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"FISH VERTEBRA" about 3 Sickle Cell Patients Followed at Laquintinie Hospital, Douala

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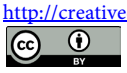

Abstract

Vertebral involvement in particular is common in sickle cell patients. We report 3 cases of "fish vertebra" fractures in sickle cell patients aged 16, 18, and 24 years old respectively at Laquintinie Hospital, Douala. When the vertebral fractures were diagnosed, the 3 patients had back pain and kyphosis deformities of the dorsal spine. Treatment with an infusion of biphosphonates (zoledronic acid at a dose of 0.5 mg·per·kg) was offered to all three patients. Two out of three patients received treatment with biphosphonates with a successful outcome. Profound vitamin D deficiency is associated with increased bone remodeling and a history of fractures. In sickle cell anemia, vertebral fractures may also result from bone fragility, which is often overlooked as aseptic osteonecrosis and osteomyelitis, which are very often suspected.

Keywords

Vertebral Fractures, Sickle Cell Anemia, Vertebral Bone Fragility

1. Introduction

Bone problems in sickle cell disease are multifactorial in origin. Vertebral involvement is particularly frequent and may result from vertebral osteomyelitis, stress fractures, vertebral vaso-occlusive crises or osteoporosis [1] 1n. Bone infarction is a debilitating and significant complication of SCD, and it may occur anywhere in the skeleton. It results directly from the sickling of red blood cells in

the bone marrow, which causes stasis of blood and sequestration of cells, Ischemia and tissue hypoxia are the consequences and, in turn, worsen the sickling process [2]. Medullary bone infarcts are far more common than osteomyelitis in patients with sickle cell disease but clinical differentiation can be difficult. [3] [4]

We report 3 cases of "fish vertebra" fractures in sickle cell patients aged 16, 18 and 24 years, followed at Laquintinie Hospital, Douala.

2. Observation

Case 1: A 24-year-old female patient, known to have SS homozygous sickle cell disease since the age of 3 years, on folic acid and hydroxyurea for 2 years, was admitted to hospital for disabling low back pain lateralized to the right with kyphoscoliosis and disabling right scapulalgia with severe anemia at 5.5 g/dl hemoglobin in a febrile context. Biological tests revealed a biological inflammatory syndrome with a predominantly neutrophilic leukocytosis of 23,000/mm^3, and an elevated CRP of 48 mg. The thick blood smear revealed a parasite density of 880 trophozoites/µl for *Plasmodium falciparum*. *Enterobacter cloacae* was isolated in both urine and blood cultures. The GeneXpert MTB/RF PCR on sputum was negative. The chest X-ray revealed a right basal alveolar consolidation. The X-ray and ultrasound of the right hip were normal. The CT scan of the lumbar spine showed an L5 fish vertebra, but the subsequent MRI revealed a biconcave compression of L5 but also of L4. She received a blood transfusion, effective antibiotic therapy, and morphine-based analgesics, 2 infusions of zoledromic acid A month apart together with calcium and vitamin D supplementation. A rigid corset was administered, and physiotherapy started in the hospital was continued at home, enabling the patient to stand up. She was then evacuated to a reference hospital in India for further treatment. She benefited from a total right hip prosthesis, which unfortunately became complicated by an infection. The prosthesis has now been removed and the patient is waiting for a replacement. She is still unable to stand upright. (**Figure 1**, **Figure 2**)

Case 2: An 18-year-old patient with SS homozygous sickle cell anemia known since the age of 4, on folic acid, was brought to the clinic with moderate back pain of 6 months' duration of mechanical origin, with kyphosis and without gibbosity. He presented with severe anemia at 6.7 g/dl, well tolerated with a good general state and in the absence of fever. Biological tests revealed a biological inflammatory syndrome with a predominantly neutrophilic leukocytosis of 12,000/mm^3 and an elevated CRP of 89.7 mg. GeneXpert MTB/RF PCR on sputum was negative, and the chest X-ray was normal. He had hypocalcemia of 86.7 mg/l and vitamin D level of 35.2 mg/l. MRI of the lumbar spine was suggestive of L2-L3 spondylodiscitis with paravertebral extension and tiered vertebral compression with a probably ischemic fish-like appearance complicated by L3 L4 and L4 L5 vertebral blocks. He received antituberculous treatment for 9 months. In terms of bone, an infusion of Zoledronic Acid was prescribed, along with calcium and vitamin D supplementation, and a rigid corset was administered.

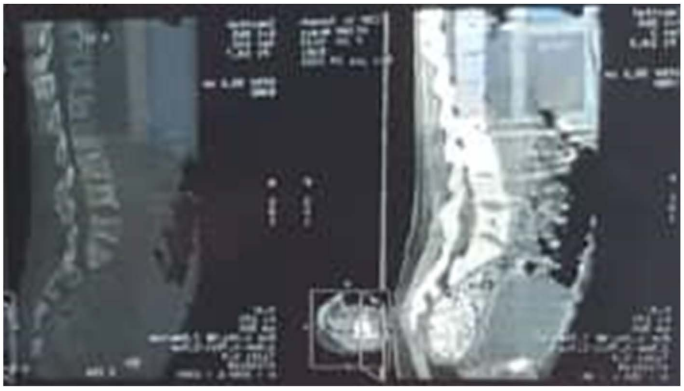

Figure 1. Case 1, CT scan of lumbar spine: biconcave compression of L5 with a probably ischemic fish-like appearance.

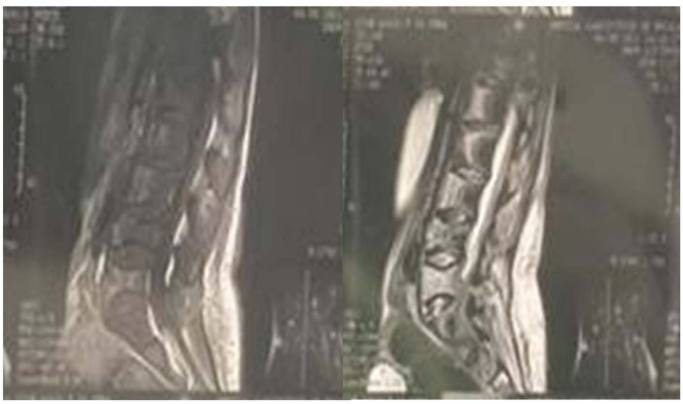

Figure 2. Case 1, MRI of lumbar spine: fish-shaped compression of the L4 and L5 vertebrae.

unfortunately, the patient did not receive the biphospnate due to lack of funds. A treatment with hydroxy urea was prescribed. (**Figure 3**)

Case 3: 16-year-old female with SS homozygous sickle cell anemia known since the age of 5 and on folic acid, referred for consultation for dorsolumbar kyphosis noted by the mother 1 year previously, with no notion of trauma associated with delayed puberty in a context of preserved general condition and apyrexia. The inflammatory work-up was normal and the hemoglobin level was 10.5 g/dl. A CT scan of the lumbar spine showed a dorsolumbar kyphosis of 53˚ at the D11 vertex against a background of benign vertebral compression and diffuse osteopenia. Calcemia was 96.5 mg/l. Vitamin D levels and biphosphonates treatment were prescribed, but the patient was not seen again. (**Figure 4**)

3. Discussion

The consecutive vertebral central depression seen in sickle cell anemia vertebrae is known as "fish-mouth" vertebrae due to the similarity of the upper vertebral inferior endplate together with the lower vertebra superior endplate and a fish with opened mouth [1]. It is thought to be secondary to subchondral infarctions of the central endplates and disk compression of the infarcted bone, leading to the

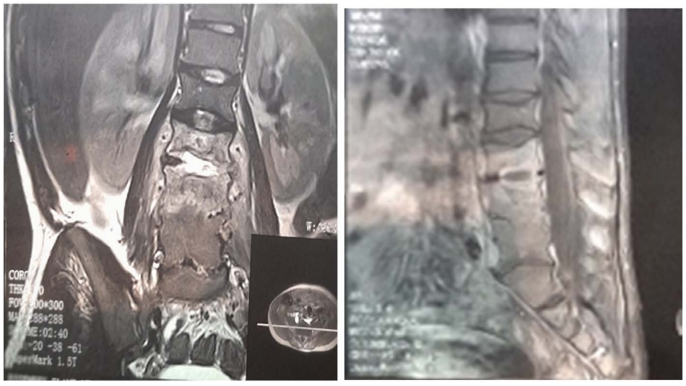

Figure 3. Case 2, MRI of lumbar spine: L2-L3 spondylodiscitis with paravertebral extension and tiered vertebral compression with a probably ischemic fish-like appearance complicated by L3L4 and L4L5 vertebral blocks.

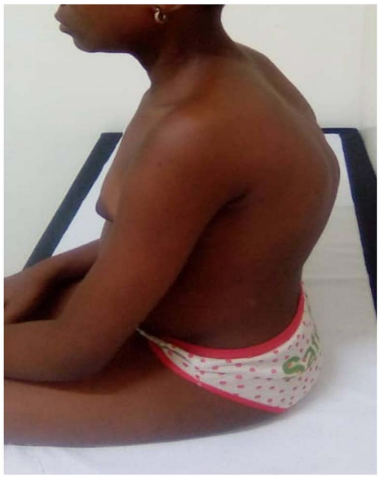

Figure 4. Case 3, Painless dorsolumbar Kyphosis in a 16-year-old female patient with SS homozygous sickle cell disease.

deformity and arching of endplates. The peripheral portions of the endplates are usually spared because of collateral circulation [2] [3]. The etiological diagnosis of "fish vertebrae" is not easy. In our 3 cases, the delay to diagnosis was 33 days or more. The causes are often interlinked, and vertebral infarction is sometimes concomitant with spondylodiscitis in the context of bone fragility, which is generally underestimated [4]. **Access to diagnosis is limited in our context:** Acute painful vaso-occclusive crisis occurs in the lumbosacral region in 2/3 of sickle cell patients. Its symptoms are dominated by hyperalgesia and disabling low back pain. In terms of imaging associated with vaso-occlusive crises, radioisotope bone scans using combined Sulphur labelled with colloid Tc-0m and Tc-99 diphosphonate is the gold standard for detecting areas of infarction in the acute phase. In practice, magnetic resonance imaging is sufficient and appears to be more sensitive. X-rays are not useful in the acute phase; however, it has been reported that the radiographic signs of bone infarction, including the "fish verte-

bra", appear as a biconcave deformation of the lumbar vertebrae with softening of the bone in the lateral parts in the late stage [5]. 2 of our 3 patients had access to MRI (Table 1) which enabled us to detect the fish vertebrea early. In Case 1 in particular, MRI enabled early detection of L4 involvement, which had not yet been visible on the CT scan performed a few days before.

Osteoporosis in sickle cell disease: myth or reality?

Osteoporosis is a generalized skeletal disorder characterized by low bone mass and deterioration in the microarchitecture of bone tissue, leading to bone fragility and increased susceptibility to fractures. According to the literature, more than 70% of adults with sickle cell disease have low bone mineral density (Table 2) and vitamin D deficiency. The associated factors found are disease severity, severe anaemia, low BMI and the SS homozygous phenotype [6] [7] [8]. In our context, access to vitamin D testing and bone densitometry is limited. It would be interesting to combine the treatment of sickle cell disease with systematic vitamin D supplementation.

Only one of our 3 patients received biphosphonates. The second was unable to obtain them due to lack of funds, and the third was lost to follow-up. In our

Table 1. Bone mineral density in patients with sickle cell disease.

Authors, place and year of study, sample size	Frequency of bone fragility (osteopenia and/or osteoporosis)	Factors associated with bone fragility
Gabriel bandzanni and al, Brésil, 2011, n = 65	81.5% (57 % ostéopénia and 24.5% ostéoporosis)	Low GFR, severe anaemia with p = 0.02, low BMI with p = 0.17
Mona Sarrai and al, 2007, NY, USA, n = 103	79.6%	Homozygous phenotype with p = 0.018; Severe anaemia with p < 0.0001, High ferritinaemia with p = 0.010, Low BMI with p = 0.003
Redonda G Miller and al, JH, 2006, USA, n = 32	72%	BMI with p = 0.007, sex M with p = 0.02, homozygous phenotype SS

Table 2. Patient adherence to treatment.

Age and exe	Visual Analogue Scale	Vitamin D	Calcema	Vitamin D supplementation	Biphosphonate treatment	Imaging for fish vertebra diagnosis
Case N˚1, 24 years old, Female	10/10, debilitating pain	Done	done	done	Done	MRI and CT scan of the lumbar spine
Cas N˚2, 18 years old, Male	5/10,	Not Done	Done	Done	Not Done	X-ray and MRI of the dorsolumbar spine
Cas N˚3 16 years old, Female	1/10	Not done	Done	lost from sight	lost from sight	CT scan of the lulbar spine

context, the lack of health coverage is a real obstacle to the care of our patients. However, complications can be disabling and irreversible, as in the case of patient number 1. An emphasis should be placed on the early detection of ischemic bone complications in sickle cell patients.

4. Conclusion

In sickle cell anemia, vertebral fractures may also result from bone fragility, which is often overlooked in favor of aseptic osteonecrosis and osteomyelitis, which are very often suspected.

Conflicts of Interest

The authors have no conflicts of interest.

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