



Clinical Group

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Case Report

Congenital pseudarthrosis of the clavicle in a 7-year-old girl: A case report treated with smooth elastic intramedullary pinning without bone grafting and literature review

Abstract

The authors report a case of congenital pseudarthrosis of the right clavicle in a 7-year-old girl. It is a rare congenital malformation. Unanimity is far from being achieved on the best therapeutic method to adopt. This case benefited from a stable elastic medullary pinning without graft with an encouraging result.

Introduction

Congenital pseudarthrosis of the clavicle is a rare clinical entity. The literature reports to date only 200 clinical cases in the world. The first known description of this pathology dates back to 1910. It is mostly described in girls and often affects the right clavicle. Bilateral, familial forms have been reported. The diagnosis is both clinical and radiological. Consensus is still far from clear to the therapeutic attitude to adopt. We report the case of a 7-year-old girl, the first one encountered in our practice for nearly 20 years, and the second case described in Africa to the best of our knowledge. We treated it surgically with a satisfactory anatomical and functional result.

Case Presentation

We report the case of a 7-year-old girl borne normally at term but with a low birth weight of 2,200kg. There is no family history of this pathology among the parents and siblings. She was brought by her parents to our outpatient department complaining of pain and swelling of the left wrist without prior trauma. On the systematic examination of this girl, a swelling of the posterior aspect of the left wrist strongly suggesting a synovial cyst was noticed. On further examination, we noticed a swelling around the middle 1/3 of the right clavicle visible and palpable under the skin. The parents reported that this swelling has been existing since the birth of the child. (Figure 1).

The comparative radiographic assessment of the two wrists showed no bone pathology (Figure 2).

However, the comparative x-ray of the 2 clavicles revealed a congenital pseudarthrosis of the right clavicle (Figure 3).

We prescribed an analgesic treatment to relieve the pain caused by the left wrist cyst. We as well suggested and discussed surgical treatment of this pseudoarthrosis of the clavicle and explained its necessity if she complains of pain, functional or aesthetic discomfort. We reviewed this child after about 1 year, complaining of an aesthetic discomfort that became more and more apparent as the child grew up. We then proposed surgical treatment and discussed it with the parents. The procedure consisted in performing a direct approach to the pseudarthrosis focus under general anesthesia. We



Figure 1: N.G., 7 years old, presenting with a palpable and visible swelling under the skin around the middle 1/3 of the right clavicle (blue arrow).

then excised the fibrosis exposing the fresh ends of the bone fragments followed by reduction and internal fixation with smooth flexible intramedullary pins (Figure 4), this construct was supplemented by an immobilization of the shoulder with an elbow-to-body sling.

The shoulder sling was removed 6 weeks after surgery.

The clinical evolution was marked by a satisfactory consolidation of the pseudarthrosis focus, this motivated us to perform implant removal on day 305 postoperatively. The radiograph of the clavicle done on day 435 shows a complete consolidation of the pseudarthrosis focus (Figure 5).



Figure 2: The comparative x-rays of the two wrists in antero-posterior and lateral views are normal.



Figure 3: Comparative x-ray of the 2 clavicles showing a congenital pseudarthrosis on the middle 1/3 of the right clavicle (white arrow).



Figure 4: The postoperative X-ray on Day 21 shows good stability of the construct without secondary displacement of the intramedullary flexible pins.



Figure 5: The last postoperative check-up on day 435 shows good consolidation with good cortication of the congenital pseudarthrosis.

Discussion

Epidemiological aspect

The most important published series emphasized the predominance of the female sex: 0.3 and 0.8 respectively according to Cadilhac et al. [1], and Di Gennaro et al. [2]. The age of our patient, 7 years, corroborates with the average age of diagnosis of congenital pseudarthrosis which varies from 7.1 years on average for Studer et al. [3] to 11.5 years for Cadilhac et al. [1].

Aetiological aspect

Congenital pseudarthrosis of the clavicle is due, according to Lloyd-Roberts et al. [4], to a defect of fusion of the 2 points of ossification of the clavicle. But the real mechanism of the origin of this pathology is unknown. The theory advanced by these authors would be the constant and abnormally high pressure exerted by the subclavian artery which is normally high on the right side. This pressure could be increased by the presence of the first ribs on this side.

The rare involvement of the left clavicle would be linked to a dextrocardia and the existence of an abnormally large cervical rib. Lombard [5] also evokes a malposition of the fetus.

The familial cases reported by Cadilhac et al. [1], and Price et al. [6], and in twins by Akman et al. [7] suggest a genetic cause.

Clinical aspect

Pseudarthrosis of the clavicle is most often an incidental discovery [8] as in our case, and rarely symptomatic. It can be revealed by pain, neurological disturbances [9] or by a disgraceful cosmetic appearance that leads parents to consult [10]. It rarely presents with complication such as the thoracic outlet syndrome [9, 11, 12 and 13]. This last eventuality would be the result of a long evolution in the adolescent or the adult age. Gamier et al. [14] reported 3 arterial complications of thoracic paralysis syndrome and recommended Doppler ultrasound of the subclavian or selective arteriography. A simple comparative X-ray of the 2 clavicles often makes it possible to make the diagnosis of congenital pseudarthrosis. In case of doubt, a CT scan with 3D reconstruction images confirms the diagnosis and helps in the choice of therapeutic methods [15]. However, congenital pseudarthrosis of the clavicle may have the appearance of post-traumatic pseudarthrosis. This eventuality, in addition to the asymptomatic aspect of our case, prompted us to observe our case for a few months before the surgical treatment which finally was very necessary to remedy the aesthetic prejudice.

Therapeutic aspect

Most authors agree on a therapeutic abstention in cases of asymptomatic pseudarthrosis with an acceptable cosmetic aspect [16, 17]. The treatment modalities depend on the age and severity of congenital pseudarthrosis. Some authors recommend excision of the fibrosis, autologous cortico-

spongy graft with internal osteosynthesis (DCP plate, pins) or external fixation (external fixator) [2, 10, 13, 18– 22]. Grogan *et al.* [23]. carried out only an excision of the pseudarthrosis while preserving the periosteum without the addition of graft or internal fixation with a satisfactory consolidation in 14 weeks in 8 children. Chandran [24] advocates excision-grafting and internal fixation by a DCP plate for faster consolidation with fewer complications compared to the use of threaded pins in a comparative study of the 2 methods of open reduction and internal fixation.

In our case we carried out a thorough excision of the fibrosis and internal fixation with intramedullary pins according to the technique described by Métaizeau [25] reinforced by immobilization of the shoulder by a elbow-to-body sling, which made it possible to obtain adequate consolidation without complication.

Conclusion

The diagnosis of pseudarthrosis of the clavicle requires a complete history, a brief clinical examination to appreciate the symptomatic, functional and aesthetic repercussions. Therapeutic abstention is still a part of the therapeutic arsenal, but literature and our case teach us that surgery in children can produce quite satisfactory results with very few complications.

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