Case Report

Congenital Pseudoarthrosis of Medial Malleolus in A Young Soccer Player - Diagnosis in Clinical setting of Ankle Sprain

Giuliano Cerulli¹, Fantasia Fabiano², Potalivo Gabriele¹, Placella Giacomo³, Sebastani Enrico³

Abstract

Introduction: We report a case of a young female soccer player affected by congenital medial bilateral malleolus pseudoarthrosis and os subfibulare. Congenital pseudoarthrosis is the failure of the bones to fuse prior or at birth. The etiology is still unknown, although frequency is high in subjects affected by neurofibromatosis or correlated syndromes, so it has been suggested that these congenital disorders may be the cause of congenital pseudoarthrosis.

Case Report: Our patient, a 16-year-old female, high level soccer player, was referred to us following a right ankle sprain during a match. She reported no medical history of tibia-tarsus joint injuries or disease. Pain, swelling and functional impairment were noted immediately after the accident. Standard radiographs in the emergency department revealed a displaced fracture of the medial malleolus and the presence of os subfibularis. The patient was transferred to our Traumatology and Orthopaedic Department to undergo malleolus osteosynthesis. Before surgery swelling, functional impairment and intense pain at the medial malleolus level were confirmed. However, there was no radiological opening of ankle, instability or pronation pain; furthermore the flexion-extension was preserved with slight pain. Twenty-four hours later a considerable remission of symptoms was evident with increased range of motion and reduction in the swelling and post-traumatic edema. A radiograph on the left ankle to compare with that of the right ankle was necessary to overcome the discrepancy between the radiological diagnosis and the clinical examination. The radiographic results of both medial malleoli were comparable although on the left the os subfibularis was absent. Since the diagnosis of fracture by the association between the radiographs and the symptomatology was doubtful, a bilateral CT was performed. The scan revealed a medial bilateral malleolus pseudoarthrosis and an accessory right subfibularis nucleus. The patient was discharged from hospital with the diagnosis of “second degree right ankle sprain in patient affected by congenital medial bilateral malleolus pseudoarthrosis”. A therapeutic-rehabilitative program was prescribed for the ankle sprain and unnecessary surgery was avoided. After 30 days there was an almost complete remission of pain. At a follow-up of six months the patient was completely asymptomatic and gradually began competitive activity.

Conclusion: An accurate history and an objective examination should be performed and correlated with the results of diagnostic procedures in order to avoid the incorrect diagnosis of a fracture needing surgery. The rarity of this ailment and the absence of consequences on long-term function, show that this disease does not justify sports activity cessation. Traumatic events at this site must be assessed properly in order to avoid being confused with malleolus fractures leading to over treatment.

Keywords: Ankle sprain, medial malleolus, pseudoarthrosis

What to Learn from this Article?

Diagnosis of congenital pseudoarthrosis of medial malleolus specially in a confusing setting of ankle sprain

Author’s Photo Gallery

¹Nicola’s Foundation Onlus, Arezzo, Italy.
²Nuova Clinica San Francesco, Foggia, Italy.
³University of Perugia, Italy.

Address of Correspondence
Prof. Giuliano Cerulli,
Orthopaedic and Traumatology Residency Program,
University of Perugia, via GB Pontani 9, Perugia, Italy.
Email: clinreslpm@gmail.com
Introduction

Congenital pseudoarthrosis is the failure of the bones to fuse prior or at birth. [1] The etiology is still unknown, although frequency is high in subjects affected by neurofibromatosis or correlated syndromes, so it has been suggested that these congenital disorders may be the cause of congenital pseudoarthrosis [2]. The most frequent site is the distal half tibia, with possible involvement of the ipsilateral fibula at the same level [3]. Nevertheless, the expression “congenital tibial pseudoarthrosis” includes many diseases including both simple tibial anterior “prucurvatus” and the severe form involving both skeletal segments of the leg and pseudoarthrosis due to the failure of the growing nucleus to fuse. This rare disease has an incidence of 1 in 250,000 individuals. The correlation with neurofibromatosis is 50-90%. [4] The etiology seems to be due to the lack of, or an insufficient ossification of the primordial growing nucleus. Biopsy has shown fibrous tissue with chondral islets and bone spurs within which electron microscopy has revealed fibroblasts inside a collagen fibrous matrix. [5] Mechanical factors, such as muscular imbalance together with altered electrochemical and electromechanical bone potential could be considered as possible causes of congenital pseudoarthrosis. [6]

Case Report

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The patient was transferred to our Traumatology and Orthopedic Department to undergo malleolus ostheosynthesis. Before surgery swelling, functional
impairment and intense pain at the medial malleolus level were confirmed. However, there was no ankle relaxation, instability or pronation pain; furthermore the flexion-extension was preserved with slight pain. Twenty-four hours later a considerable remission of symptoms was evident: increase of the R.O.M. and reduction in the swelling and the post-traumatic edema. A radiograph on the left ankle to compare with that of the right ankle was necessary to overcome the discrepancy between the instrumental diagnosis and the clinical examination. The radiographic results of both medial malleoli were comparable although on the left the os subfibularis was absent. (Fig 3).

Since the diagnosis of fracture by the association between the radiographs and the symptomatology was doubtful, a bilateral CT was performed. The scan revealed a medial bilateral malleolus pseudoarthrosis and an accessory right subfibularis nucleus (Fig. 4-5-6). The patient did not have any history of trauma in the past and also the bilateral nature of the disease forced us to think in terms of congenital etiology.

The patient was discharged from hospital with the diagnosis of “second degree right ankle sprain in patient affected by congenital medial bilateral malleolus pseudoarthrosis”. A therapeutic-rehabilitative program was prescribed for the ankle sprain and unnecessary surgery was avoided. After 30 days there was an almost complete remission of pain.

At a follow-up of six months the patient was completely asymptomatic and gradually began competitive activity.

**Discussion**

Any discrepancy between clinical diagnosis and radiological findings should be carefully evaluated.

The classification most commonly used to evaluate pseudarthrosis is that described by Boyd as it includes clinical examination, radiological signs and findings prognosis. Nevertheless this patient did not fit any type. The presence of os subfibularis lead the formation of os tibialis and secondary pseudarthrosis during developmental phase of the epiphysis. [7] The radiographs revealed a different morphology between
medial and lateral malleolus. Accessory growing nucleus generally have smooth and defined borders as were observed in the peroneal malleolus. On the contrary, the radiographs of the medial malleolus showed jagged borders and spurs, consistent with pseudarthrosis. [8,9] Heterotypical ossification can occur in any malleolus. It is not known if they are present as an anatomical variation or as the consequence of chronic microtrauma. The radiographs show them as two different entities, but they may represent one anatomical entity [10], which may undergo repeated stress during sports activities until avulsion occurs. [11] As shown with this patient, the chance finding should not induce the physician to prescribe competitive sport cessation. Generally os-accessory appears between the age of 7 and 10 years. Os tibial is present in 20% of healthy ankles, os fibular in 1%. Ogden [11], evaluating 103 patients, observed possible bilateral accessory ossification often diagnosed following an accident leading to ankle sprain. By evaluating the bone borders, a differential diagnosis between accessory nucleus and pseudarthrosis is possible. [12] Although uncommon, several cases of stress fractures in young athletes have been described in literature. [13] The Salter-Harris [14] and the Ogden classifications were consulted but they did not describe our case properly. The total absence of café-au-lait spots and neurofibromas excluded the presence of Neurofibromatosis type 1. The patient’s brother’s and mother’s radiographs did not show any pathological features. Unfortunately the father’s radiographs were unavailable. The absence of family correlations lead to the conclusion that this was probably just a sporadic case.

**Conclusion**

Congenital pseudoarthrosis of medial malleolus is very rare may be due to disturbance in epiphyseal development. These may be confused with traumatic fractures as seen in [1] our case and may lead to inadvertent surgery. Careful assessment of bone borders and clinical examination of instability will lead to accurate diagnosis. Conservative management will be enough in these cases and presence of pseudoarthrosis may not justify cessation of sports activity.

**Clinical Message**

In medicine many unusual situations may occur that can lead the physician to reach an incorrect diagnosis. This case-report shows the importance of sharing information and experiences.

**References**