

Spinal Involvement in Adult Patients with Sickle Cell Disease

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ABSTRACT

Background Data: Sickle cell disease (SCD) is the most common inherited blood disease that has obvious effects on the osteoarticular system and the spine. Despite Although vaso-occlusive crises and osteomyelitis are the most frequent complications requiring hospitalization for patients with SCD, the spinal involvement in adult patients with SCD has not been fully explored.

Study Design: Retrospective clinical case series.

Purpose: To elaborate on various types of spinal affection in adults with SCD and to discuss the effectiveness of conservative management in these patients.

Patients and Methods: Between January 2018 and December 2019, a retrospective study was conducted on 21 adults with SCD who presented to the neurosurgery clinic with back pain. The mean age was 27.4 ± 7.84 (range, 17–49) years, while 14 were females and 7 were males. The patients were divided into two groups according to the cause of back pain where Group I (N=12) included the patients who presented with back pain due to SCD related causes like osteonecrosis or infection, while Group II (N=9) included the patients who had back pain due to non-SCD etiologies such as vertebral disc protrusion or facet arthropathy. All patients were offered conservative management as a first-line treatment. Back pain was evaluated using the Wong–Baker Faces Pain Scale.

Results: Group I of 12 patients (57.1%) had SCD related causes including 11 patients (91.7%) with vertebral osteonecrosis (4 of them (33.3%) had associated osteoporosis) and one patient (8.3%) with acute lumbar osteomyelitis. Of the 11 patients who presented with osteonecrosis, 7 patients (63.6%) had an acute painful crisis, and the remaining 4 (36.4%) presented with chronic pain due to bone infarcts. In 8 patients (72.7%), the osteonecrosis involved the lumbar spine, while the thoracic and lumbar spines were involved in 3 patients (27.3%). Group II of 9 patients (42.9%) had non-SCD etiologies, including 2 patients (22.2%) who had facet arthropathy, 4 patients (44.4%) disc protrusion, and 3 patients (33.3%) mixed pathology. All patients were managed conservatively. The mean follow-up was 15.2 ± 5.38 (range, 3–24) months. Overall, the mean pre-management Wong–Baker Faces Pain Scale improved from 5.57 ± 1.121 (range, 4–8) to 1.95 ± 0.59 (range, 1–3) at the last follow-up.

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Conclusion: The most common SCD related spinal pathologies in adults are infarction, infection, and osteoporosis. The majority of SCD patients presenting with low back pain can be managed conservatively regardless of the associated pathology. (2020ESJ202)

Keywords: Spinal involvement; Sickle cell disease; Degenerative; Osteonecrosis.

INTRODUCTION

Sickle cell disease (SCD) is the most common inherited blood disorder.²⁹ It is an autosomal recessive genetic disorder resulting from a mutation in the beta-globin gene where the amino acid valine replaces glutamic acid in position six of the beta globin chain leading to the formation of an abnormal hemoglobin HbS molecule.²⁶ This abnormal HbS protein chain can polymerize reversibly in the presence of hypoxia into a gelatinous network of fibrous polymers that increases the viscosity, makes the RBC membrane rigid, and causes dehydration resulting in a sickle shape. The sickle cells lose their flexibility and are abnormally sticky resulting in episodes of microvascular occlusion and premature hemolysis. The pathophysiology of the manifestations of sickle cell anemia results from rigid adherent cells occluding the small vessels, leading to tissue ischemia and gradual end-organ damage. The vaso-occlusive crises may be precipitated by hypoxia, dehydration, fever, infection, acidosis, and exposure to cold.^{1,25}

Osteoarticular involvement represents one of the most common clinical presentations of SCD where it may present acutely such as painful vaso-occlusive crises or as a source of chronic disability. Painful vaso-occlusive crises and osteomyelitis are the most frequent complications requiring hospitalization for patients with SCD.^{4,21,24} However, spinal involvement in SCD, marked mainly by vertebral osteonecrosis and osteomyelitis, has been little studied.

This study aimed to describe the clinical manifestations of different types of spinal involvement in adult patients with SCD and to discuss the effectiveness of conservative management in these patients.

PATIENTS AND METHODS

This is a retrospective descriptive clinical case series that has been conducted on 21 adult patients with SCD spinal involvement and who were referred to Neurosurgery Outpatient Clinic in Najran Armed Forces Hospital. All patients with SCD aged ≥ 16 years complaining of back pain and referred by the internal medicine team to the neurosurgery clinic during two years from January 2018 to December 2019 were recruited for this study. The patients < 16 years were excluded from the study.

At the neurosurgery clinic, history of all patients was taken; they were subjected to general and neurological examination and laboratory and radiological investigations. This consisted of complete medical history including the history of previous vaso-occlusive crises, physical neurological examination, back pain assessment using the Wong–Baker Faces Pain Scale, blood tests for complete blood count (CBC), C-reactive protein (CRP), and the posterior-anterior and lateral plain radiographs of the segment of the spine likely to be affected. MRI of the spine was done to confirm the occurrence of vertebral osteonecrosis, to differentiate it from infection in suspected cases, and to detect non-SCD related causes of back pain. DEXA scan was conducted on patients with confirmed vertebral osteonecrosis on MRI of the spine.

The patients were divided into two groups according to the cause of back pain: Group I included the patients who presented with back pain due to SCD related causes like osteonecrosis or infection; Group II included the patients who had back pain due to non-SCD etiologies such as vertebral disc protrusion or facet arthropathy. The differentiation between the osteonecrosis and osteomyelitis was based on MRI findings together with the clinical and laboratory findings. High-grade fever and

markedly elevated inflammatory markers were in favor of osteomyelitis. The MRI findings of the spinal osteonecrosis included multifocal vertebral end-plate depression in all cases. Moreover, in the acute phases, there was associated marrow edema pattern near the vertebral end-plates in terms of low-signal intensity in T1WIs and heterogeneous high-signal intensity in T2 and STIR images. The spinal osteomyelitis presented with irregular vertebral end-plates, marrow edema pattern, and heterogeneous contrast uptake extending into the intervening disc substance (spondylodiscitis) with paraspinal small abscess formation.

All patients were offered conservative management as a first-line treatment. This included bed rest, pain management, medical osteoporosis prevention, and lumbar support. For acute pain management, tramadol 50 mg cap given twice daily after meal for 2 weeks, paracetamol 1 gm injected intravenously four times a day for 5 days, and pethidine hydrochloride 50 mg/ml 1 ml ampoule given by intramuscular route once a day PRN were used. Meloxicam 15 mg once daily was used for pain management in the chronic setting. Alendronate sodium 70 mg, one tablet per week, one hour before breakfast with plain water only for six months, was used for osteoporosis prevention. For the patients with spinal osteomyelitis, ceftriaxone 2 g IV once daily and flucloxacillin 500 mg IV three times daily for four weeks followed by ciprofloxacin 500 mg tab twice daily for another four weeks were used. The patients who had non-SCD etiologies were offered medical treatment including analgesics, neuropathic pain medications for associated neuropathic complaints, and physiotherapy. Surgical treatment was to be considered in patients with infections not responding clinically and laboratory to medical treatment in patients with persistent back pain ≥ 8 on the Wong–Baker Faces Pain Scale.

During the follow-up visits in the outpatient clinic, the patients were assessed clinically and when needed radiologically. The follow-up assessment was scheduled at 3-month intervals or in-between if the patients developed acute symptoms.

Improvement of the patients was considered on clinical backgrounds including pain assessment using the Wong–Baker Faces Pain Scale.

RESULTS

During the two-year period of the study, 21 adult patients with SCD suffered from back pain presented to our Neurosurgical Outpatient Service. The male/female ratio was 1:2 (7/14) (Table 1). The mean age was 27.4 ± 7.84 (range, 17–49) years and only one patient (4.8%) aged > 40 years (Table 2). All patients were known to be SS homozygous sicklers. Two female patients (9.5%) had associated thalassemia, while the other two male patients (9.5%) had G6PD.

The patients were divided into two groups. Group I included twelve patients (57.1%) who presented with back pain due to SCD related causes. They included 11 patients (91.7%) who presented due to vertebral osteonecrosis (Figure 1) (4 of them (33.3%) had associated osteoporosis) and one patient (8.3%) due to acute lumbar osteomyelitis. Of the 11 patients who presented with osteonecrosis, 7 patients (63.6%) had acute painful crises and the remaining 4 (36.4%) presented with chronic pain due to bone infarcts (Figure 2). In 8 patients (72.7%), the osteonecrosis involved the lumbar spine, while the thoracic and lumbar spines were involved in 3 patients (27.3%) (Figure 3). None of our patients had involvement of the cervical spine or thoracic spine alone. All patients in this group improved on conservative management; no surgical intervention was needed for the patients in this group. In this group, the mean pre-management Wong–Baker Faces Pain Scale improved from 6 ± 1.21 (range, 4–8) to 2 ± 0.60 (range, 1–3) at the last follow-up.

Group II included nine patients (42.9%) with SCD who presented with back pain due to non-SCD etiologies, including 2 patients (22.2%) who had facet arthropathy, 4 patients (44.4%) disc protrusion, and 3 patients (33.3%) mixed pathology (disc protrusion and facet arthropathy).

In 7 patients (77.8%), a single-level lumbar spine was involved: L5/S1 level in 4 patients (57.1) and L4/5 in 3 patients (42.9%). The double-level lumbar spine was involved in the remaining 2 patients (22.2%): one patient (50%) had L3/4 and L4/5 affection, while the other one (50%) had L4/5, L5/S1 involvement. The patients also in this group had a good response to conservative measures. In this group, mean pre-management Wong–Baker Faces Pain Scale improved from 5 ± 0.71 (range, 4–6) to 1.88 ± 0.61 (range, 1–3) at the last follow-up (Table 1).

The mean follow-up period was 15.2 ± 5.38 (range, 3–24) months. Three patients presented,

during the follow-up period, with ≥ 5 recurrent acute vaso-occlusive crises involving extraspinal systems (Table 3). Two of these 3 patients were a female had an associated thalassemia, and a male had involvement of the thoracolumbar spine in a previous attack. Moreover, 2 of these 3 patients had an associated osteoporosis (Figure 4). The patients with recurrent vaso-occlusive crisis involving extraspinal systems were managed by the internal medicine team and orthopedic team as there was frequent involvement of multiple joints. Their management included blood transfusion, proper hydration, hydroxyurea, and folic acid supplement.

Table 1. Summary of data of study patients' groups.

Parameters		Group I (N=12): sickle cell disease presentation	No.	Group II (N=9): non-sickle cell disease presentation	No.
Age/years		24.4±4.91 (range, 17–34)		30.4±9.14 (range, 19–49)	
Gender	Male	3		4	
	Female	9		5	
Back pain etiology		Osteonecrosis	11	Facet arthropathy	2
		Osteomyelitis	1	Disc disease	4
				Mixed	3
Wong–Baker Faces Pain Scale	Pre-management	6±1.21 (range, 4–8)		5±0.71 (range, 4–6)	
	Last follow-up	2±0.60 (range, 1–3)		1.88±0.61 (range, 1–3)	
Follow-up duration/months		15.3±6.40 (range, 3–24)		15±40 (range, 8–21)	

Table 2. Age distribution of the patients in the study.

Age/ years	Total No. (%)	Group I (N=12): sickle cell disease	Group II (N=9): non-sickle cell disease
16 -	4 (19%)	3	1
20 -	7 (33.3%)	5	2
25 -	6 (28.6%)	3	3
30 -	2 (9.5%)	1	1
35 -	1 (4.8%)	0	1
40 -	0 (0%)	0	0
45 -	1 (4.8%)	0	1
Total	21 (100%)	12	9

Table 3. Incidence of recurrence of vaso-occlusive crisis in patients with vertebral osteonecrosis (N=11).

Number of recurrent attacks	Number of the patients	Percentage
1-2	4	36.4%
3-4	4	36.4%
5 or more	3	27.2%
Total	11	100%

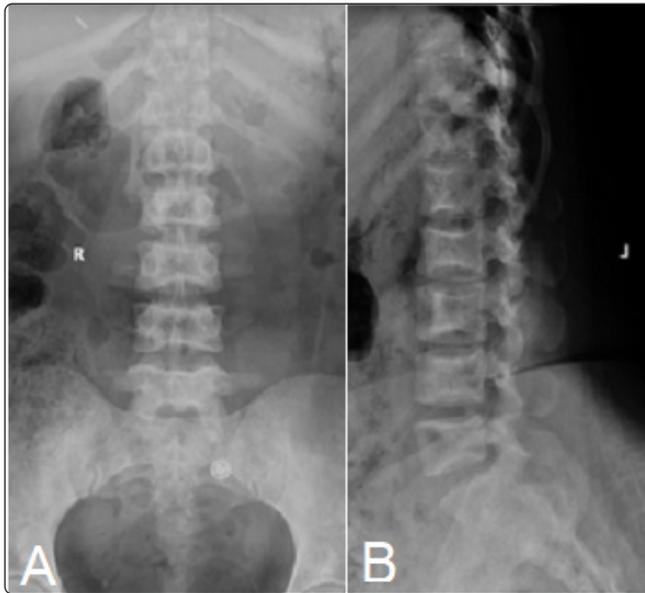


Figure 1. A 35-year-old female patient: (A) AP view lumbarosacral plain radiograph and (B) lateral view showing H-shaped configuration of the scanned lumbar vertebral bodies with central depressions of the vertebral end-plates. Coarse trabeculation and increased bone density of the examined bones are noted.



Figure 2. MRI lumbarosacral spine sagittal images: (A) T1WIs and (B) T2WIs of a known patient of sickle cell anemia and associated thalassemia showing diffuse red marrow hyperplasia as evidenced by diffuse low-signal intensity of the examined vertebrae on T1WIs and T2WIs.

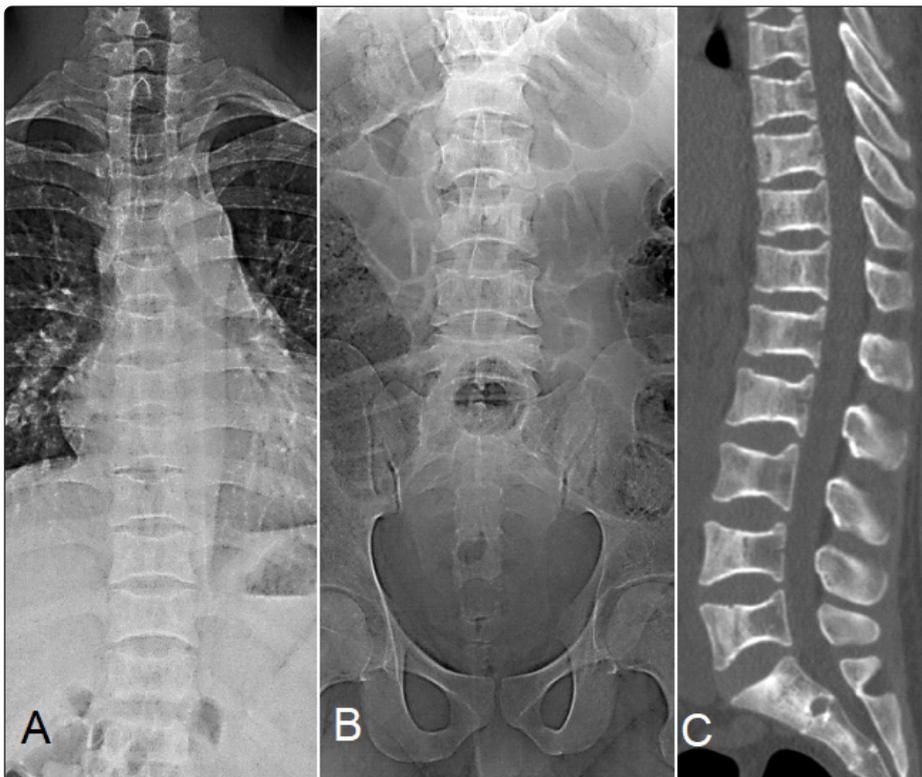


Figure 3. H-shaped configuration, coarse trabecular configuration, and increased bone density of the dorsal and lumbar spine are evident on (A) plain radiograph AP view of the thoracic spine, (B) AP view of the lumbosacral spine, and (C) MSCT-scan thoracolumbar spine sagittal images of a 25-year-old male patient.

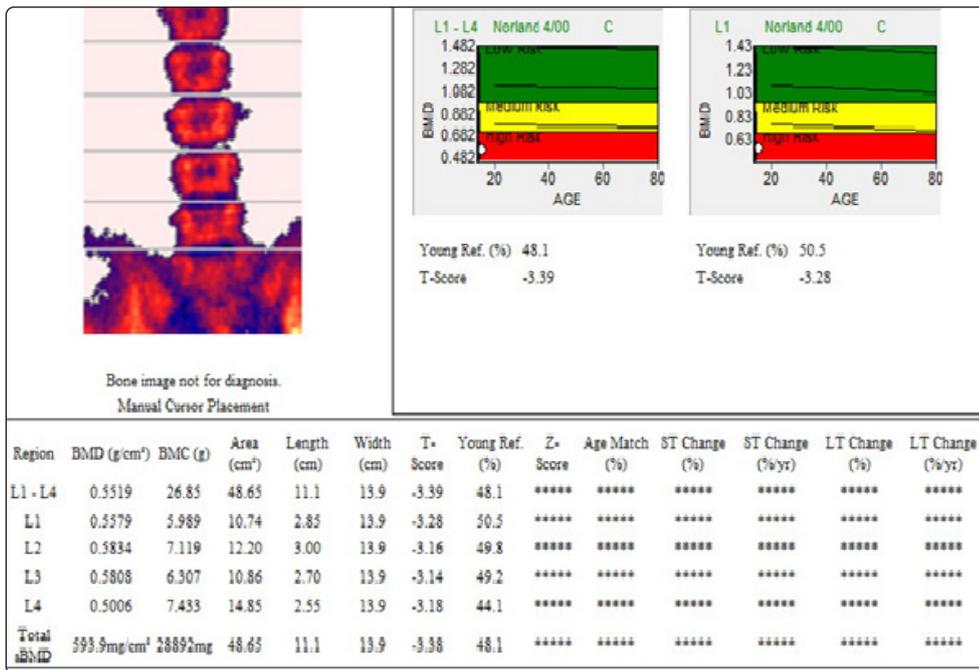


Figure 4. Osteoporosis at the lumbar spine (T score is -3.39) noted on BMD done for a 20-year-old male patient with SCD and G6PD who had recurrent vaso-occlusive crises.

DISCUSSION

Sickle cell disease is considered as a multisystem disease, as it causes multiple abnormalities, especially in the musculoskeletal system, yet, in many cases, it does not contribute to mortality. The affection of the spine and extremities is the most common clinical manifestation in these patients. The patients with SCD can present with acute bony problems, including painful vaso-occlusive crises, stress fractures, osteomyelitis, vertebral collapse, bone marrow necrosis, dental problems, and orbital compression syndrome. On the other hand, patients with SCD can present with chronic bony problems, including osteopenia, osteoporosis, avascular necrosis, and chronic arthritis.^{2,28} The most common of the spine pathologies affecting patients with SCD is vaso-occlusive crises.^{2,14,22} Severe back pain and tenderness over the involved vertebrae are the most significant clinical spinal manifestations of the vaso-occlusive crisis.⁵ In this study, eleven patients presented with vertebral osteonecrosis due to vaso-occlusive crisis, representing more than 90% of SCD related spinal involvement. These crises affect almost all SCD patients and have recurrent attacks throughout life.

Although the microvascular occlusion is the key event in acute painful crises, its pathogenesis is complex and involves the activation of leukocytes, platelets, endothelial cells, and RBCs containing HbS. These events can occur in any organ, but they are particularly common in the bone marrow.^{2,22} Seven patients in the study group presented with an acute painful crisis. In two-thirds of the patients, the lumbosacral spine is involved, while the thoracic spine is involved in 20% of the patients.²⁵ This is a little different from our study where the lumbar spine was involved in 70% of patients and both the lumbar and dorsal spine were involved in the remaining patients. The vertebral body is supplied centrally by the long branches of the vertebral nutrient artery, while peripherally it is supplied by short perforating branches of the periosteal vessels.¹ The longer end vessels are more affected than the shorter vessels in the vaso-occlusive crisis, resulting in destructive events. Moreover, the combination of vaso-occlusive infarction, compressive forces along the spine, secondary infection, and reactive bone response resulted in vanishing vertebra on the plain radiograph.²³ Regarding the imaging of the vaso-occlusive crises, plain radiographic examinations are usually not

conclusive in confirming or excluding osteonecrosis in the acute phase of the vaso-occlusive crises, as typically the bone appears normal.¹² However, there are many radiographic signs of the chronic phase of bone infarction, including the fish vertebra sign and vanishing of the vertebra in a patient with SCD where bone softening and ischemia of the middle part of the vertebral growth plate appear as fish vertebra sign, in the form of biconcave deformity and exaggeration of the concave curvature of the top and bottom surface of one or multiple vertebral bodies.²² Other radiological findings in the posterior-anterior and lateral radiographs of the spine include a stepladder effect, persistent anterior vertebral notching, coarse trabecular pattern, biconcavity of the bodies, massive vertebral collapse, and compression deformities.^{13,15} Magnetic resonance imaging and radioisotope bone scans might be more sensitive in detecting bony infarcts.¹² Radioisotope bone scans including combined Tc-99m-labeled sulfur colloid (for bone marrow uptake measurement) and Tc-99m diphosphonate (for bone uptake measurement) can recognize the acute phase of bone infarction.⁷

The second most common spinal pathology affecting the patients with SCD is osteomyelitis. It is usually triggered by vaso-occlusive crises, yet clinically it can be difficult to differentiate between spinal infarction and infection in patients with SCD as both conditions present with fever, tenderness over the involved vertebrae, and elevated inflammatory markers. Infection is suggested as a more likely diagnosis than infarction in the presence of high fever and erythema. The incidence of infection of the spine is markedly increased in patients with SCD. This is due to several factors including bone ischemia, splenic dysfunction, impaired complement activity, and excess iron secondary to increased hemolysis which is an important bacterial nutrient². In our study, one patient with SCD developed spinal osteomyelitis. Although the most common organism responsible for osteomyelitis in SCD patients is believed to be *Salmonella* with *Staphylococcus aureus* being the

second most common pathogen⁶, there are some reports of anaerobic osteomyelitis with *Bacteroides fragilis* where the patients had a good response to intravenous antibiotics.^{2,3,9}

Spinal column osteoporosis is common in patients with SCD due to reduced bone mineral density secondary to bone marrow hyperplasia. These patients usually have reduced vitamin D levels as well, hence an increased risk of pathological fractures.^{5,16,20} The incidence of low bone mineral density in adult patients with SCD ranged from 64% to 72% according to some studies when compared with age-, race-, and sex-matched controls.^{19,20} These results are higher than the findings in our study where about one-third of the patients with vertebral osteonecrosis were diagnosed to have osteoporosis. This may necessitate the need to perform more bone DEXA scan screening of the patients. The lumbar spine is the best site for assessment of bone mineral density according to a Brazilian study for children and young adults with SCD.¹⁸

The cause of back pain in these patients may also be not related to SCD like all other presentations of pain in the patients with SCD. In our study, about 40% of the adult patients with SCD had back pain due to non-SCD related causes including disc protrusion, facet arthropathy, and mixed pathology. That is why back pain should be fully evaluated in patients with SCD, particularly those patients who present with neuropathic symptoms as it becomes clear that other causes of back pain can be seen in patients with SCD and should be treated.

In this study, all patients were managed conservatively. Conservative management is the main line of management to treat the spinal affection in adult SCD patients. Symptomatic therapy of acute painful episodes, rest, orthosis, and osteoporosis medical therapy is usually employed for the treatment for vertebral osteonecrosis in adult patients with SCD.³ In patients with SCD who have spondylodiscitis in the absence of a neurologic deficit, the treatment is conservative too with intravenous and oral antibiotics.¹⁷ Ceftriaxone

and oxacillin are the usually used antibiotics as their spectrum covers *Salmonella* and *Staphylococcus aureus* which are believed to be the most common causative organisms of osteomyelitis in patients with SCD.⁶

The indications for surgical intervention in SCD patients include failure of antibiotic treatment therapy for osteomyelitis or spinal abscesses.⁶ Various anesthetic factors have to be considered during spine surgery in these patients to prevent vaso-occlusive crises, including the need to avoid hypoxia, dehydration, hypothermia, and circulatory stasis. Additionally, it is noted that rehabilitation in the postoperative period had been shown to reduce the incidence of vaso-occlusive crisis.¹¹ Spine surgery in adult patients with SCD has many complications including the collapse of osteoporotic vertebrae which is a very common complication in these patients. The implants may lose purchase and eventually pull out leading to a high rate of instrumentation failure in these patients.⁸ The techniques used to overcome these problems include augmenting of pedicle screws with acrylic cement or with resorbable cement, the use of screws coated with osteoinductive material, to use larger diameter screws, and expanding screws.^{8,10,27}

The limitations of this study include the relatively small number of adult patients with SCD who presented with back pain and the short follow-up period for 2 years as many patients presented with recurrent vaso-occlusive crises. Further studies with more patients with a longer follow-up period are recommended.

CONCLUSION

The spinal affection in adults with SCD may be due to SCD related causes or non-SCD related etiologies. The most common SCD related pathologies include vasoocclusive crises, osteomyelitis, and osteoporosis. They may have an acute or chronic presentation. The majority of SCD patients presenting with low back pain

can be managed conservatively regardless of the associated pathology.

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الملخص العربي

إصابة العمود الفقري عند البالغين المصابين بمرض الخلايا المنجلية

البيانات الخلفية: مرض الخلايا المنجلية هو أكثر أمراض الدم الوراثية شيوعًا والتي لها تأثيرات واضحة على الجهاز العظمي المفصلي و العمود الفقري. علي الرغم من أن الأزمات المؤلمة الوعائية والتهاب العظم هي المضاعفات الأكثر شيوعًا التي تتطلب دخول المستشفى للمرضى الذين يعانون من مرض الخلايا المنجلية، إلا أن مشاركة العمود الفقري في المرضى البالغين الذين يعانون من مرض الخلايا المنجلية لم تُدرس كثيرًا.

تصميم الدراسة: دراسة حالات إكلينيكية بطريقة الاستعادة.

الغرض من الدراسة: توضيح الأنواع المختلفة من تأثير العمود الفقري لدى البالغين المصابين بمرض الخلايا المنجلية وفعالية العلاج التحفظي في هؤلاء المرضى.

المرضى و الطرق: بين يناير 2018 وديسمبر 2019 ، تم إجراء دراسة بأثر رجعي على واحد وعشرين شخصًا بالغًا مصابًا بمرض الخلايا المنجلية الذين قدموا إلى عيادة جراحة الأعصاب يعانون من آلام الظهر. كان متوسط العمر 7.843 ± 27 (المدى 17-49) سنة بينما كانت 14 من الإناث و 7 من الذكور. تم تقسيم المرضى إلى مجموعتين تبعًا لسبب آلام الظهر، حيث تضمنت المجموعة الأولى المرضى الذين عانوا من آلام الظهر بسبب أسباب مرتبطة بمرض الخلايا المنجلية مثل تنخر العظم أو العدوى، في حين شملت المجموعة الثانية المرضى الذين يعانون من آلام الظهر بسبب المسببات غير مرض الخلايا المنجلية مثل بروز القرص الفقري أو اعتلال المفاصل. عُرض على جميع المرضى العلاج التحفظي كخط أول للعلاج. تم تقييم آلام الظهر باستخدام مقياس الألم من الوجوه وونغ بيكر.

النتائج: ضمت المجموعة الأولى اثني عشر مريضاً (57.1%) لديهم أسباب مرتبطة بمرض الخلايا المنجلية بما في ذلك 11 مريضاً (91.7%) يعانون من تنخر العظم الفقري ، أربعة منهم (33.3%) مصابين بهشاشة العظام ومريض واحد (8.3%) مصاب بالتهاب حاد بالعظم القطني. من بين المرضى الأحد عشر الذين أصيبوا بمرض تنخر العظم، يعاني سبعة مرضى (63.6%) من أزمة مؤلمة حادة ، بينما عانى الأربعة الآخرون (36.4%) من ألم مزمن بسبب احتشاء العظام. في ثمانية مرضى (72.7%) ، اصاب تنخر العظم العمود الفقري القطني ، بينما أصيب العمود الفقري الصدري والقطني في ثلاثة مرضى (27.3%). ضمت المجموعة الثانية تسعة مرضى (42.9%) لديهم مسببات غير مرض الخلايا المنجلية شملوا مريضين (22.2%) مصابين باعتلال المفاصل ، وأربعة مرضى (44.4%) كان لديهم نتوء القرص وثلاثة مرضى (33.3%) لديهم أمراض مختلطة. تم علاج جميع المرضى بعلاج تحفظي بدون اللجوء للجراحة. كان متوسط فترة المتابعة 5.382 ± 15.2 (المدى 3-24) شهرًا. تحسن مقياس الألم من الوجوه وونغ بيكر في المجموعة بأكملها من 1.12 ± 5.57 (المدى 4-8) قبل العلاج إلى 0.589 ± 1.95 (المدى 1-3) في المتابعة الأخيرة.

الاستنتاج: أكثر أمراض العمود الفقري المتعلقة بمرض الخلايا المنجلية عند البالغين هي الاحتشاء والعدوى وهشاشة العظام. يمكن علاج غالبية مرضى الخلايا المنجلية الذين يعانون من آلام أسفل الظهر بالعلاج التحفظي بغض النظر عن الأمراض المرتبطة بها.