Atypical presentation of spondylitis in a case with sickle cell disease

İlker Devrim[1], Ateş Kara[1], Güler Kanra[1], Muharrem Yazıcı[2], Şafak Güçer[3]
Fatma Gümrük[4], A. Bülent Cengiz[1], Gülten Seçmeer[1]

Units of [1]Infectious Diseases, [3]Pathology, and [4]Hematology, Department of Pediatrics, and [2]Department of Orthopedics and Traumatology, Hacettepe University Faculty of Medicine, Ankara, Turkey

SUMMARY: Devrim I, Kara A, Kanra G, Yazıcı M, Güçer S, Gümrük F, Cengiz AB, Seçmeer G. Atypical presentation of spondylitis in a case with sickle cell disease. Turk J Pediatr 2005; 47: 369-372.

Salmonella osteomyelitis is rarely seen and vertebral involvement by this organism is very rare, especially in childhood. Salmonella should be considered in the list of etiologic agents of osteomyelitis in sickle cell disease. Herein we report a five-year-old boy with sickle cell disease and *Salmonella paratyphi B* spondylitis who was diagnosed with atypical clinical findings of infection. We would like to emphasize the importance of differentiation between osteomyelitis and infarction even if there is no obvious sign of infection in the sickle cell patient even at atypical localization.

Key words: osteomyelitis, spine, vertebra, sickle cell disease.

Salmonella osteomyelitis is rarely seen, representing 1-4% of all bone infections[1]. It usually develops in immunocompromised patients, most typically in sickle cell disease (SCD). The overall incidence of osteomyelitis in SCD is approximately 2% to 5%[2]. Among the organisms responsible for osteomyelitis, a predominance of *Staphylococcus aureus* has been described; however, a recent review of the reports of the last two decades has indicated the importance of Salmonella in SCD[3]. Osteomyelitis usually starts in the medullary cavity of the humerus, radius, tibia, femur or ulna, in descending order of frequency[4]. A review of previous reports indicates that vertebral involvement by this organism is very rare, especially in children. The presentation of osteomyelitis with fever, bone pain and local swelling is usually acute.

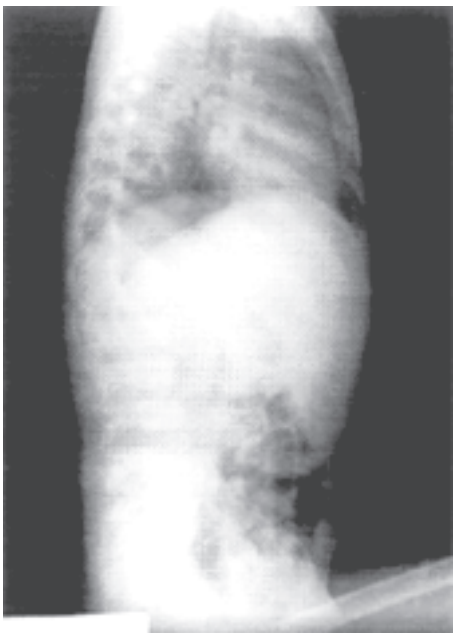
We report a case with SCD, in whom *Salmonella paratyphi B* vertebral osteomyelitis was diagnosed without typical clinical findings of infection. We would like to emphasize the differential diagnosis of osteomyelitis even at an atypical site with unexpected presentation in patients with SCD.

Case Report

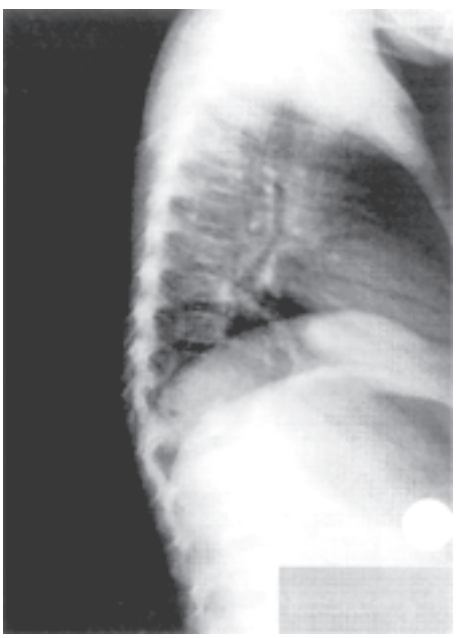
A five-year-old boy with homozygous SCD was admitted to our hospital with complaints of back pain, difficulty in walking, kyphosis and pain in the lower thoracic spine area. Those symptoms had begun about one month before admission. There was no history of gastroenteritis or fever.

Physical examination revealed that the patient was in discomfort and had a tender painful area at the T10-T12 level. He held both hips in flexion position to reduce the pain and was unable to stand up; other system findings were normal. Laboratory examination showed hemoglobin (Hb) 3.5 g/dl, peripheral leukocyte count 10,200/mm^3, with 62% neutrophils, 30% lymphocytes and 8% monocytes, and platelet count 477,000/mm^3. The reticulocyte count was 5%. The peripheral blood smear showed sickling and target cells with anisopoikilocytosis. Hemoglobin electrophoresis demonstrated the presence of HbS at a level of 24.5%. C-reactive protein was 3.99 (normal limit 0-0.8), but the sedimentation rate was 6 mm/hour. Magnetic resonance imaging (MRI) revealed compression and collapse at the T10 level; similar lesions

without collapsing features at the levels of the T8-T9 and T12-L3 vertebrae were also noted (Figs. 1a and 1b: initial plain X-ray, Fig. 2 MRI finding). The agglutinating titers against typhi-O antigen and paratyphi-b-0 antigens were 1/200, and repeated cultures of blood, stool and urine were negative, including for



(a)



(b)

Fig. 1a, 1b: Initial lateral radiographs.

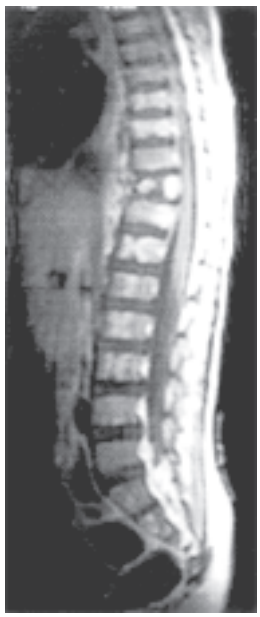


Fig. 2. MRI image shows mild compression at T10 level and disc narrowing at the T10-T11 level. Image revealed high signal intensity on the vertebral bodies at multiple levels.

Salmonella and tuberculosis. The clinical symptoms did not decrease by consequent exchange transfusions, and the second MRI on the 17th day of admission (Fig. 3) revealed that compression had increased and the potential space decreased. Anterior vertebrectomy, curettage, corpectomy and stunt grafting were performed, and at the second stage posterior

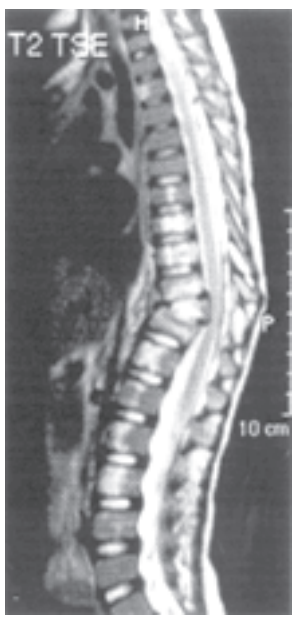


Fig. 3. Seventeen days after admission, significant kyphosis and spinal cord compression were observed.

instrumentation and fusion were added to the procedure (Figs. 4 and 5). The pathology of the surgical material showed an active chronic inflammation of the bones, and the *Salmonella paratyphi B* grew from two cultured bone materials, but all stool and blood cultures taken repeatedly were negative. He was given intravenous ciprofloxacin for two weeks and three partial exchange transfusions. In the postoperative course, posterior instrument was removed at 7th month of follow-up. Further follow-up for 12 months was uneventful.

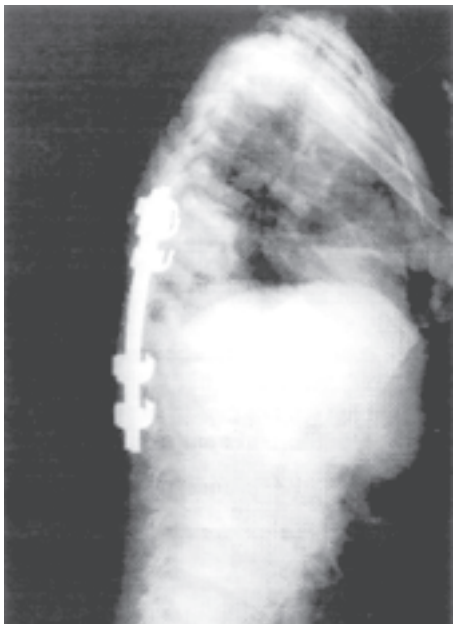


Fig. 4. Anterior corpectomy and stunt grafting were followed by the posterior instrumentation and fusion (early post-op).

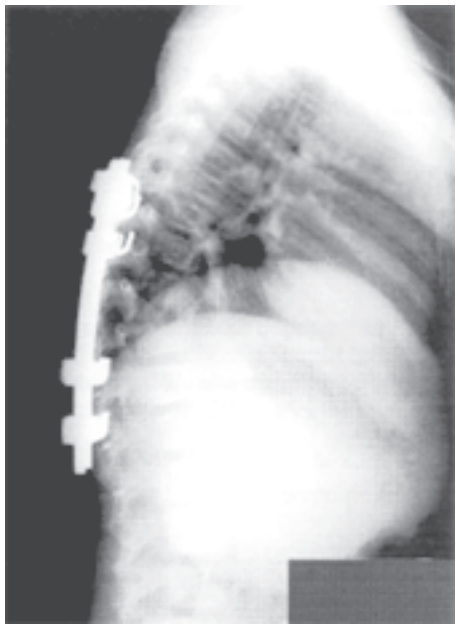


Fig. 5. Anterior corpectomy and stunt grafting were followed by the posterior instrumentation and fusion (late post-op).

Discussion

Patients with SCD are highly susceptible to Salmonella infections, especially in developing countries. The pathogenesis of illness due to bacterial infections is not understood yet but functional asplenism, impaired microcirculation, defective phagocytosis and hematogenous involvement may be responsible[5]. An increased rate of intestinal Salmonella carriage in patients with SCD has not been shown, but Salmonella may enter the bloodstream through micro-infarction in the intestinal mucosa.

Bone involvement in SCD consists of bone marrow hyperplasia, microvascular thrombosis, infarction and aseptic necrosis, which may play a predisposing factor for bacterial growth[6]. Sickling of the red cells results in thromboembolic infarcts in bone, leading to painful crises and also osteomyelitis[7]. Common sites of osteomyelitis in SCD are upper end of the femur, and the shafts of the tibia, radius, ulna and humerus[8]; the phalanges of both hands and feet, the ribs, vertebra, tarsal bones and innominate bone were less often involved[7]. Salmonella osteomyelitis represents 1-4% of all bone infections[1], and osteomyelitis of the spine is a rare condition, with an incidence of 0.45% of all cases of osteomyelitis[9]. Although several studies revealed Salmonella species as the most isolated organism in osteomyelitis cases associated with SCD[6,8,10-13], there is insufficient information about the real incidence of osteomyelitis of the spine associated with SCD.

Bone pains and frequent crises, whether they are due to osteomyelitis or thromboembolic infarcts of the bone, account for up to 80% of the hospital admissions of the patients with SCD[8, 14-16]. In a large series reviewing SCD, 20 of 70 patients with complete hemoglobin defects had at least one hospitalization for the treatment of osteomyelitis during a 10-year period[10].

Our case had no clinical or laboratory findings suggesting Salmonella infection, and his family serological and cultural tests were all negative.

In conclusion, rapid diagnosis and appropriate treatment of the infections are very important in order to prevent irreversible skeletal and neurological injury. Differential diagnosis between osteomyelitis and infarction should be done because of the similar presenting symptoms of both diseases, even if there is no obvious sign of infection. Therefore, if patients with SCD who complain of back pain do not improve despite partial exchange transfusions, infection should be considered. Biopsy and cultures from affected bone may be of great value in isolating the microorganism. Surgical intervention is necessary for both etiology and treatment.

REFERENCES

1. Van Oye E, Vassialiadis P. Oste'omyelite a' salmonella et HE'moglobinopathie. In: Van Oye E (ed). The World Problem of Salmonellosis (Monographiae Biologicea; Vol XIII). Philadelphia: 1964: 233-242.
2. Syrogiannopoulos GA, McCracken GH Jr, Nelson JD, et al. Osteoarticular infections in children with sickle cell disease. Pediatrics 1986; 78: 1090-1096.
3. Burnett MK, Bass JW, Cook BA. Etiology of osteomyelitis complicating sickle cell disease. Pediatrics 1998; 101: 296-297.
4. Adeyokunnu AA, Hendrickse RG. Salmonella osteomyelitis in childhood: a report of 63 cases seen in Nigerian children of whom 57 had sickle cell anaemia. Arch Dis Child 1980; 55: 175-184.
5. Epps CH Jr, Bryant DD 3rd, Coles MJ, et al. Osteomyelitis in patients who have sickle-cell disease. Diagnosis and management. J Bone Joint Surg Am 1991; 73: 1281-1294.
6. Kooy A, de Heide LJ, ten Tije AJ, et al. Vertebral bone destruction in sickle cell disease: infection, infarction or both. Neth J Med 1996; 48: 227-231.
7. Ebong WW. Acute osteomyelitis three years after a closed fracture in an adult with sickle-cell anemia. A case report. J Bone Joint Surg Am 1980; 62: 1196-1198.
8. Onuba O. Bone disorders in sickle-cell disease. Int Orthop 1993; 17: 397-399.
9. D'Souza CR, Hopp PG, Kilam S. Osteomyelitis of the spine due to Salmonella: case report, review of clinical aspects, pathogenesis and treatment. Can J Surg 1993; 36: 311-314.
10. Engh CA, Hughes JL, Abrams RC, et al. Osteomyelitis in the patient with sickle-cell disease. J Bone Joint Surg Am 1971; 53: 1-15.
11. Specht EE. Hemoglobinopathic salmonella osteomyelitis. Orthopedic aspects. Clin Orthop 1971; 79: 110-118.
12. Laurence B. Etiology of osteomyelitis in patients with major sickle hemoglobinopathies. J Pediatr 1981; 99: 411-413.
13. Piehl FC, Davis RJ, Prugh SI. Osteomyelitis in sickle cell disease. J Pediatr Orthop 1993; 13: 225-227.
14. Al-Dabbagh AA, Acquaye JK, Al-Baghdadi TM. Radiological bone changes in sickle cell disease in Western Saudi Arabia. Saudi Med J 1989; 10: 367-371.
15. Anionwu E, Walford D, Brozovic M, et al. Sickle-cell disease in a British urban community. Br Med J (Clin Res Ed) 1981; 282: 283-286.
16. Booz RE, Urbaniak JR. Radiological diagnosis of sickle cell disease. J Bone Joint Surg Br 1975; 59: 12.