction of the arm. Adson’s test was negative and nerve conduction studies were within normal values. A diagnosis of thoracic outlet syndrome due to congenital pseudoarthrosis of the clavicle was made. As the patient’s symptoms and signs failed to improve following a 6 month course of exercises and physiotherapy, surgical decompression was undertaken. Specifically, through a transverse incision centred on the middle edge of the clavicle, the pseudoarthrosis and the sternal fragment of the clavicle were resected. Following resection, the absence of

Fig. 1. Chest X-ray. Congenital pseudoarthrosis of the clavicle: lack of union at the middle third of the clavicle, decrease of the medular diameter of the sternal medial fragment, located at a higher position than the lateral fragment.
arterial, venous and neural compression was confirmed and so the first rib was left in situ. Post-operatively, recovery was uncomplicated, the patient has been rendered asymptomatic and the oedema has improved.

Discussion

Normally, the clavicle is formed from the fusion of two ossification centres, which appear between the 5th and 7th weeks of intrauterine life. The prevalence and aetiology of true congenital clavicular pseudoarthrosis is unknown. One theory takes into account the observation that the condition is commoner on the right (but on left in patients with dextrocardia) and proposes that it is due to pressure exerted by the subclavian artery (which lies higher on the right). A second suggests it is due to the separation of the two primary ossification centres. The condition usually presents during childhood as a small, slow growing, usually asymptomatic, lump over the middle third of the right (10% cases are bilateral) clavicle which rarely requires treatment. In adults, the condition has been associated with thoracic outlet syndrome. In such cases, the requirement for, and the nature of surgical intervention remains controversial. Combinations of corrective osteotomy, bone graft interposition and internal fixation have been proposed but can be associated with significant morbidity. In the present case, simple resection of sternal segment of the clavicle led to an excellent symptomatic and cosmetic outcome.

References


Accepted 6 January 2003