Case Report

A 25-year-old woman was seen on Christmas eve at the emergency department because of a subacute pain in the left leg of increasing severity. Two years previously, during pregnancy, she had presented with bilateral avascular necrosis of the femoral head and was diagnosed with sickle cell β-thalassaemia. Before this, she had never experienced any haemolytic crises and was therefore unaware of this diagnosis. Because of severe functional impairment, the orthopaedic surgeon was planning bilateral femoral head replacement. Apart from the pain in her left leg her history at presentation was unremarkable. She came to the emergency department because she suspected a sickle cell crisis. At physical examination she did not appear ill. Her BMI was 19 kg/m². She had no fever. The left leg was shortened and positioned in endorotation. Laboratory tests were normal apart from known haemolysis. Because of the increased pain the orthopaedic surgeon was consulted who ordered an X-ray (figures 1 and 2). She was admitted to the internal medicine ward and treated with hyperhydration and adequate pain medication. Several days later she was discharged.

What is your diagnosis?

See page 295 for the answer to this photo quiz.

A woman with a painful hip

M. van der Valk*, M.H. Godfried

Department of Internal Medicine, Academic Medical Centre, University of Amsterdam, Meibergdreef 9, 1105 AZ Amsterdam, the Netherlands, *corresponding author: e-mail: m.vandervalk@amc.uva.nl

Photo Quiz

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Case Report

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Figure 1. Eight months prior to presentation

Figure 2. At presentation
The diagnosis is progression of bilateral avascular necrosis of the femoral head and advanced pregnancy. Unfortunately, the pregnancy was not seen by the orthopaedic surgeon, the resident internal medicine and the consulted resident radiology. Two weeks later she was seen again, this time because of a gastroenteritis. At physical examination she was found to be approximately 35 weeks pregnant. Because of deterioration of the foetal condition a caesarean section was performed the same evening without any complications. The birth weight of the baby was 2530 grams.

The patient was completely unaware of the pregnancy. Up until the last month she claimed to have had cyclic menstruation-like blood loss. Although the pathogenesis of this phenomenon is unknown, the prevalence of cyclic blood loss in pregnancy is estimated between 0.2 and 2.8%. Factors that might have contributed to the ‘denial’ of pregnancy apart from the cyclic blood loss can be the fact that in sickle cell disease 21% of neonates are small for the gestational age. Interestingly 70% of individuals presenting with denial of pregnancy report cyclic blood loss. In contrast to general belief, no differences in demographics and/or education level were found compared with a control group.

This case clearly demonstrates that one can very easily miss what one is not looking for.

REFERENCES