

# Pseudodystrophy of the ankle and lower leg in a 9-year-old girl

Sascha VAN NUIJS, Jan BERGER, Marek WOJCIECHOWSKI, Isabelle D'HOORE, Marc DRIESSENS

From Antwerp University Hospital, Antwerp, Belgium

A 9-year-old girl presented with pain, swelling, redness and functional impairment of the left foot after a minor trauma. Clinical assessment revealed atrophy of the left calf and cyanosis and coldness of the left foot. Bone scintigraphy showed diffusely decreased tracer uptake in the left lower leg. Further examinations were normal. Pseudodystrophy was diagnosed and intensive physiotherapy was started. This resulted in complete functional recovery.

Pseudodystrophy is typically found in children, adolescents and young women. The clinical features usually include severe pain at a joint or part of a limb with major functional disability, cyanosis, coldness and oedema or atrophy. This is caused by disuse of the affected limb after a minor trauma.

The physical lesions may be improved or cured by means of intensive physiotherapy, sometimes combined with medication. As psychogenic factors often play an important role, one of the important elements of treatment is psychotherapy.

The most important differential diagnosis is reflex sympathetic dystrophy (RSD). The distinction can be made by bone scintigraphy.

**Keywords** : pseudodystrophy ; children ; arthralgia ; scintigraphy ; reflex sympathetic dystrophy.

## **INTRODUCTION**

Pseudodystrophy is a condition clinically resembling reflex sympathetic dystrophy (RSD). The distinction between both disorders is based on clinical features and bone scintigraphy. The mainstay of treatment is physiotherapy. Pseudodystrophy (PD) is a little known problem. There are no data regarding incidence or prevalence of the disorder. We describe a 9-year old girl with PD at the distal part of the left leg.

## CASE REPORT

A 9-year-old girl presented with pain and functional impairment of the left foot. The complaints started a few months earlier after distortion of her ankle. She was already treated with analgesics and physiotherapy, but the symptoms only worsened and she refused to use her foot. Walking was impossible without the aid of crutches.

Clinical assessment revealed marked atrophy of the left calf (circumference 22 cm on the left, com-

E-mail : marek.wojciechowski@uza.be

<sup>■</sup> Sascha Van Nuijs, MD, Resident.

<sup>■</sup> Marek Wojciechowski, MD, Senior Staff Member.

Department of Paediatrics, Antwerp University Hospital, Antwerp, Belgium.

<sup>■</sup> Jan Berger, MD, Resident.

<sup>■</sup> Marc Driessens, MD, PhD, Senior Staff Member.

Department of Physical Medicine and Rehabilitation, Antwerp University Hospital, Antwerp, Belgium.

<sup>■</sup> Isabelle D'Hoore, MD, Senior Staff Member.

Department of Nuclear Medicine, Imeldaziekenhuis, Bonheiden, Belgium.

Correspondence : Marek Wojciechowski, Department of Paediatrics, University Hospital of Antwerp, Wilrijkstraat 10, 2650 Edegem, Belgium.

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*Fig. 1.* — Decreased bone density over the left ankle and hind-foot.

pared to 25 cm on the right side). The left foot was cyanotic and felt colder than the right. Passive joint mobility was painful but not limited. Arterial pulsations were slightly weaker at the left foot. There were no other abnormalities at clinical examination. Paedopsychiatric assessment was normal.

Full laboratory investigation was normal, with no signs of inflammation. Radiography of the left foot and ankle showed osteoporosis (fig 1), echography and MRI were normal. A duplex examination was also normal. Bone scintigraphy showed decreased tracer uptake at the left leg and ankle (fig 2). Thus, pseudodystrophy was diagnosed.

Intensive physiotherapy was started by means of active and passive mobilization and gait rehabilitation with progressive weight bearing on the left foot. Four months later, the girl walked most of the time without crutches and only slight atrophy of the left calf persisted. Eventually, complete functional recovery was obtained.

#### DISCUSSION

RSD and PD share many similarities : both are frequently initiated by trauma and present with pain, functional disability, atrophy or oedema,



*Fig. 2.* — Bone scintigraphy showing diffuse decreased uptake in the left lower leg.

cyanosis and coldness at a joint or part of a limb. Nevertheless, they are different conditions. In stage 1 RSD there is an increase in regional blood flow whereas decreased blood flow is observed from the onset in PD. Inflammatory changes as found in stage 1 RSD are never observed in PD (1). In stage 2 RSD, there is an increasing stiffening of the affected joint, due to sclerosis and retraction of the ligaments and tendons. In PD, passive joint mobility remains normal. If there is a limited range of motion, it is caused by compulsive posture of muscle groups, as is indicated by the fact that joint mobility is normal under anaesthesia. PD typically occurs in children, adolescents and young female adults. RSD, on the other hand, is found principally in the adult population without gender preference.

Bone scintigraphy in PD shows normal or, most frequently, decreased tracer uptake. In adults with stage 1 and 2 RSD there is typically an increased uptake. This is not the case in children suspected of RSD where increased as well as normal or decreased uptake can be seen (2,3), although some of the cases reported as RSD may have been PD. When decreased radioisotope uptake is found on bone scintigraphy together with a compatible clinical picture, the diagnosis of PD should be considered, and replace what was earlier described as RSD with negative bone scan (1).

Little is known about the pathophysiology. Deregulation of the sympathetic nervous system is associated with RSD while PD is considered to be due to disuse. Psychogenic and social factors seem to play an important role in many cases of PD (1). Important efforts should therefore be made to detect and, if possible, to treat the psychological disorders and social problems.

The diagnosis of PD in our patient was based on the clinical symptoms and signs, the absence of joint limitation and inflammatory changes and the decreased tracer uptake on bone scintigraphy. In contrast to other case reports, paedopsychiatric assessment was completely normal. This could be a reason why good therapeutic results were obtained with ambulatory rehabilitation. Indeed, admission to a rehabilitation unit is frequently necessary to implement a full-time rehabilitation scheme and to withdraw the patient from her or his environment.

The mainstay of treatment is physiotherapy. Normal use of the affected limb must be encouraged by means of active and passive mobilization and gait rehabilitation with progressive weight bearing. If needed, physiotherapy can be complemented with vasodilating, neuroleptic or analgesic drugs; however, aggressive methods of treatment (such as sympathectomy) should be avoided.

With a correct diagnosis and comprehensive treatment plan, pseudodystrophy can be cured as in our patient. This is a very important message to be given to the patient.

# CONCLUSION

Pseudodystrophy is a little known problem in children that clinically resembles reflex sympathetic dystrophy. However, these two disorders are different and require a different management. The mainstay of treatment of pseudodystrophy is intensive physiotherapy, preferably in a specialized centre. Psychological or social problems are often present, but sometimes, as in our patient, no underlying psychosocial problems can be identified.

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