Findings that shed new light on the possible pathogenesis of a disease or an adverse effect

Congenital pseudarthrosis of clavicle: illustrated operative technique and histological findings

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Summary

Congenital pseudarthrosis of the clavicle is a rare condition present at birth but often diagnosed later in childhood. Indications for surgical treatment include pain, deformity or neurovascular compromise. Reconstruction usually involves resection of the pseudarthrosis, placement of iliac crest bone graft or graft substitute and internal fixation. In this paper, we present a case of congenital pseudarthrosis of the clavicle in a 9-year-old boy who was treated with plate fixation and bone autograft. The majority of patients who undergo surgery because of cosmetic or functional problem heal well and proceed with a normal, unrestricted life. However, for those patients who are not bothered by the cosmetic appearance of the pseudarthrosis and are asymptomatic in that they are not functionally limited, non-surgical treatment is a viable option.

BACKGROUND

Congenital pseudarthrosis of the clavicle (CPC) is a rare condition of unresolved aetiology and pathogenesis. The condition was initially described by Fitzwilliams¹ as a distinct variant of cleidocranial dysostosis in 1910. CPC has since appeared in a number of studies involving more than 200 patients. However, very few articles present the histology of the lesion²⁻⁴ along with intraoperative pictures illustrating the surgical technique.

This malformation appears to be 25–80% more common in females than males. Usually, the condition is unilateral and occurs on the right side, but in approximately 10% of the cases involvement is bilateral. Left-sided unilateral involvement is extremely rare and is often associated with dextrocardia or cervical ribs.⁵

Congenital pseudarthrosis of the mid-portion of the clavicle occurs when an environmental insult or anatomic or mechanical event disrupts diaphyseal membranous ossification. The clavicle is the first bone to undergo primary ossification. In congenital pseudarthrosis of the clavicle, the two primary ossification centres fail to unite. The two portions are connected by a fibrous bridge that is contiguous with the periosteum, and a synovial membrane develops.⁵

Certain anatomical abnormalities are predisposing factors for CPC. It is believed that cervical ribs or vertically oriented upper ribs may cause compression of the subclavian artery between the clavicle and first rib.⁶ The exaggerated arterial pulsations and pressure which result could influence the development of a pseudarthrosis. This theory suggests that the increased incidence of right-sided CPC may be related to the fact that the right subclavian artery is normally elevated to a point directly underneath the clavicle.³

A painless mass or swelling over the clavicle is the usual presenting concern. The patient has no history of trauma,

and shoulder and arm movement are normal. At the site of the pseudarthrosis, the ends of the clavicular segments are enlarged and there is a degree of motion between them.⁷ The deformity tends to become more obvious as the child grows and marked mobility is found at the site of the pseudarthrosis. Moderate pain may be present around the shoulder girdle and upper arm, while a few patients appear to have functional problems such as weakness of the arm and limited shoulder abduction.⁷

CASE PRESENTATION

A 9-year-old boy presented with a visible lump over the right clavicle. His recent history included a fall which had increasingly become painful; shoulder and arm movements were normal although the patient had a certain degree of difficulty during overhead activities. Plain radiographs (figure 1) revealed clear separation but no displacement of the clavicular diaphysis. The CPC is atypical in that the medial segment does not override the lateral segment, presenting instead an example of a broad-ended pseudarthrosis.

TREATMENT

Surgical treatment was offered to correct the appearance and the functional problems in the right shoulder and arm.

At operation, a thick pseudocapsule around the pseudarthrosis was found. The bone end on each side looked smooth and was covered by fibrocartilage (figure 2). Material was obtained from the bone ends for histology and the operative specimen showed sclerosis of subchondral bone and a well-formed cartilage cap at the site of the pseudarthrosis (figure 3).

Excess bone was removed to contour both edges and was used as local morcelised bone graft. The area of the pseudarthrosis was reduced, held with a reduction forceps

Figure 1 Preoperative radiographs reveal clear separation with hypertrophy of both bone ends.

and a lag screw was inserted from anterior to posterior. Subsequently, a five-holed reconstruction plate was contoured and applied on the superior surface of the clavicle as a neutralisation plate and the length of the clavicular diaphysis was re-established. A polysling was used for 6 weeks for immobilisation. Overhead and repetitive movements were prohibited for 12 weeks.

OUTCOME AND FOLLOW-UP

Postoperative radiographs showed complete union of the pseudarthrosis after 8 months (figure 4), and the patient regained a full pain-free range of motion and symptom-free function of the shoulder.

DISCUSSION

Congenital pseudarthrosis of the clavicle is different from obstetric fracture, post-traumatic non-union and cleidocranial dysostosis.

An obstetric fracture of the clavicle should be suspected when there is a history of a difficult delivery, pseudoparalysis of the arm with no voluntary limb movement and pain on passive movement, or when a massive callus is seen radiographically.⁶ The feature that distinguishes congenital pseudarthrosis from cleidocranial dysostosis is the absence of other skeletal abnormalities such as skull

Figure 2 At operation both bone ends looked smooth and were covered by cartilage.

Figure 3 Histology of the operative specimen shows subchondral bone sclerosis with a well-formed cartilage cap at the side of the pseudarthrosis.

deformities, small size of the facial bones, scoliosis and deficiencies of the pelvis.⁵

Surgical treatment should be considered for symptomatic patients with dysfunction of the arm,⁸ when patients are embarrassed by the appearance of the lump,^{2 9} and to prevent thoracic outlet syndrome in the future.¹⁰ Surgical treatment consists of excision of the pseudarthrosis and internal fixation of the bone ends with bone grafting. In this case, the patient was complaining of mild pain and a certain degree of discomfort during overhead activities along with a slight increase in the size of the mass. These symptoms prompted us to proceed to surgical treatment of this congenital deformity.

In childhood, congenital pseudarthrosis of the clavicle does not usually disturb function and if the prominence can be accepted, it is a benign condition. The reported complications of surgery include sepsis, scars on the shoulder and at the donor site of bone grafts and delayed or non-union requiring reoperation.^{7 11} Brachial plexus injury has also been reported.¹²

Figure 4 Postoperative radiograph shows complete healing of the area of the pseudarthrosis with no signs of hardware loosening.

Learning points

- ▶ Even simple resection of the pseudarthrosis to improve the cosmetic appearance has sequelae which include a potential difference in length and underdevelopment of the distal fragment with drooping of the shoulder.^{6 8}
- ▶ Conservative management appears to leave mild swelling with no functional disability, and should therefore be considered in asymptomatic individuals.
- ▶ Radiographic and histological findings in congenital pseudarthrosis are different from other forms with an acquired lesion.²

Competing interests None.

Patient consent Obtained.

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