SMGroup

SM Journal of Neurology and Neuroscience

Clinical Perspective

Avascular Necrosis of Vertebral Body associated to its Sickle Cell Disease - A Rare Cause of Back Pain

Aaron Hilis[1]*, Girish Kulkarni[2], Ameet Kulkarni[1] and Qin Fang Goh[3]

[1]*Department of Neurosurgery, Queens Medical Centre, UK*
[2]*Department of Spinal Surgery, Queens Medical Centre, UK*
[3]*School of medicine, University of Nottingham, UK*

Article Information

Received date: Oct 22, 2018
Accepted date: Oct 25, 2018
Published date: Oct 26, 2018

***Corresponding author**

Aaron Hilis, Department of Spinal Surgery, Queens Medical Centre, Nottingham, UK,
Email: hilis@hotmail.com

Abstract

Avascular Necrosis (AVN) of a vertebral body or osteonecrosis is an uncommon entity. AVN has been described in the context of several underlying lesions; one of them is Sickle Cell Disease (SCD).

This is a case report of Avascular Necrosis (AVN) of a vertebral body in a young patient diagnosed with Sickle Cell Disease (SCD).

Introduction

Avascular necrosis of a vertebral body or osteonecrosis is an uncommon entity. Avascular necrosis has been described in the context of other underlying lesions, such as malignancy, infection, radiation therapy, systemic steroid treatment, and less commonly with vasculitides and trauma (Kummell's disease). (a) One of the disorders related to AVN is Sickle Cell Disease (SCD).

Sickle cell disease is a genetic disorder affecting both alleles of the haemoglobin beta gene. There is multisystem involvement in SCD, and red blood cells are sickled (banana-shaped) as well as sticky and rigid. These sickle cells can block blood flow in small blood vessels of the body. When blood flow is blocked in vessels that supply bone, the bone does not get enough oxygen and the bone tissue may die. There are many complications in SCD, AVN being the commonest complication affecting the epiphysis of long bones such as the femoral and humeral heads and femoral condyles. Osteonecrosis of vertebrae is a rare complication in sickle cell disease. Sickle cell trait, where only one haemoglobin beta gene allele is affected in a heterozygous individual, has also been reported to be a rare cause of AVN.

Sickle cell disease is commonly encountered in the Central and West African sub-region [1,2].

A Case

A young 13-year-old female presented to the spinal outpatient department with an 8 month history of lower back ache and pain while walking. She also complained of bilateral hip pain. The pain was mainly localized to the lower back with no radiation; there was no tingling or sensory complaint. The pain was worse with walking, hence restricting her movements and forcing her to use two crutches. She had some relief of pain only in supine position. There was no history of fever, weight loss, loss of appetite, white discharge pervagina, trauma to back or weakness of lower limbs.

On examination she was tender on spine palpation, paraspinal muscles spasm was present and there was no neurological deficit. There was pain on bilateral hip movement.

The patient is of Ghanaian descent and had recently immigrated to the UK. She was diagnosed with sickle cell disease as a neonate, and there was no evidence of history of previous blood transfusion. The diagnosis of homozygous sickle cell disease was confirmed on admission by performing hemoglobin electrophoresis, which showed a hemoglobin S percentage of 80% and hemoglobin F of 2.9%.

X-ray and MRI of the hip joint showed avascular necrosis of bilateral femoral heads.

Her hip pain was managed with IV analgesia. Despite the improvement of her hip pain, the patient continued complaining of lower back pain which prompted a lumbar spine X-ray. The X-ray showed bone softening and resultant compression of vertebrae by adjacent intervertebral disks (Figure 1). A slight dextro-scoliosis was noted (Figure 2), which was attributed to her on-going hip pain.

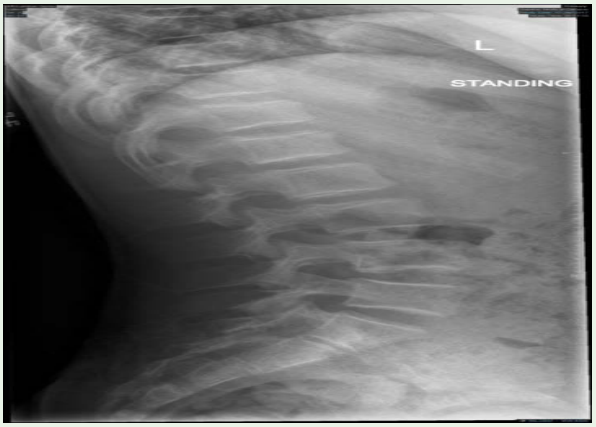

Figure 1: Lateral radiograph of the thoraco-lumbar spine shows the effects of bone softening and resultant compression of vertebrae by adjacent intervertebral disks (arrows).

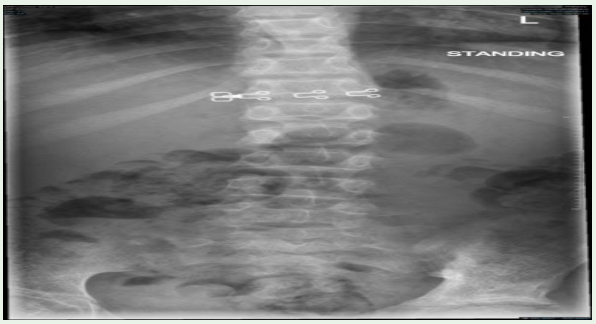

Figure 2: AP Radiograph thoraco-lumbar spine shows mild dextro-scoliosis, secondary to her right hip pathology.

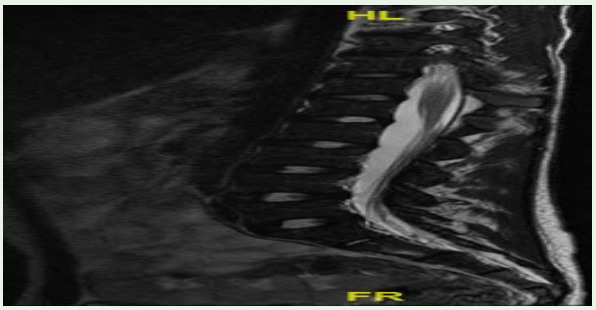

Figure 3: T2W MRI showing marrow infiltration and H shaped vertebra typical of SCD.

Magnetic resonance imaging with contrast revealed diffuse abnormal marrow signal intensity affecting L5, S1 and S2 vertebra, suggestive of osteonecrosis (Figures 3 & 4). There was bone marrow hyperplasia and multilevel H shaped vertebrae which are pathognomonic of sickle cell disease.

As per the treatment, the orthopaedic team provided her with supportive measures for her hip lesion.

The patient's back pain has remained quiescent and we have currently opted for a wait and watch policy.

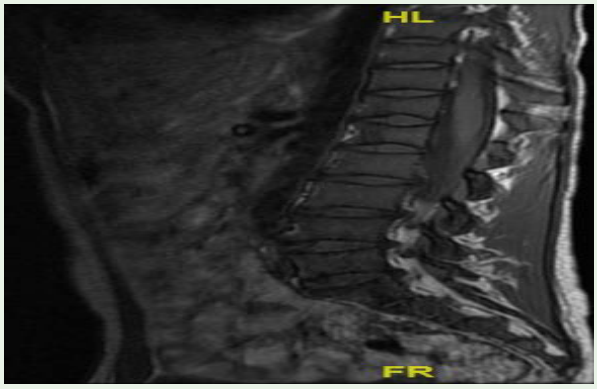

Figure 4: T1W MRI showing marrow infiltration at L5, S1 and S2, and H shaped vertebra.

Discussion

Spontaneous back pain is uncommon in teenagers. Usually there is a history of trauma or markers of inflammation in children suffering from lower back pain.

AVN of VB secondary to sickle cell disease is a rare cause of back pain. Bone changes in sickle cell disease occur due to marrow hyperplasia, tissue ischemia and infarction due to vaso-occlusion. Vertebral collapse can thus readily occur following weakening of the bone from thrombosis or infarction. Vertebral collapse is common following osteoporosis involving the vertebrae. The vertebral changes consist of central deterioration with preserved peripheral areas of the vertebral end-plates. This core destruction is a result of the anatomic blood supply to the end-plates. The central portion of the vertebral end-plate derives its blood supply from the long branches of the vertebral nutrient artery while the peripheral part of the end-plate is provided by short perforating branches of the periosteal vessels. The longer end arterial vessels are more likely to exhibit vaso-occlusion and destructive events [3].

Hip joint pathologies can mimic back pain and also can cause scoliosis in growing skeletons.

A high index of suspicion is necessary to make the correct diagnosis, and early management in these children is important to improve outcomes.

Management of sickle cell disease can be complex, involving plasmapheresis, bone marrow transplantation and even chemotherapy in extreme cases. As the disease progresses, it can cause further AVNs elsewhere in the spinal column and hence multimodal treatment is recommended involving discussion with the paediatric haematologists, parents and the surgical team.

References

1. Dean J, Schechter AN. Sickle cell anemia: Molecular and cellular bases of therapeutic approaches (first of three parts). N Engl J Med. 1978; 299: 752-763.
2. Ebong WW. The treatment of severely ill patients with sickle cell anemia and associated septic arthritis. Clin Orthop Relat Res. 1980; 149: 145-159.
3. Sadat-Ali M, Ammar A, Corea JR, Ibrahim AW. The spine in sickle cell disease. Int Orthop. 1994; 18: 154-156.

Citation: Hilis A, Kulkarni G, Kulkarni A and Goh QF. Avascular Necrosis of Vertebral Body associated to its Sickle Cell Disease - A Rare Cause of Back Pain. SM J Neurol Neurosci. 2018; 4(1): 1021s.