ORIGINAL ARTICLE

Sickle-cell hip necrosis and intraosseous pressure

M.M. Mukisi a,∗, K. Bashoun a, F. Burny b,1

a Orthopedic-Traumatology Department, CHU de Pointe-à-Pitre, 97159 Pointe-à-Pitre, Guadeloupe
b Orthopedic-Traumatology Department, cliniques universitaires de Bruxelles, hôpital Erasme, Bruxelles, Belgium

Accepted: 6 January 2009

KEYWORDS
ONFH; Sickle-cell disease; Intraosseous pressure

Summary
Introduction. — Osteonecrosis of the femoral head (ONFH) is a frequent complication of sickle-cell disease. Numerous studies have demonstrated increased intraosseous pressure (IOP) in idiopathic necrosis and necrosis secondary to corticotherapy or alcohol poisoning. Several reports have testified to the clinical interest of decompression by drilling which, when performed in the early course of the pathology, can arrest or slow evolution. To the best of our knowledge, no studies have reported IOP increase in sickle-cell ONFH. The present study sought to show that intraosseous hyperpressure plays a role in the physiopathology of sickle-cell, like idiopathic, ONFH.

Materials and methods. — Sixteen intraosseous pressure (IOP) measurements were taken: eight in adult sickle-cell disease patients, four in sickle-cell trait carrying ONFH patients (AS) and four in non-sickle-cell ONFH patients (AA).

Arterial blood-pressure equipment with bone-puncture needle was used to measure IOP in the great trochanter body. Three IOP measurements were made after zero calibration: before drilling (direct pressure: IOP-1), after hyperpressure test but before drilling (IOP-2), and after drilling (IOP-3).

Results. — The present, admittedly short, series displayed elevated predrilling IOP-1 and IOP-2, reduced after drilling (IOP-3). Abnormal IOP fell after drilling performed for evolutive symptomatic ONFH. Significant differences in IOP-1 and IOP-2 were found, these being higher in the ’’sickle-cell disease’’ and ’’sickle-cell trait carriers’’ groups (p < 0.05). Only in the sickle-cell groups was there a significant correlation between pain score and hyperpressure level, with significantly reduced pain after drilling.

Discussion. — The elevated IOP levels found in symptomatic sickle-cell hips were comparable to those reported in the literature. Ischemia due to femoral head sinusoid occlusion by falciform globules with secondary intraosseous hyperpressure is the cause of the pain and of the onset and evolution of ONFH. The drilling tunnel acts as a safety valve, achieving real decompression of the segment involved and immediate postoperative reduction in or disappearance of pain.

∗ Corresponding author.
E-mail address: mukisi@ool.fr (M.M. Mukisi).
1 Thanks to Ms M. Donkerwolcke for her help in revising the article.

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doi:10.1016/j.otsr.2009.01.001
Introduction

Osteonecrosis of the femoral head (ONFH) is a frequent complication of sickle-cell disease, with an incidence estimated at 30–40% [1–7]. The literature contains no reports of hyperpression in sickle-cell-disease-related ONFH, although it could explain the severe pain associated with vaso-occlusion episodes. Bone decompression is the key to cessation of pain and favorable evolution of infarction and osteomyelitis lesions [8]. The present study seeks to demonstrate that hyperpression is at work in sickle-cell-disease-related ONFH, and the interest of early decompression drilling (Fig. 1).

Material and methods

Patients

Ten patients aged between 18 and 50 years, presenting with grade I, II or III uni- or bilateral ONFH (grade III being defined by subchondral dissection without loss of sphericity) underwent decompression drilling and pressure measurement in the Pointe-à-Pitre teaching hospital in Guadeloupe between October 2000 and December 2006. Six were sickle-cell patients (3 SS, 3 SC; 8 hips), two were sickle-cell trait carriers with ONFH (AS; 4 hips) and two were ONFH patients free of sickle-cell disease (AA; 4 hips). The classification used was that of Arlet et al. [9], adapted for modern imaging by ARCO [10].

Sixteen intraosseous pressure (IOP) measurements were taken: eight on adult homozygotic sickle-cell disease patients, four on necrotic hips in sickle-cell trait carriers (generally considered to be free of risk of severe complication, but in whom necrosis has nevertheless been reported in some rare cases) [11], and four on necrotic hips in sickle-cell-free patients. Ethical considerations precluded using the contralateral hip of the same patient as control [12].

Clinically, the patients presented with hip pain, limping and movement impaired by pain on rotation, especially internal. Pain intensity was assessed, before and after drilling, on a 0—100 numeric assessment scale, the patient setting a marker between the two extremes according to the intensity of pain experienced at a given time [13]. Sickle-cell disease patients are hyperalgesic, making pain assessment difficult due to under- or overestimation. Subjective estimation was therefore backed up by taking account of three degrees of analgesics consumption (I, IIa/b, III), with intensity thus classified as: 0 ''no pain'', 0—50 ''moderate pain'' relieved by I or IIa analgesics (non- or low morphine), and > 50 ''intense pain'' relieved by IIb or III analgesics (medium or high morphine) [14]. Pre- and postdrilling pain scores (PSpre and PSpost, respectively) were recorded. Diagnostic imaging (standard X-ray, CT and MRI) determined the osteonecrosis grade, following Arlet et al. [9].

IOP measurement

The technique, based on Arlet et al. [9], was simple and adapted to sickle-cell disease patients, who are fragile under anesthesia [15,16]. Taking our lead from intraosseous perfusion techniques, we employed a Jamishidi DJ4008X puncture needle, 8G × 10 cm [17]. Pressure was measured under visual control before and after decompression drilling of the femoral head. Measurement was feasible only in the great trochanter body (Fig. 1), by direct puncture 4 cm under the tip of the greater trochanter after perforation by a thin square-tipped awl and introduction of a bone-puncture needle connected up to a pressure sensor by a sterile tube filled with physiological saline. The patient was installed in dorsal decubitus, with the sensor at heart-level on the midaxillary line. The final value after stabilizing the mean IOP curve was read directly from the anesthesia ECG monitor. Correct cancellous positioning of the needle was confirmed by regular pressure-curve variation synchronized to the heartbeat. Direct measurement was followed up by an evoked hyperpressure test (intraosseous injection of 5 ml heparinized saline into the trochanter) to disclose any infraclinical pathology undetectable at baseline. Three measurements were made per
Table 1  IOP (mm/Hg) sickle cell patients (SS, SC).

<table>
<thead>
<tr>
<th>Patient/side</th>
<th>Type</th>
<th>Age</th>
<th>Age grade</th>
<th>IOP1</th>
<th>IOP2</th>
<th>IOP3</th>
<th>IOP2-IOP1</th>
<th>IOP1-IOP3</th>
<th>IOP2-IOP3</th>
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<tbody>
<tr>
<td>1L</td>
<td>SS</td>
<td>34</td>
<td>I</td>
<td>49</td>
<td>55</td>
<td>26</td>
<td>6</td>
<td>23</td>
<td>29</td>
</tr>
<tr>
<td>2R</td>
<td>SC</td>
<td>27</td>
<td>II</td>
<td>79</td>
<td>101</td>
<td>49</td>
<td>22</td>
<td>30</td>
<td>52</td>
</tr>
<tr>
<td>2L</td>
<td>SC</td>
<td>27</td>
<td>III</td>
<td>87</td>
<td>103</td>
<td>55</td>
<td>16</td>
<td>32</td>
<td>48</td>
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<tr>
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<td>SC</td>
<td>27</td>
<td>III</td>
<td>87</td>
<td>103</td>
<td>55</td>
<td>16</td>
<td>32</td>
<td>48</td>
</tr>
<tr>
<td>4L</td>
<td>SS</td>
<td>18</td>
<td>III</td>
<td>77</td>
<td>91</td>
<td>42</td>
<td>14</td>
<td>35</td>
<td>49</td>
</tr>
<tr>
<td>5L</td>
<td>SC</td>
<td>22</td>
<td>III</td>
<td>78</td>
<td>82</td>
<td>50</td>
<td>4</td>
<td>28</td>
<td>32</td>
</tr>
<tr>
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<td>11</td>
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<tr>
<td>6R</td>
<td>SC</td>
<td>28</td>
<td>III</td>
<td>82</td>
<td>90</td>
<td>34</td>
<td>8</td>
<td>48</td>
<td>56</td>
</tr>
<tr>
<td>Mean</td>
<td></td>
<td></td>
<td></td>
<td>72 ± 15</td>
<td>84 ± 18</td>
<td>42 ± 10</td>
<td>12.1</td>
<td>29.9</td>
<td>42</td>
</tr>
</tbody>
</table>

Significance: IOP1-IOP3, \( t = 7.98 \) (\( p < 0.001 \)); IOP2-IOP1, \( t = 5.65 \) (\( p < 0.001 \)); IOP2-IOP3, \( t = 9.44 \) (\( p < 0.001 \)).

Table 2  IOP (mm/Hg) sickle-cell trait patients (AS).

<table>
<thead>
<tr>
<th>Patient/side</th>
<th>Type</th>
<th>Age</th>
<th>Arlet grade</th>
<th>IOP1</th>
<th>IOP2</th>
<th>IOP3</th>
<th>IOP2-IOP1</th>
<th>IOP1-IOP3</th>
<th>IOP2-IOP3</th>
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</thead>
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<tr>
<td>1L</td>
<td>AS</td>
<td>50</td>
<td>II</td>
<td>98</td>
<td>102</td>
<td>41</td>
<td>4</td>
<td>57</td>
<td>61</td>
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<tr>
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<td>AS</td>
<td>50</td>
<td>II</td>
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<td>AS</td>
<td>50</td>
<td>II</td>
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<td>113</td>
<td>49</td>
<td>33</td>
<td>31</td>
<td>64</td>
</tr>
<tr>
<td>2R</td>
<td>AS</td>
<td>50</td>
<td>II</td>
<td>72</td>
<td>88</td>
<td>35</td>
<td>16</td>
<td>37</td>
<td>53</td>
</tr>
<tr>
<td>Mean</td>
<td></td>
<td></td>
<td></td>
<td>87 ± 20</td>
<td>100 ± 16</td>
<td>43 ± 9</td>
<td>14.3</td>
<td>44</td>
<td>58.3</td>
</tr>
</tbody>
</table>

Significance: IOP1-IOP3, \( t = 7.30 \) (\( p < 0.01 \)); IOP2-IOP1, \( t = 2.07 \) (NS); IOP2-IOP3, \( t = 22.6 \) (\( p < 0.001 \)).

Statistics

Hyperpressure was studied according to genetic group by comparison of means for small-sample paired series, using Schwartz’s formula [18]: \( t = \frac{m_s}{s/N^{1/2}} \), where \( t \) is read from Fisher and Yates’ table (DOF = \( N – 1 \); Tables 1–4).

Series 1 and 2 (sickle-cell disease SS and SC, or carrier AS) taken together were compared to series 3 (AA) by the formula: \( \frac{(M_A – M_B) \sqrt{S^2/N_A + S^2/N_B}}{N_A + N_B – 2} \), where \( S^2 \) stands for variance, taken as common (Tables 1 and 2).

Pain score (PSpre minus PSpost, equaling 1 or 2) according to the difference between IOP-2 and IOP-3 was assessed by the same formula with variance calculated as \( S^2 = \frac{\sum (x – m_A)^2 + \sum (x – m_B)^2}{n_A + n_B – 2} \).

Results

The 16 IOP measurements (Tables 1–3) showed elevation associated with both idiopathic and sickle-cell ONFH, and reduction following decompression drilling.

The study lacked power to disclose any difference in IOP values between the three series. Comparing series 1 and 2 (sickle-cell disease SS and SC, and carrier AS, respectively) taken together to series 3 (idiopathic necrosis) disclosed a significant difference for IOP-1 and IOP-2, which were higher in sickle-cell disease patients and trait carriers (\( p < 0.05 \)), but not for IOP-3 (postdrilling).

Comparing degree of hyperpression according to pain intensity as per Schwartz [18] showed no correlation for all patients and genotypes taken together. In sickle-cell disease patients and trait carriers (12 hips), in contrast, pain was reduced in nine hips (75%) and unchanged in three (25%, NS, \( p = 0.0833 \)), and the pressure difference before and after drilling (IOP-2 minus IOP-3) was \( 44.4 ± 9 \) mm/Hg for the nine hips showing hip: two before drilling (1 direct, IOP-1; one following the hyperpressure test, IOP-2), and one after core decompression (IOP-3).
Discussion

Larsen [19] and Miles [20] demonstrated the ischemic impact of intraosseous hyperpressure in the physiopathology of necrosis of the femoral head. The elevated IOP values found here in pathological hips of sickle-cell disease patients are comparable to those in the literature [9,21—24]. IOP-1 and IOP-2 were higher homozygotic (SS) and double heterozygotic (SC) sickle-cell subjects with necrosis than in sickle-cell-free (AA) patients. Our results support Ratcliff et al.’s contention [11] that bone abnormality is found in heterozygotic (AS) subjects, even if more rarely.

IOP was measured by a manometer in mm/Hg. It can be measured in two parts of the hip: femoral head and trochanter body, values taken in the former being 5 mm/Hg than in the greater trochanter [9]. Normal values do not exceed 30 mm/Hg (24 mm for Ficat et Arlet [22], and 26 mm for Klaer et al. [24]). Values above 30 mm/Hg are considered pathological [9,23]. Arlet et al. [9] provided reference values for IOP in the superior femoral epiphysis of adults with and without hip necrosis. IOP-3 fell significantly with decompression drilling, without, however, always passing the 30 mm/Hg threshold in sickle-cell disease (SS, SC) or trait-carrying (AS) patients.

Like Hauzeur et al. [23], reporting idiopathic ONFH, we found no correlation between degree of hyperpressure and sickle-cell-disease-related ONFH grade (Tables 1 and 2). Likewise, drilling produced an analgesic effect, with reduction or cessation of pain. The drilling hole is open after puncture/ablation of the osseous core and acts as an outlet, providing real decompression of the treated segment. It promotes conjunctive tissue colonization and thus vascularization of the femoral head by peritrochanter vessels the neovascular interpretation of necrotic bone-lesion repair put forward by Arlet et al. [9], which is compatible with the concept of cell repair by addition of bone marrow with drilling, as proposed by Hernigou et al. [25].

Initial necrosis cannot be seen on X-ray and can only be diagnosed on MRI; while the latter may be feasible in many countries, sickle-cell disease prevails in populations who have no such access. X-ray and clinical evolution indicates that early decompression drilling in clinical ONFH prevents necrosis developing into disabling osteoarthritis.

Conclusion

Pressure measurement is of diagnostic value in sickle-cell patients presenting with a symptomatic hip. Independently of surgery, manometry can be performed under local anesthesia to confirm early diagnosis of ONFH where MRI is not readily available. It can be performed quite straightforwardly, without pressure sensor, using a simple water column (physiological saline) and a three-way tap. The effectiveness of decompression drilling can thereby be assessed by comparing peroperative IOP-1 and IOP-3 values.

X-ray and clinical evolution indicates that early decompression drilling in ONFH prevents necrosis developing into disabling arthroscopic lesions. Patients experience definite hip pain reduction immediately after drilling. All reports concur on this. Decompression drilling in sickle-cell-disease-related necrosis serves two purposes: bone decompression with pain reduction after drilling in ONFH, and arresting the ischemic process before the femoral head loses sphericality or sequelae ensue from acute bone infarction.

References


