



Relative rates and features of musculoskeletal complications in adult sicklers

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The purpose of this study was to prospectively look for the relative rates and features of musculoskeletal complications in a sample of adult homozygous SS sicklers in Yaoundé.

During a 3-year period, known homozygous SS sicklers aged sixteen years or more, with suspicion or evidence of locomotor system disease, including leg ulcer, were consecutively investigated through complete medical history, clinical examination, full blood count, C-reactive protein, standard radiographs of the area of complaint, and, when necessary, CT scan and pus analysis. Those patients with no definite diagnosis were excluded.

The study group comprised 84 patients aged 16 to 51 years (mean age : 22 years), with a male/female ratio of 0.75. Four of them (4.5%) were older than 40 years. Thirty five (41.6%) presented a total of 50 lesions of aseptic osteonecrosis, which were located in the hips in 25 cases (50%), in the lumbar spine in 20 cases (40%), in the humeral head in four cases (10%) and in the talar body in one case. The hip necrosis was grade I in 6 cases, grade II in four, grade III in 11 and terminal in four. Multiple sites of necrosis were observed in six patients. Nineteen (22.6%) of the sicklers came on with 36 malleolar ulcers, more frequently in males (sex ratio : 5/1) and 28 (78%) located on the medial side. Fifteen sites of osteomyelitis were noted in 14 patients (17.8%) and septic arthritis in six (7%). Less frequent complications were impingement syndrome, gout osteoarthropathy, stress fracture, subtalar fusion, knee osteoarthritis, tendonitis of the anterior tibialis, and recurrent dislocation of the patella. All patients were managed conventionally, except for advanced aseptic necrosis in which the indication for arthroplasty was delayed till the terminal stage.

As suggested by another recent report from Senegal, efforts should be made to improve the life expectancy of sicklers in Sub-Saharan African countries, by acting on education, social and medical care. Orthopaedic surgery should focus on reducing the failure rate of joint replacement in terminal stages of osteonecrosis and designing core decompression trials in early stages.

INTRODUCTION

Adult patients with sickle cell disease may be affected at any time in their life by three frequent musculoskeletal complications : osteonecrosis,

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malleolar ulcers, and chronic osteomyelitis. With rates ranging from 37 to 50% among adults sicklers, osteonecrosis has been reported as the most frequent of these complications (8, 12), followed by either malleolar ulcer, found in up to 25% of patients (11), or osteomyelitis which is most frequent in childhood (3). However, due to genetic and haematological variations, economic and cultural specificities, and finally, differences in access to medical facilities, it may be possible that the relative rates and clinical features of these complications fluctuate as a function of the area or the time. It therefore appears necessary, for any community with an homogeneous population of sicklers, to cyclically analyse the frequencies and pictures of these orthopaedic diseases in order to focus research efforts, design trials and hopefully, improve the management, the quality of life and the life expectancy of these patients.

The purpose of this study was to prospectively investigate the relative rates and features of different musculoskeletal complications including malleolar ulcers, in a population of homozygous SS adults sicklers, attending the haematology clinic of the central hospital of Yaoundé (Cameroon), a sub-Saharan African town.

PATIENTS AND METHODS

The study was a prospective investigation of orthopaedic conditions in a group of adult sickle cell patients attending the haematology clinic of the central hospital of Yaounde (CHY). All sicklers aged sixteen or above with suspicion or evidence of any musculoskeletal disorder except acute episode of pain, but including leg ulcer, were consecutively referred by the haematologist team to the orthopaedic clinic during a three years period, from may 2000 to June 2003. In the orthopaedic clinic, the patient underwent a program of systematic clinical, biological and imaging investigations in two steps. The first step consisted of complete medical history, physical examination, blood sampling for full blood count (FBC), C-reactive protein (CRP), pus sampling for bacteriological analysis in case of acute infection and standard antero-posterior and lateral radiographs of the segment or joint of complaint. The second step was indicated only if, after the first, no clear definite diagnosis was established or, for operative treatment or follow-up needs ; it consisted of standard CT

Table I. — Demography of the study group

Range of age	number	percentage	Cumulated percentage
16 to 20	39	46.5%	46.5%
21 to 25	22	26%	72.5%
26 to 30	10	12%	84.5%
31 to 35	6	7.5%	92%
36 to 40	3	3.5%	95.5%
41 to 45	0	00%	95.5%
46 to 50	3	3.5%	97.5%
51 & more	1	1%	99%
Total	84	100%	100%

Scan for painful joints with normal radiographs. Any patient without a diagnosis after this second step was treated symptomatically, returned to the haematology clinic and was excluded from the sample, as well as any one who was lost early before the end of the investigation program. All those with a confirmed orthopaedic condition benefited of conventional management. For those with chronic bone infection, an operative sampling for bacteriological analysis was done. All were advised to be seen at 3 and 6 months after discharge for follow-up. All these informations were collected and used for calculation of basic statistical values such as frequencies, percentages, ratios and means.

RESULTS

Demography

During the three-year period of the study, 97 adult sicklers were referred by the haematology team with a suspicion or evidence of musculoskeletal complications (MSC). Four of them refused the protocol, three were lost before the end of the investigation program, and six were screened negative, while 84 (89.7%) presented with confirmed MSC. The male/female ratio of the sample was 0.75, the minimal age was 16 years, the maximal age 51, the mean age 22 and only four patients (4.5%) were aged above 40 (table I).

Musculoskeletal complications

All 84 patients were well known SS homozygous sicklers ; their mean haemoglobin concentration was 7.8 g/100 ml.

Table II. — Relative rates of musculoskeletal complications in 84 SS homozygous adults sicklers

Type of musculoskeletal complications	Number of patients	Percentage of patients	Number of complications	Relative rate of complications
Aseptic necrosis	35	41.5	50	43%
Malleolar ulcer	19	22.5	36	31%
Osteomyelitis	15	18	15	13%
Septic arthritis	6	7	6	5%
Impingement syndrome	2	—	2	—
Gout osteoarthropathy	2	—	2	—
Stress fracture	1	—	1	—
Osteoarthritis of the knee	1	—	1	—
Spontaneous subtalar fusion	1	—	1	—
Tendonitis of tibialis anterior	1	—	1	—
Recurrent dislocation of the patella	1	—	1	—
Total	84	100	116	100%

Thirty five (41.6%) of them presented a total of 50 aseptic osteonecroses, more frequently in females (male/female ratio of 0.35); they were located in the hips (fig 1f) in 25 cases (50%), in the lumbar spine (fig 1d) in 20 cases (40%), in the humeral head (fig 1e) in four cases (10%) and in the talar body (fig 1c) in one single case. Based on Ficat's classification, the hip necrosis was grade I in 6 cases, grade II in four, grade three in eleven and terminal in four. Multiple sites of necrosis were observed in six patients : seven in one patient (both hips, one shoulder, three vertebrae and one talar body), six in another one (both hips, one shoulder and one vertebra), and two in four cases (both hips) ; 29 individuals presented osteonecrosis in a single site (19 in a hip, 14 in a vertebra, and 2 in a shoulder). Nineteen (22.6%) of the sicklers came on with 36 malleolar ulcers (fig 1a), more frequently in males (male/female ratio of 5), with 28 lesions (78%) located on the medial side. Fifteen osteomyelitis lesions (fig 1g) were observed in 14 patients (17.8%) while septic arthritis occurred in six (7%). Nine other less frequent orthopaedic conditions were observed in this series : two impingement syndromes (fig 1h), two gout osteoarthropathies (fig 1i), one stress fracture (fig 1b), one subtalar fusion (fig 1j), one osteoarthritis of the knee, one tendonitis of the anterior tibialis, and finally, one recurrent dislocation of the patella (table II).

Management and outcome

The eleven patients with grade III osteonecrosis of the hip and two with grade II lesions, who refused surgery, were treated conservatively with intermittent oral pain killers, crutches and life style counselling. Six with grade I and two with grade II underwent core decompression, while four others, with a terminal stage disease, were referred overseas for total hip arthroplasty. One of the 13 patients managed conservatively died four months later of acute pulmonary infection, one was lost to follow-up, three felt better, six felt no change while three did worse subjectively and on imaging at the sixth month. Seven of the eight patients with hip coring felt better at both check ups and their radiographs presented no change, while one felt no benefit although he did not worsen. The osteonecrotic talar body was resected and the ankle was grafted and fixed with an external fixator ; at six months, the patient was totally painless and the arthrodesis appeared fused on imaging.

Fifteen of the leg ulcers were managed with just bed rest, simple dressing and counselling, while four of them needed further debridement and skin graft ; all with good results immediately and after six months except three which recurred, but were finally controlled.

The osteomyelitis lesions were managed by conventional debridement, external fixation, targeted

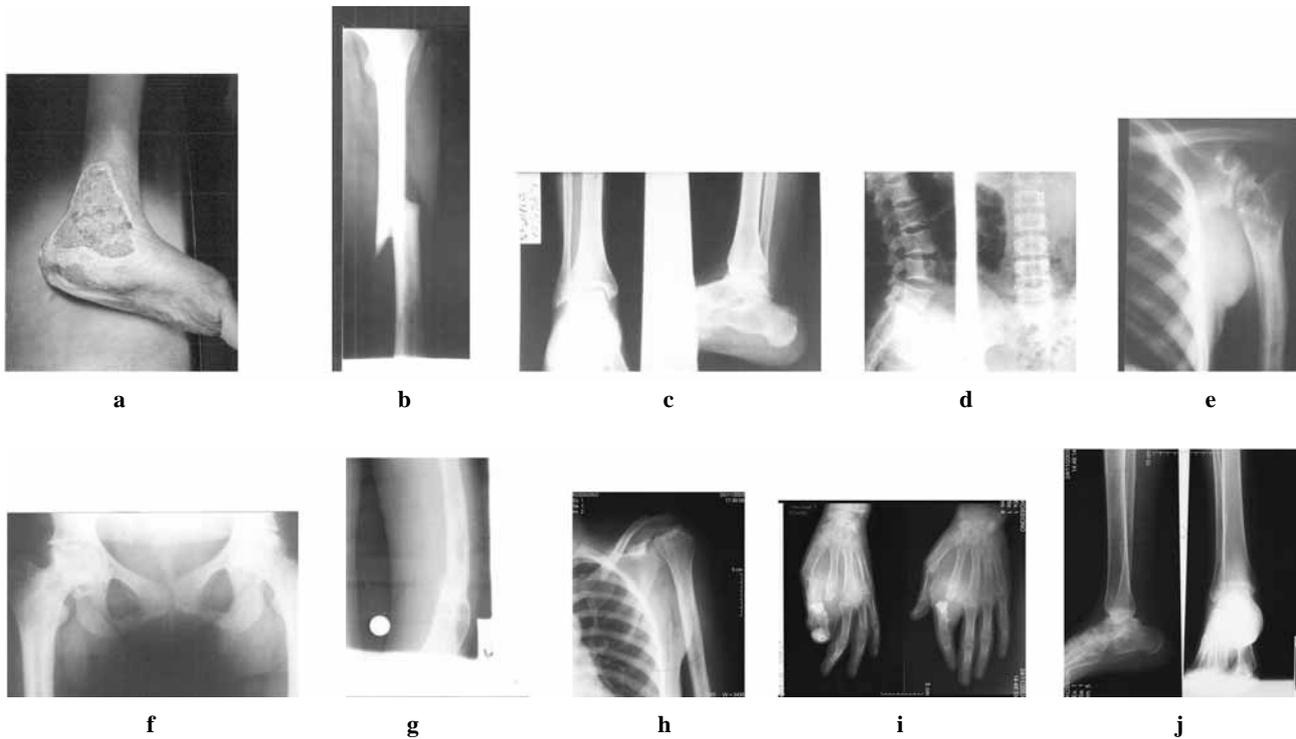


Fig. 1. — Showing 10 musculoskeletal complications in SS adults sicklers : (a) medial malleolar ulcer, (b) mid-shaft femoral stress fracture, (c) osteonecrosis of the talar body, (d) osteonecrosis of lumbar spine, (e) osteonecrosis of the humeral head, (f) osteonecrosis of both femoral heads, (g) osteomyelitis of the femoral shaft, (h) impingement syndrome of the shoulder, (i) arthritis nodosa of the hand and, finally, (j) spontaneous subtalar fusion.

antibiotic, and delayed bone and soft tissue reconstruction ; they all dried up after an average hospitalisation of six weeks. The septic arthritis responded well to surgical debridement and targeted antibiotics except for one patient who died of septicaemia and acute renal failure two days after the procedure.. The shoulder and spinal conditions were managed conservatively, the subtalar fusion and the knee osteoarthritis benefited from simple counselling, the stress fracture was internally fixed while the patellar dislocation underwent a lateral release combined with a medial advancement ; none of these patients complained at the six-month review.

DISCUSSION

This study presents the relative rates and features of musculoskeletal complications including

some which have been rarely, if ever, reported before, in a sample of 84 adult homozygous SS sicklers from a sub-Saharan African community. We hope that information gained in this study may help to improve the diagnosis and treatment of sicklers with orthopaedic complains, and finally, to foster further research. From the demographic point of view, less than 5% of our patients were aged above forty, which is similar to the findings of Diop *et al* (4) in Senegal, but very low, compared to 50% reported by Platt *et al* (9) in the United States. This poor life expectancy in sub-Saharan African sicklers may be related to many factors like the absence of hydroxyurea therapy which may improve the survival in this disease (10), the low educational background, the poor life style, and the limited access to medical facilities as suggested by Yetunde *et al* (13) in Nigeria. It is our opinion that, with increasing focalisation of the attention, fund-

ings and human resources on the HIV/AIDS pandemics, it will remain a challenge to improve the care of sicklers and thus, their life expectancy in sub-Saharan Africa.

Osteonecrosis ranks first among musculoskeletal complications, and is mainly located at the hip, as is currently reported in the literature (8, 12). Conservative management has been widely suggested for this complication rather than arthroplasty because of the known poor results of surgery (12, 7), the high rate of associated peri-operative complications (5, 12) and, finally, the poor life expectancy of these patients. At the beginning, it was also our policy to delay any major procedure until the advanced stage. However, we learned from this study that core decompression was efficient in stage I and II. We support the opinion of Vichinsky *et al* (12) that this procedure may be promising and that a trial on its long-term outcome should be designed. Malleolar ulcer ranks second among complications, with a rate of 22% close to the 25% found by Sawhney *et al* (11). Osteomyelitis presented no specific feature, and was managed according to our former reported protocol (1) with good result. Septic arthritis was the fourth musculoskeletal complication in our patients, but usually appeared as an immediate life-threatening issue ; one patient died from the generalisation of his sepsis. It should therefore be reminded that, as assumed by Mancini *et al* (6), in any acute infection in these patients, close monitoring and prompt aggressive treatment are warranted. The management of stress fractures has been discussed in our previous report (2). The recurrent dislocation of the patella was managed as usual since in our opinion, it was not specifically associated with sickle cell disease. None of all other less frequent musculoskeletal complications was life threatening ; they were therefore treated conservatively.

CONCLUSION

More efforts are needed to improve the life expectancy of Sub-Saharan patients with sickle cell disease. These effort should focus on education, social, and medical care. Surgery should focus on

reducing the peri-operative complications and failure rate of joint replacement in terminal stages of osteonecrosis ; for early stages, core decompression trials should be designed.

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