MORBUS ODELBERG-VAN NECK – A RARE ENTITY IN CHILDHOOD

Narcis Flavius Tepeneu[1,2]

Abstract

Morbus Odelberg-van Neck (van Neck disease, van Neck-Odelberg disease, ischiopubic "osteochondrosis", synchondrosis ischiopubic syndrome, assymetric ischiopubic syncondrosis syndrome, osteocondrosis of the ischiopubic syncondrosis) is a benign skeletal abnormality in children involving a hyperostosis of the ischiopubic syncondrosis seen on radiographs. Children complain of groin or buttocks pain and it's radiological features mimic osteomyelitis, fracture, tumors or posttraumatic osteolysis or even the normal skeletal development. This leads usually to unnecessary workup. It is often considered a diagnosis of exclusion, as laboratory values are usually normal and routine radiographic workup may be nonspecific.

The paper presents two cases of van Neck disease in which the correlation between the clinical, laboratory and imaging data enabled the diagnosis of Morbus Odelberg-van Neck.

Key words: Morbus Odelberg-van Neck, hip pain, children, ischiopubic "osteochondrosis", ischiopubic syncondrosis (IPS)

Introduction

The ischiopubic syncondrosis is a cartilaginous joint between the os pubis and os ischia. It is a temporary cartilaginous joint which is present at birth and undergoes complete ossification before puberty.

In 1923 Odelberg described 3 patients who had rarefying lesions of the ischium of doubtful etiology with pain in the hip, limping and limitation of hip movement [1]. All patients were operated and the histological investigation revealed a non-specific inflammation. In 1924 van Neck reported two similar patients upon whom he had operated and applied the term ischiopubic osteochondritis for the first time [2]. It was first considered a pathological fenomenon. Later, because of it's apparent benignity and spontaneous healing the terms of osteochondritis or osteochondrosis were used less and less. Nowadays, it seems to be clear, that widening of the ischiopubic junction is a process of normal skeletal growth, at least in asymptomatic individuals.

The development of the pubic bone begins in the fetus during the fifth or the sixth month, when the center of ossification is formed in the horizontal ramus at the edge of the obturator foramen. The ischium normally begins to ossify in the superior ramus in the fetus during the fifth month. As growth progresses, the cartilage between the ischial and the pubic rami is replaced by bone until fusion occurs. This area, called the "ischiopubic synchondrosis," (IPS) undergoes fusion relatively early in childhood, whereas in the acetabular region, synostosis does not occur until puberty.

In early childhood, enlargement of this synchondrosis is bilateral; however, in older children, it is commonly unilateral. Usually, the fusion of the ischial and pubic bones develops without any clinical symptoms. Recognition of the entity as a normal variant is important for radiologists when interpreting a pediatric pelvic radiograph[4,5]. One of the key questions to ascertain is wheather the region is painful or not.

Material and method

Two cases of Morbus Odelberg-van Neck are presented.

The *first case* is of a nine- year-old child from the rural area presenting with hip pain on the left side. The patient could walk, but his limp was obvious. He was a active football player, yet he denied traumatic events. From the patients history a insiduos appearance of the complaints is to be noted, the pain began about five weeks prior to presentation to the hospital. After sports the pain increased, so that the child developed a limp, which led to not being able to play football. The parents observed the limp but initially sought it came from a trauma in sports and it would go away. When the limp did not disappear after 2 weeks of observation at home they brought the child to the hospital. No medication was administered at home. There was no history of fever at home.

There was no palpable swelling of the IPS, the child complained about groin and hip pain, there was no limb length discrepancy, no local swelling or inflammatory signs, yet there was a contracture of the adductor muscles on the left side with restriction of external and internal rotation of the hip in comparison to the healthy right side.

Laboratory tests revealed a slight increase in the C-reactive protein (CRP) and the erythrocyte sedimentation rate (ESR). Procalcitonin and Anti-streptolysin-O (ASO) titer was normal. The blood culture did not reveal any pathogenic germs. The Mantoux tuberculinic test was normal. Anteroposterior radiographs of the pelvis showed an enlarged left ischiopubic syncondrosis characterized by a focal area of osteolysis (Figure 1). A diagnosis of van Neck disease was made.

[1]University of Medicine and Pharmacy ''Victor Babes'' Timisoara
[2]Emergency Children's Hospital ''Louis Turcanu'' Timisoara
E-mail: nftepeneu@yahoo.com

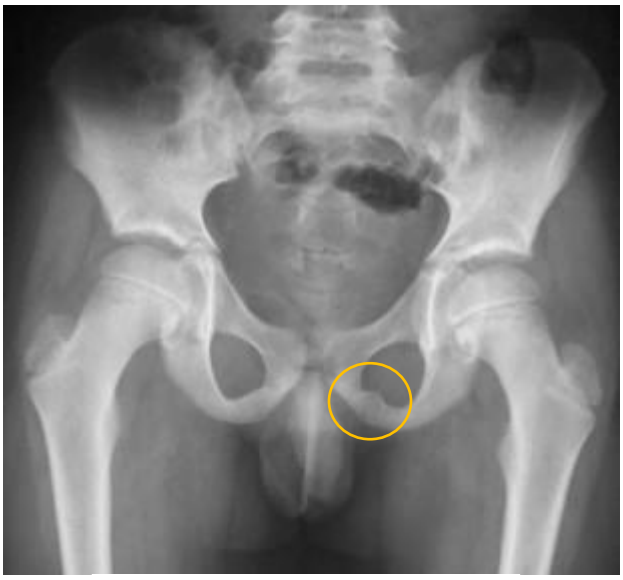

Figure 1. Radiography of the pelvis.

The child was treated anti-inflammatory therapy (NSAIDs) and bed rest for about 3 weeks. He was also prescribed crutches for ambulation and was advised to not be engaged in any sports. A clinical control of the patient 1 month later showed a remarkable improvement in symptoms, with an almost pain free patient. Laboratory tests revealed normal values. The ambulation with crutches was prescribed for a further month because of improvement of symptoms and the fact that the patient was very handy with them, despite of his relatively small age. At two month of treatment there was complete regression of symptoms. The patient was allowed to walk, but further sports interdiction was prescribed. A follow-up control at 4 months showed a virtually pain free patient, the laboratory tests revealed normal values so the patient was allowed to practice sports. He was eventually lost from our evidence, because the family moved to a foreign country.

The *second case* is of a 8 year old obese male patient presenting to the hospital with diffuse intermittent pain in the right groin region. The pain began about 3-4 weeks ago. The child was operated 6 years ago because of a undescended testes on the right side. 14 days before a foreign body (wooden piece) was removed by the family doctor from the scrotal skin. The mother of the child initially thougt it was scrotal pain, then a more detailed anamnesis revealed that the child was presumabily hit by another child with the foot in the pelvic region. There was no history of fever at home.

An ultrasound of the testis revealed no abnormalities. The ultrasound and radiographs of the pelvis and of the right hip (Figure 2,3) revealed also no abnormalities. Anteroposterior radiographs of the pelvis showed an enlarged bilateral ischiopubic syncondrosis, which was interpreted as the normal appearance of the ischiopubic syncondrosis.

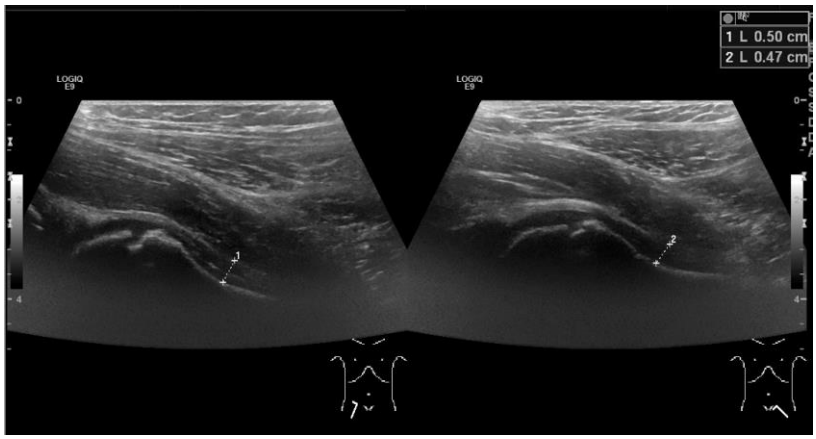

Figure 2 – Ultrasonography of the hips which shows no sign of intraarticular abnormalities.

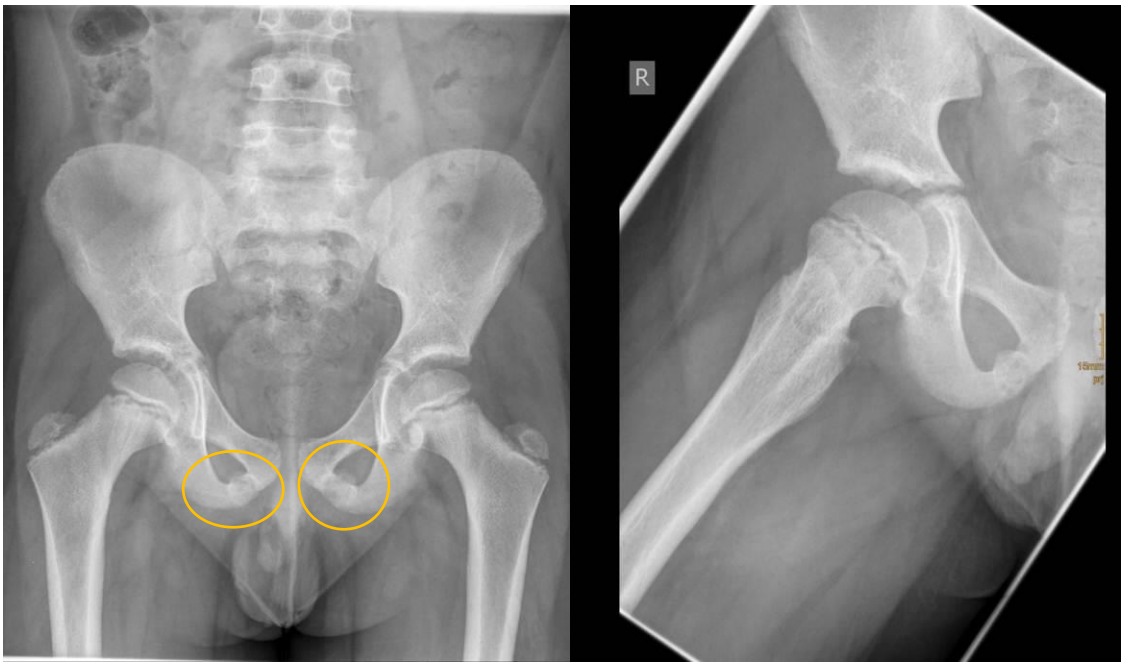

Figure 3 – Radiographs of the pelvis and the right hip.

Clinically both testes were descended, of normal shape and size. The was no sign of a inguinal hernia. The clinical examination revealed no limb length discrepancy, no local swelling or inflammatory signs, yet there was a contracture of the adductor muscles on the right side with restriction of external and internal rotation of the hip in comparison to the healthy left side. There was also a marked limp.

The child was initially treated with bed rest/ walking with with crutches and Ibuprofen p.o at home. There was no improvement of the pain, so a MRI of the pelvis was indicated.

The MRI images showed moderate oedema of the perilesional soft tissue and muscles and a fusiform enlargement of the right ischiopubic syncondrosis (Figure 4).

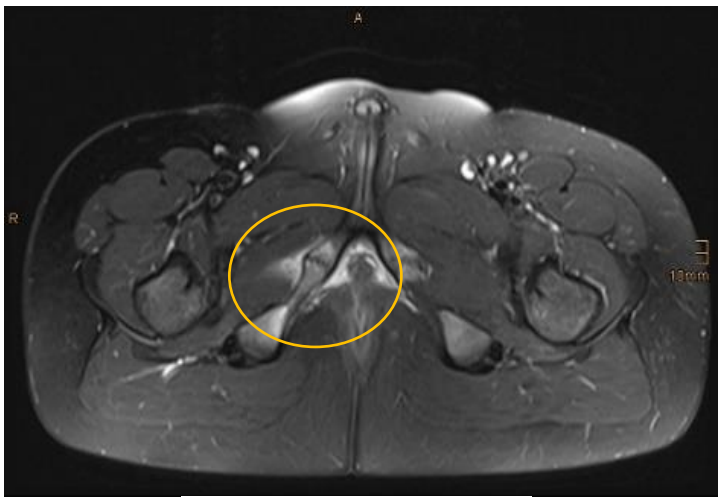

Figure 4 – MRI of the pelvis.

Laboratory values showed normal leucocites, ESR and a elevation of CRP of 1,5 mg/dl (normal value 0,5 mg/dl). Anti-streptolysin-O (ASO) titer was normal, also workup for rheumatoid diseases and Lyme disease. Under suspicion of a osteitis/osteomyelitis the pacient received a treatment with Clyndamicin 3x300 mg/day p.o for 3 weeks. The next week his CRP was in normal range and it continued to be with weekly followup. A follow-up radiography of the pelvis showed no dynamic of the image in comparison to Figure 4. Because the symptoms had a fluctuating character with

pain-free periods and then again impossibility to walk, a orthopaedic consultation was done. The pediatric orthopaedic surgeon recommened a bone scintigraphy. The bone scintigraphy showed no abnormalities. (Figure 5).

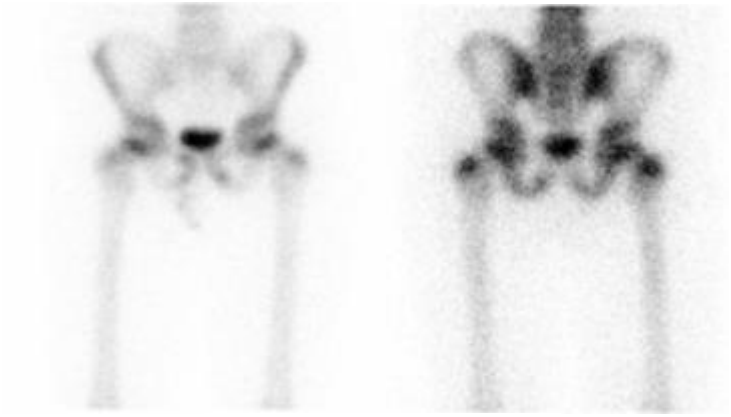

Figure 5 – Bone scintigraphy.

A neurologic exam by an pediatric neurologist was normal. Because the patient continued to have symptoms a biopsy under general anesthesia was proposed. The family refused.

As an alternative a five day i.v. therapy with Iloprost Trometamol (Ilomedin) 10 μg/0,5 ml was begun. The child was released in good condition from the hospital.

A follow-up MRI 6 weeks later showed that the local oedema was regredient and only discrete.

The soft tissues and muscles were normal. There were no other abnormalities (Figure 6).

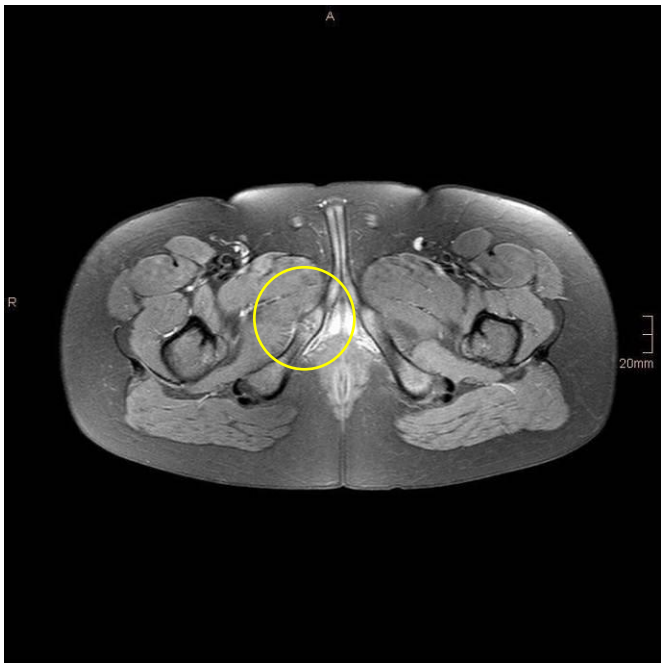

Figure 6 – Follow –up MRI of the pelvis.

Clinically the patient was also painfree and had a normal walking pattern .He was restrained from sports for another 4 weeks. At follow-up in 8 weeks the patient was painfree and had no complaints. He was again allowed to practice sports.

Discussion

In 1923 Odelberg [1] reported three patients (two boys aged eleven and fifteen years and a girl thirteen years) who had rarefying lesions of the ischium of doubtful etiology, with pain in the hip, a limp and limitation of movement. In each case the lesion in the ischium was scraped out. Bacteriological examinations for tuberculosis were negative, and histological investigation suggested non-specific inflammation. A fourth boy of eleven years who had been

operated on at another hospital was included in this report. In this child, and in the first of the other three children, a perineal fistula developed after an initial exploration. In retrospect it seems likely that these were cases of pain in the hip with altered ischio-pubic synchondroses. The descriptions of the findings at operation in these cases are very limited and the nature of the material submitted for histology was not recorded.

In 1924 Van Neck [2] reported two similar patients upon whom he had operated, and applied the term ischio-pubic osteochondritis for the first time. In one, a girl ofeight years, he found pain, limitation of hip movement, and a swollen labium majus. Radiographs showed the ischio-pubic synchondrosis to be enlarged and rarefied; and it was palpable per rectum. Suspecting osteomyelitis he incised the lesion but found only blood and friable bone, a small fragment of which was reported by one pathologist as a small-cell sarcoma, and by another as an osteochondritis. In the second child, a girl ofeleven, with similar clinical and radiological findings, operation revealed a hard bony swelling the size of a cherry; this was resected. Microscopically the cut surface of the swelling showed an irregular cartilage seeded with dark bony granules and islets of bone; no pus, sequestra or inflammatory tissue were present.

The histological report concluded that this was " osteochondritis, the bone and cartilage in the zone of endochondral bone formation showing inflammatory lesions of no specific character". Both patients recovered satisfactorily.

In 1956 Caffey and Ross [3] investigated the ischio-pubic synchondrosis in 549 radiographs of apparently normal children, with the object of estimating the time of closure of the synchondrosis and the incidence of swelling and uneven mineralisation. The ages of these children, of whom 48 % were boys, ranged from two to twelve years. They found that the fusion age was variable and extended from four to twelve years, but was commonest between nine and eleven years. Bilateral changes were more frequent than unilateral in a group of 246 children with swelling and uneven mineralisation; girls (134) were slightly more often affected than boys (112), and the greatest incidence was between five and ten years; 92 % of the changes occurred in this age range. They concluded that swelling of the ischio-pubic synchondrosis, with or without uneven mineralisation, is present at some time in almost all, or perhaps all, children.

Nowadays, it is well-known that the asymptomatic ischiopubic osteochondrosis is part of the normal fusion process. But when clinical symptoms are associated with these radiographic changes, must they be regarded as part of the normal growth process or as pathological?

Bernard et al [6] agree with the term "osteochondrosis" when clinical symptoms are associated with radiographic abnormalities and compare this entity with other osteochondroses such as Osgood-Schlatter's disease or Sinding-Larsen-Johansson's disease. Several authors [7,3] do not regard ischiopubic osteochondritis as a specific disease but suppose that the radiographic changes described are transitory stages in the normal fusion of the synchondrosis. Caffey and Ross found that more than 50% of asymptomatic children may present swelling and demineralization of the ischiopubic synchondrosis [3]. Neitzschman [8] considers this entity as part of the normal fusion process even though there is associated pain.

MRI was thought to be helpful in the differentiation of ischiopubic synchondrosis from other pathologic conditions because of its excellent tissue characterisation. However, most MRI findings in ischiopubic synchondrosis are non-specific and may add to the confusion concerning this physiological condition [9]. According to Herneth et al [10], typical MRI features of ischiopubic osteochondrosis involve signal alteration and contrast enhancement of the bone marrow, which is hyperintense on T2-weighted and STIR sequences and hypointense on T1-weighted sequences. Irregular swelling of the adjacent soft tissue is typically present and appears hyperintense on T2-weighted and STIR sequences. But only the fibrous "bridging" described by the same authors seems to be a characteristic MRI feature of the ischiopubic synchondrosis [8]. As described by Herneth et al, we also observed a band-like area in the center of the ischiopubic synchondrosis in the second case, which was hypointense on all sequences, consistent with a fibrous bridging. This finding, as well as the well defined margins of the ischiopubic bone on MRI, are reassuring to rule out a neoplastic lesion. The other MRI findings, however, were non specific.

Some differential diagnosis must be mentioned. Acute hematogenous ischiopubic osteomyelitis is rare. Symptoms include a limp, fever, pain with rotation of the hip. Laboratory tests show an elevated erythrocyte sedimentation rate, elevated C reactive protein and a positive blood culture.

MRI shows obvious myositis, abscess and free fluid surrounding the ischiopubic synchondrosis.

Stress fractures are common lesions in athletes and typically present with hyperintense bone marrow oedema on T2W images and a hypointense fracture line perpendicular to the long axis of the superior pubic ramus.

Tumors such as Ewing's sarcoma usually present with permeative bone distruction and extension into the soft tissue as well as neoplastic impregnation clinical symptoms.

Conclusions

Ischiopubic "osteochondrosis" is a well-known finding on conventional radiographs of both symptomatic and asymptomatic children. In this case report, the authors recall that the "atypical" radiologic appearance of the ischiopubic synchondrosis in children may be confused with other pathology, especially if it is discovered unilaterally. There is data that suggests that Morbus Odelberg-van Neck results from an excessive pull of the hamstring tendon on the ischial tuberosity – this pattern of oedema may suggest stress reaction and callus formation as a mechanism of ischiopubic hypertrophy[11].

MRI findings are strongly suggestive of oedema of the bone and adjacent soft tissue that may also be present in inflammation, tumour or trauma. Most MRI findings in ischiopubic synchondrosis are non-specific and may add to the confusion. Since MRI findings also seem to be non

specific, their interpretation warrants great care and a good knowledge of the physiological nature of ischiopubic synchondrosis.

Knowledge of this condition is essential to make sure it is not mistaken for stress fracture, infection or tumor in symptomatic children. "Cure" of the condition could be effected by bed rest alone or in association with anti-inflammatory drugs.

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Correspondence to:

Narcis Tepeneu
I. Nemoianu Street, no.2
Timisoara
Romania
E-mail: nftepeneu@yahoo.com