Isolated congenital pseudarthrosis of the fibula: A report of two cases and review of the literature

T. Cherrad^{*}, H. Zejjari, H. Bousbaä, J. Louaste, K. Rachid, L. Amhajji

^a Department of Orthopedic Surgery and Traumatology, Military Hospital Moulay Ismail Meknes, HMMI, BP 50000, Morocco

ARTICLE INFO

Article history:

Received 26 November 2016

Accepted 3 July 2018

Available online 5 July 2018

Keywords:

Congenital pseudarthrosis

Fibula

Valgus

1. Introduction

Pseudarthrosis of the fibula is frequently associated with a pseudarthrosis of the tibia, but it becomes uncommon when it's isolated. Isolated congenital pseudoarthrosis of the fibula (ICPF) is usually considered a less severe condition than congenital pseudoarthrosis of the tibia (CPT). However, its site most frequently near the ankle leads to severe valgus and instability of this weight-bearing joint.¹

Due to its bad functional results such as deformity, instability and severe osteoarthritis; many of operative techniques are proposed. They are intended to correct either the deformation, either the pathogenic mechanisms, or both at the same time.

Through two observations and a review of the littérature we discuss the treatment of this rare entity.

2. Case 1

A 13 year old boy complained for pain on walking with limping and progressive deformation of the right ankle. Examination showed valgus ankle deformity with medial malleolus protrusion. No family history of neurofibromatosis was noted. Spine examination was normal, and no Café-au-lait spots were observed.

Ankle X-ray images showed congenital pseudarthrosis at the

lower third of the fibula with no tibial deformity; concisely it was Dooley type 3 with 78° in lateral distal tibial angle (LDTA) (Fig. 1).

The patient underwent osteosynthesis of the fibula by screwed plate and intercalary bone graft associated with internal screw epiphysiodesis of distal tibial cartilage. Postoperatively, the patient was immobilized in a leg cast for 6 weeks and then weight bearing was allowed with an ankle–foot orthosis. The evolution after two years is favorable with disappearance of clinical signs and 90° in LDTA (Fig. 2).

3. Case 2

A 12 year old boy, without medical history, was referred to our department for a deformation of the right ankle with pain and limping. Examination showed a normal spine, and no Café-au-lait spots. A severe valgus deformity of the ankle was present on standing. Plain radiographs of the ankle (Fig. 3a and b) showed ICPF type 3 of Dooley with 59° in LDTA.

The patient underwent frontal osteotomy of the tibia to improve LDTA and distal tibiofibular synostosis, also an epiphysiodesis of the medial malleolus was realized.

Postoperatively, the patient was immobilized in a leg cast for 2 months and then weight bearing was allowed with an ankle–foot

^{*} Corresponding author.

E-mail address: taoufikcherrad@gmail.com (T. Cherrad).

Fig. 1. ICPF type 3 of Dooley with 78° at LDTA (case 1).

orthosis. The outcome was favorable after 48 months with clinical and radiological improvement (LDTA 95°) (Fig. 3c).

4. Discussion

ICPF is a very uncommon condition with fibular's continuity deficiency, which often sits at the distal third or just proximal to the ankle joint.²

Dooley et al.³ described a 4-part classification for congenital pseudarthrosis of the fibula (CPF), but only types 2 and 3 are ICPF:

- Type 1: prepseudoarthrotic fibular bowing with ankle varus and anterolateral tibial bowing.
- Type 2: ICPF with neutral ankle alignment.
- Type 3: ICPF with ankle valgus.

- Type 4: CPF with late development of congenital pseudarthrosis of the tibia.

As the fibula bears only 7–15% of the weight across the ankle, pseudarthrosis may be clinically asymptomatic. Thus, level (the more distal, the more likely the development of deformity) and type of pseudarthrosis (gap, relation between bone ends and quality of bone ends) probably influence the development of valgus deformity.⁴ Dias⁵ has also noted a close relation between the fibula shortening and progressive valgus deformity of the ankle.

Although the ICPF may be asymptomatic, the progressive valgus deformity of the ankle (Dooley type 3) causes limping and pain at walking. This has been seen in the Dooley type 3; however, we have not documented progressive valgus in a Dooley type 2 deformity. Patients with ICPF may have radiological changes in the tibia, even pathological fracture due the lack of lateral stability of the ankle.²

Treatment is recommended only in case of deformity of the ankle (type 3), for type 2 surveillance is required.⁶ The surgery aims to correct the deformation directly (valgus) or its mechanisms, or both at the same time.⁴

Procedures that aim to realignment of the ankle by corrective supramalleolar osteotomy of the tibia should be used for patients older than 10 years, and must generally be combined with other therapeutic methods to prevent recurrence, as the case of our second patient.⁶

To correct the deformity, various procedures have been proposed for each pathogenic mechanism (pseudarthrotic tissue, lack of lateral stability of the ankle, shortening of the fibula and abnormal load transmitted to the ankle). Various therapeutic methods can be combined to improve results.⁴

The pseudarthrotic tissue accelerated the deformity by effect of attachment, forming a kind of a cord that aggravates the valgus. This abnormal tissue is also responsible of resorption of the bone graft.⁷ Consequently the first step of treatment is the resection of the pseudarthrotic tissue.

The lateral ankle support provided by the fibula is lost in the ICPF, which causes valgus and instability at walking. Langenskiöld has recognized that the distal tibiofibular synostosis stabilizes the ankle and prevents valgus progression. Bone grafting is often necessary in this synostosis even if there is a risk of resorption. A syndesmotic screw, like we have done for our second patient, may

Fig. 2. Result after 2 years with 90° at LDTA (case 1).

Fig. 3. Case 2;
- 3a and 3b: ICPF type 3 of Dooley with 59° at LDTA.
- 3c: result after 4 years with 95° at LDTA.

be used to provide primary stability.⁸

The shortening of the fibula is mainly caused by the absence of the downward thrust provided by the development of the proximal growth plate of the fibula and secondarily by the reduced stimulation of the distal physis. To enable transmission of push, fibular continuity restitution procedures have been proposed. However these surgical techniques are generally complex situations, requiring several operations with a real risk of failure (resorption of the graft, pseudarthrosis recurrence). The most used techniques are autogenous bone grafting,^{6,9} the allograft,¹⁰ methods using transfer the bone by Ilizarov technique,⁶ and the technique using a distal pedicled returned periosteal flap and sutured at the pseudarthrosis site.¹¹

Fibular osteosynthesis is indicated when the ankle alignment is neutral, the distal fibular fragment is wide and long enough to be internally fixed and finally when the gap of the pseudarthrosis can be approximated or filled with bone graft at the time of internal fixation. Our first patient met all the conditions and was treated by fibular osteosynthesis.⁹

Another worst factor of the deformity is uneven load transmission to the distal tibia. The increased compressive stress in the latero-distal tibia delays growth of this region, and in contrast provides accelerated growth at the medial portion.⁵ The result is progressive lateral wedging of the epiphysis and growth modifications of distal physis with aggravation of deformity; thus the internal hemi-epiphysiodesis is proposed as solution.¹⁰

In summary, isolated forms of the PCF require treatment for the deforming type. Different methods are proposed either isolated or associated considering the patient's age, the rate of evolution, the importance of nonunion, and radiographic features.

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

Consent

The patients and their parents are informed about our paper and we have their consent.

Statements

All authors state have not received grant support or research funding and they declare that there is no conflict of interest regarding the publication of this paper.

References

1. Tudisco C, Bollini G, Dungi P, et al. Functional results at the end of skeletal growth in 30 patients affected by congenital pseudarthrosis of the tibia. *J Pediatr Orthop Part B*. 2000;9:94–102.
2. Martus Jeffrey E, Johnston Charles E. Isolated Congenital Pseudarthrosis of the Fibula. A comparison of fibular osteosynthesis with distal tibiofibular synostosis. *J Pediatr Orthop*. 2008;28:825–830.
3. Dooley BJ, Menelaus MB, Paterson DC. Congenital Pseudarthrosis and bowing of the fibula. *J. Bone Joint Surg. Br.* 1974;56:739–743.
4. Lampasi Manuele, Antonioli Diego, Gennaro Giovanni LuigiDi, Magnani Marina, Donzelli Onofrio. Congenital pseudarthrosis of the fibula and valgus deformity of the ankle in young children. *J Pediatr Orthop B*. 2008;17:315–321.
5. Dias LS. Valgus deformity of the ankle joint: pathogenesis of fibular shortening. *J Pediatr Orthop*. 1985;5:176–180.
6. Ng BKW, Saleh M. Fibula pseudarthrosis revisited treatment with Ilizarov apparatus: case report and review of the literature. *J Pediatr Orthop B*. 2001;10:234Y237.
7. Dal Monte A, Donzelli O, Sudanese A, et al. Congenital pseudarthrosis of the fibula. *J Pediatr Orthop*. 1987;7:14–18.
8. Langenskiöld A. Pseudarthrosis of the fibula and progressive valgus deformity of the ankle in children: treatment by fusion of the distal tibial and fibular metaphyses: review of three cases. *J. Bone Joint Surg. Am.* 1967;49:463–470.
9. Cho TJ, Choi IH, Chung CY, et al. Isolated congenital pseudarthrosis of the fibula: clinical course and optimal treatment. *J Pediatr Orthop*. 2006;26:449–454.
10. DiGiovanni CW, Ehrlich MG. Treatment of congenital pseudarthrosis of the fibula with interposition allograft. *Orthopedics*. 1998;21:1225–1228.
11. Trigui Moez, de Billy Benoit, Metaizeau Jean-Paul, Clavertd Jean-Michel. Treatment of congenital pseudarthrosis of the fibula by periosteal flap. *J Pediatr Orthop B*. 2010;19:473–478.