

Slipped capital femoral epiphysis as manifestation of a rare endocrinological disease

C.M. Beukhof^{1*}, F.C. van Biezen², W.W. de Herder^{1*}

Departments of ¹Internal Medicine, Sector of Endocrinology and ²Orthopedic Surgery, Erasmus Medical Center, Rotterdam, the Netherlands, *corresponding authors: +31 (0)10-70 40 704, e-mail: carolienbeukhof@gmail.com

CASE REPORT

Patient A presented at the age of 15 to the emergency department with acute hip complaints after a fall. At the age of six years, nodules were removed from her tongue. Examination revealed a flexion, abduction and exorotation contracture of the left hip with a decreased range of motion. Radiology confirmed a slipped capital femoral epiphysis (SCFE) of the left hip (*figure 1*). The SCFE was surgically treated with cannulated screw fixation. Peroperatively the blood pressure remained stable.

A few months later the patient was referred to an ENT specialist because of a nodule in the neck. Fine needle aspiration of the lymph node revealed medullary thyroid carcinoma. The patient had thickened lips, nodules on the tongue and a marfanoid appearance (*figure 2*).

Patient B visited an orthopaedic surgeon at the age of 16 because of pain of the left hip on exertion for the last six months. On examination left hip flexion was diminished; however, abduction and adduction were within the normal range. Radiology of the left hip showed that the capital

femoral epiphysis had slipped in a dorsomedial direction. Surgical treatment with a cannulated screw fixation was performed successfully.

At the age of 17, the patient returned with an SCFE of the contralateral hip. Radiology confirmed a slip of the right epiphysis of the femoral head. No complications occurred during screw fixation.

Preoperatively, a nodule in the thyroid had been noticed for which the patient was referred to a paediatrician. Thin needle biopsy showed no malignancy. Because of progressive growth of this tumour in the following years, which resulted in cosmetic complaints, the patient underwent a right hemi-thyroidectomy. Pathological examination surprisingly identified medullary thyroid carcinoma.

WHAT IS YOUR DIAGNOSIS?

See page 94 for the answer to this photo quiz.

Figure 1. A typical example of an acute slipped capital femoral epiphysis



Figure 2. a) A patient with neurofibromas of the tongue and eyelids, thick lips and marfanoid phenotype. b) Close-up of neurofibromas of the tongue (permission granted)



SLIPPED CAPITAL FEMORAL EPIPHYSIS AS MANIFESTATION OF A RARE
ENDOCRINOLOGICAL DISEASE

DIAGNOSIS

Slipped capital femoral epiphysis (SCFE) can be a manifestation of the multiple endocrine neoplasia syndrome type 2 (MEN 2).¹ MEN 2 syndrome is subdivided into MEN 2a and MEN 2b and both have medullary thyroid carcinoma as the most common feature. However, hyperparathyroidism is characteristic for MEN 2a whereas patients with MEN 2b can be recognised by neurofibromas of the tongue and marfanoid habitus.²

The diagnosis of acute SCFE is easier than that of chronic SCFE.³ Both disorders present with pain in the hip or with referred pain in the knee. Patients with acute SCFE typically have a contracture by flexion, abduction and external rotation. However, in patients with chronic SCFE the only presenting symptom can be a mild limp.³

SCFE can be difficult to diagnose on anteroposterior radiographs.³ For chronic SCFE a lateral radiograph according to Lauenstein (hips in 90° flexion and maximal abduction) is advised.⁴ SCFE often occurs bilaterally, therefore bilateral imaging at presentation and also during follow-up is indicated.⁴ Treatment of acute and chronic SCFE is surgical.⁴

Awareness of the association between MEN 2 and SCFE could help to identify patients earlier. This is crucial in order to prevent metastatic medullary thyroid carcinoma. Pheochromocytoma can also be part of MEN 2 and could cause severe hypertensive crisis or arrhythmias perioperatively. Complications of SCFE are avascular necrosis, chondrolysis and coxarthrosis if the diagnosis is missed.

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