9 knee arthroplasties for sickle cell disease

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Sickle cell disease sometimes presents with knee arthropathy secondary to osteonecrosis. We report our experience with 9 total knee arthroplasties in 5 patients. The joint fluid, synovium, capsule, and excised bones were sent for culture. Tissue cultures from 5 knees grew organisms. The outcome was: 6 excellent, 1 good, 1 satisfactory, and 1 poor. Infection in 1 case led to removal of the prosthesis and subsequent arthrodesis. 1 patient had persistent peroneal nerve palsy. There were no hematological complications. We suggest that osteonecrosis in sickle cell disease should be considered septic.

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In sickle cell disease a vicious cycle of local hypoxia and further sickling of red cells may infarct an area of bone which may lead to arthropathy (Sennara and Grory 1978). Necrosis of the femoral head occurs in 19–31 percent of all sicklers, often bilaterally (Bishop et al. 1988, Bennett and Namnyak 1990). The knees are less frequently affected. Total hip arthroplasty for this disease has a high complication rate (Rand et al. 1987, Hanker and Amstutz 1988, Clarke et al. 1989). To our knowledge, there have been no reports on total knee replacements in sickle cell disease. We have retrospectively reviewed 9 total knee arthroplasties in 5 patients.

Patients and methods

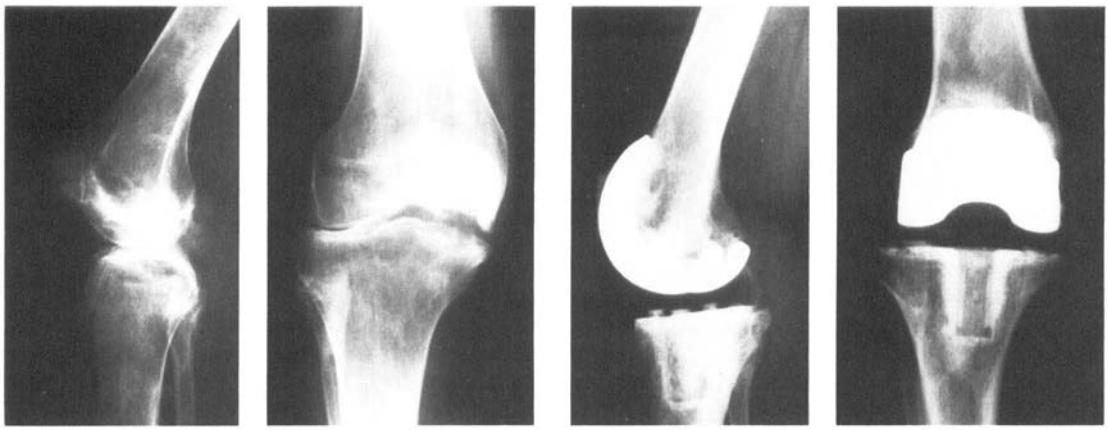
Between 1975 and 1990, 105 patients with sickle cell disease were admitted to our hospital. 36 of these patients were seen for orthopedic complications, notably 17 with necrosis of the head of the femur. 3 men and 2 women had bilateral knee involvement (Table 1). Their mean age at the time of surgery was 23 (17–29) years. 4 of the patients had been bedridden or wheelchair-bound for at least 1 year prior to surgery. Symptoms and signs were assessed pre- and postoperatively by the Hospital for Special Surgery (H.S.S.) knee rating score (Insall et al. 1976). All had severe

Table 1. Observations in 5 patients with 9 total knee arthroplasties for sickle cell disease (SS)

A	B	C	D	E			F	G	H		I		J	K	L
				HbS	HbF	HbA									
1	29	f	16	38	1	62	l	–	20–40	0–100	30	87	11	Excellent	
							r	*Salmonella*	25–40	0–100	32	90	11	Excellent	
2	22	m	18	78	20	2	l	–	0–40	0–90	36	89	3	Excellent	
							r	*Proteus Penneti*	0–90	0–100	39	91	2.5	Excellent	
3	22	m	19	75	24	1	l	*Staph. epidermidis*	30–80	0–95	14	91	3	Excellent	
							r	*Staph. epidermidis*	60–80	0–105	12	91			
4	24	m	17	93	2	5	l	*Enterobacter cloacae*	75–125	0–0	28	59	2.5	Poor	A, B
5	17	f	12	82	18	–	l	–	60–90	10–90	14	80	1	Good	B
							r	–	60–70	15–90	14	64	1	Satisfactory	C

A Case
B Age at operation
C Sex
D Age at onset of disease
E Hb electrophoresis in percentage
  HbS Sickle hemoglobin
  HbF Fetal hemoglobin
  HbA Adult hemoglobin
F Side
G Tissue cultures
H Motion range in degrees; pre, post
I Knee score; pre, post
  85–100 excellent
  79–85 good
  60–69 satisfactory
  <69 poor
J Follow-up time (yr)
K Outcome
L Comments
  A Fusion for infection
  B Temporary peroneal nerve palsy
  C Persistent peroneal nerve palsy

Figure 1. Case 1. A 29-year-old woman with sickle cell arthropathy.



Osteonecrosis of the lower end of the femur and upper end of the tibia.

11 years after total knee arthroplasty.

destruction of the knee joints (Figure 1), with severe pain at rest.

Preoperatively, all patients had hemoglobin electrophoreses to determine the level of sickle cell hemoglobin (Hbs); 4 patients (2–5) received preoperative blood transfusions to bring the Hbs level down to 45 percent. 4 patients underwent bilateral total knee replacements and 1 on the left side only. In 2 patients, 3 total hip arthroplasties were performed for femoral head necrosis. All patients received prophylactic antibiotics, Cafazoline 1 g intravenously perioperatively, and then every 6 hours for another 48 hours. 3 patients also received Gentamicin 80 mg intravenously every 8 hours for 48 hours.

The cemented Howmedica Kinematic and uncemented Howmedica PCA total knee systems were used. During surgery, the joint fluid, synovium, capsule and excised bone were sent for culture and histology. The antibiotics were changed according to the perioperative tissue cultures and sensitivity. In patients with positive tissue cultures, appropriate antibiotics were continued for a minimum of 6 weeks. In 2 knees hamstring release was performed to correct severe flexion contractures. 3 knees were treated postoperatively in an above-knee plaster splint for 2 weeks before commencing range-of-motion. In the remaining 6 knees, mobilization was commenced on the 3rd postoperative day. The mean follow-up was 5 (1–11) years.

Results

Out of the 9 arthroplasties, tissue cultures from 5 knees grew organisms (*S. epidermidis, Enterobacter cloacae, Proteus penneti* and *Salmonella*). The mean preoperative range-of-motion was 37°–73° of flexion. At the final follow-up, this had improved to 3°–85°. All the knees were clinically stable. 5 patients had no pain at follow-up, 2 had occasional pain and 1 patient was taking regular analgesics. The radiographs at the final follow-up showed no signs of loosening (Figure 1 B). Infection in Case 4 led to removal of the prosthesis and arthrodesis. There were no hematological complications. The results were 6 excellent, 1 good, 1 satisfactory, and 1 poor, based on the H.S.S. Knee Score.

Discussion

Clarke et al. (1989) reported deep infection in 4 of the 17 primary and revision hip arthroplasties for avascular necrosis (AVN) of the femoral head secondary to sickle cell disease. Sebes and Kraus (1983) reported one deep infection in 13 patients who had hip arthroplasties for sickle cell arthropathy of the hip. This high incidence of infection following joint replacement has been attributed to functional asplenia, immune system abnormalities, and relatively poor perfusion of blood in bone, secondary to sickling. We think that the main reason for such a high infection rate following a total joint replacement is the presence of active bacteria in the necrotic bone and tissue. Tissue cultures were pos-

itive in 5 knees out of 9 in our series, which supports the theory that often sickle cell osteonecrosis is septic rather than aseptic (Lifeso 1985).

The technical difficulties and the high complication rate in managing sickle cell hip arthropathy are well recognized (Clarke et al. 1989). Many of these problems are also present in the knee, the most troublesome being the presence of bacteria. The technical problem of correcting severe knee flexion contractures must also be addressed, specifically in relation to common peroneal nerve injury. Aggressive soft-tissue releases, including hamstring lengthening with traction or serial casting, should, perhaps, be considered prior to the knee arthroplasty in those patients with severe flexion contractures. Because of the frequent presence of a variety of pathogenic bacteria, an aggressive broad-spectrum prophylactic antibiotic routine should be instituted.

The prevention of sickle cell crises postoperatively is also of prime importance in these patients. This can be achieved by preoperative blood transfusion to bring the hemoglobin S level to less than 45 percent. The prevention of dehydration, hypothermia and hypoxia postoperatively is also important (Newton and Milner 1982).

Despite the complications, we feel that total knee arthroplasty is a valid option in dealing with sickle cell arthropathy of the knee.

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