CASE REPORT

Congenital pseudarthrosis of the clavicle treated by Masquelet technique

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SUMMARY

Congenital pseudarthrosis of the clavicle (CPC) is a very rare pathology of which over 200 cases have been reported. Usually discovered during the first months of life, CPC is characterised by a definitive bone defect in the middle third of the clavicle. Generally asymptomatic, the pseudarthrosis of the clavicle can cause aesthetic issues and functional symptoms indicating a surgical repair. Different reconstruction techniques have been reported with various complications. We present a 14-year-old boy diagnosed with CPC and concerned about the cosmetic aspect of a swelling of his right clavicle. We performed a two-stage surgical repair using the induced membrane technique described by Masquelet. A complete bone union has been obtained, and patient is satisfied with the cosmetic appearance. In our opinion, the Masquelet technique is a safe and reproducible procedure to treat CPC in high-risk older patients with long gap allowing a rapid and lasting bone union.

BACKGROUND

Congenital pseudarthrosis of the clavicle (CPC) is a rare congenital pathology that is characterised by a definitive bone defect in the middle third of the clavicle. It is usually discovered during the first months of life, and it is more common in girls and in the right side.¹ Generally asymptomatic, pseudarthrosis of the clavicle can be responsible for aesthetic issues and functional symptoms indicating a surgical repair.^{2,3} Surgery is still debated but always includes excision of the non-union with or without bone grafting and stabilisation.⁴ Rare and various postoperative complications are reported, but the non-union is the most seen.⁵

The induced membrane technique described by Masquelet *et al*^{6,7} for bone regeneration in the

Figure 2 Resection of the hypertrophic bone extremities and fixation by a centromedullary Kirschner wire.

metaphyseal and diaphyseal regions of long bones has recently shown its effectiveness in the treatment of congenital pseudarthrosis of the tibia in children.⁸

The aim of our work is to describe the successful use of the Masquelet technique to address a case of congenital pseudarthrosis of the right clavicle.

CASE PRESENTATION

A 14-year-old boy was referred to our paediatric orthopaedic surgery department with swelling in the midpoint of the right clavicle found by the parents when he was 6 months. A conservative management was proposed at that age in another department. At adolescence, the boy and the parents were concerned about the cosmetic aspect and demanded surgery. There were no history of obstetrical trauma or previous fracture and no familial similar disorders. On examination, the mass-like swelling of the middle third of the clavicle was not painful. There was no functional, vascular or neural deficit at the upper extremity.

INVESTIGATIONS

The radiological assessment has shown a typical aspect of CPC with a hypertrophic non-union

Figure 1 Radiological aspect of congenital pseudarthrosis of the clavicle.

Figure 3 Replacement with surgical cement.

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Figure 4 Radiological aspect after the first step.

(figure 1). The bone gap was calculated to 2.3 cm (13% of the total length of the clavicle).

TREATMENT

A two-stage surgical repair using the induced membrane technique was proposed. Initially, resection of the hypertrophic bone (figure 2), recanalisation of the medullary canal and replacement of a 3.5 cm gap with 35 cm³ of surgical cement (polymethyl-methacrylate with standard viscosity) were completed with fixation by a centromedullary Kirschner (K) wire (figures 3 and 4). The second stage was done 6 weeks later by carefully opening the induced membrane and removing the cement by fragmentation leaving in place the K-wire. The bone at both ends was decorticated. The left cavity surrounded by membrane was filled with autologous corticospinous fragmented graft (figure 5) taken from the anterior iliac crest. The membrane was sutured, and the skin was closed without drainage. The shoulder was immobilised by Mayo clinic technique for 8 weeks.

OUTCOME AND FOLLOW-UP

Complete bone union has been obtained 10 weeks after the second stage, and wire removal was performed at that time under local anaesthesia (figure 6). The examination showed a pain-free shoulder function with good cosmetic results (figure 7). With 10 months follow-up, no complication has been observed, and the patient is satisfied with the cosmetic appearance.

DISCUSSION

CPC is a rare pathology of which over 200 cases have been reported in the literature.¹ It is largely documented as being a girl right-sided condition with left-sided cases (5%) usually associated with dextrocardia or situs inversus.¹ Currently, most

Figure 5 Radiological aspect after the second step.

Figure 6 Complete bone union has been obtained after 10 weeks.

authors believe that the condition is caused by extrinsic pressure exerted on the budding clavicle by the adjacent pulsatile subclavian artery.⁹ Characteristically, radiological features of CPC include clear separation in the middle portion, with the medial fragment positioned above the lateral fragment. The lateral extremity is more voluminous than the medial extremity which is atrophic with sclerotic closure of the medullary canal.¹⁹

The indications for surgery were mainly due to the aesthetic impairment, whereas in other cases, they were due to progressive pain, functional limitation, late-starting thoracic outlet syndrome or a combination of these factors.^{2 10}

Authors agree that the best time for surgery is between 4 and 7 years suggesting that early treatment in all cases avoids complications.^{4 11–16} Furthermore, in older child, the size of the gap tends to increase, and more difficulties may occur specially with a longer time to heal, non-union and prominent hardware.^{11 15–17}

There are multiple surgical options for the treatment of CPC all including excision of the pseudarthrosis plus internal fixation (plates and screws, K-wires and intramedullary Steinmann pins) or external fixation associated or not to bone grafting.¹⁸

The most commonly used technique is the excision of the non-union, bone grafting with an internal fixation; but, what still makes debate is whether to use plates or pins.

Literature reports contradictory results for each technique with different complications: infection, implant failure, delayed or non-union, splintering of the bone and brachial plexus or vascular injury.^{1 4 5 18–21}

Chandran *et al*⁴ in a comparative study assure that plate fixation achieved more reliable and quicker union with fewer complications compared with pin fixation. Comparing the two techniques, Persiani *et al*¹⁴ found the opposite result and recommend internal fixation with K-wires. Studer *et al*¹⁵ report 57% of delayed bone union (>1 year) using plates, whereas Lorente *et al*¹⁶ report no complication using K-wires in six cases.

Figure 7 Patient's cosmetic appearance.

Learning points

- Congenital pseudarthrosis of the clavicle (CPC) is a rare congenital pathology whose treatment is still debated.
- Surgery is indicated for cosmetic issues and functional impairment.
- The Masquelet technique allows early and lasting bone union in the treatment of CPC especially in high-risk older patients with long gap.

We agree with Persiani *et al* who assert that based on their experience and the literature review, the most effective surgical procedure involves resection of the pseudarthrosis, iliac bone grafting and internal fixation with K-wires.

Even if it seems that CPC remains an easy condition to be treated, it should be noted that the non-union remains the most common and most serious complication. Persiani *et al* report 30% of non-union and Di Gennaro *et al* 26%,¹⁷ whereas Studer *et al*¹⁵ find 57% of delayed bone union (>1 year).

In our 14-year-old boy with CPC, we have judged that the risk of non-union was high because of his age and the important gap after resection of the atrophic bone (3.3 cm representing 19% of clavicle total length).

To further bone union and avert complications, we used the induced membrane technique described by Masquelet *et al*⁶ who have recently shown its effectiveness in the treatment of congenital pseudarthrosis of the tibia in children.^{8–20} The two-step technique allowing during the interval the revascularisation of marginally viable tissue surrounding the bone defect and formation of the self-induced periosteal membrane²² secreting growth factors decreases the risk of non-union.

Today, the Masquelet technique has become the reference treatment method to respond to the main situations of bone loss encountered in paediatric orthopaedics.²²

Complications of the induced membrane technique are rare and represented by pseudarthrosis and graft resorption. To avoid these complications, the method requires technical rigour on two fronts: cement coverage of the extremities and assembly stability.^{8–22} The disadvantage of using a two-step technique and removing the material as a third operating time can be avoided using local anaesthesia as what we have done. This early removal of the K-wire becomes an advantage compared with the period plates that must be left in place and the risk of infection that it implies.

Only one case of CPC treated by the Masquelet technique has been reported by Gouron *et al*² who assert that this method avoids the disadvantages of plate fixation and can be used in case of surgical revision after massive lysis of the bone graft.

It is also important to note that many authors report a duration to achieve radiological union from 3 months to 25 months.^{4–5–14–15–17–21–24} In our case, the use of the induced membrane technique seems to shorten the time of radiological union (10 weeks) compared with other techniques^{4–5–14–15–17–21–24} as it is the case when using this technique in the other aetiologies of bone loss,²² but further studies must confirm this postulate.

In our opinion, the Masquelet technique allows early and lasting bone union in the treatment of CPC especially in high-risk older patients with long gap.

Contributors HA conceived the study, collected and analysed the data and participated in surgery and follow-up. MAA operated and followed up the patient and contributed to the analysis and interpretation of the case report and also to the concept of the study. LC and KA contributed to the refinement of the study and approved the final manuscript.

Competing interests None declared.

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