Diagnostic value of whole body MRI in patients with sickle cell disease

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Purpose

Introduction

• Sickle cell disease (SCD) is an inherited (AR) abnormality of B-globin chain resulting in a spectrum of hemolytic anemias.

• The most common type of SCD is sickle cell anemia (SCA). It occurs when both globin genes are abnormal (Hb SS).

• Other types of SCD are caused by combinations of hemoglobin (Hb) S with normal (Hb SA) or other abnormal hemoglobins, such as hemoglobin C [Hb SC] or B- thalassemia [Hb S- thal].

• The disease is most prevalent in Africa, Middle East, Mediterranean countries, and India. In western countries, it is more common in those of African origin.

Pathophysiology of SCD

• In hypoxic conditions, Hb S molecule is deoxygenated causing polymerization and resulting in loss of erythrocyte flexibility.

• Repeated cycles of oxygenation and deoxygenation cause irreversible membrane damage and formation of sickled cells.

• These cells are less deformable resulting in microvascular occlusion and haemolytic anaemia, which are typical of the disease

- The aim of work is to define the spectrum of abnormalities in sickle-cell disease using whole body MRI.

Methods and Materials

Patients
This study included 27 patients (15 Males, 12 Females) known to have sickle cell disease based on clinical examination and laboratory investigation (hemoglobin electrophoresis), their ages ranged between (4-35 years) with a mean age of 12 years. All patients were referred from hematology and orthopedic department to the MRI unit of the department of Radiodiagnosis, from August 2006 to October 2007.

- All the patients complaining of bony pain and some of them bony swellings.
- 9 patients complaining of hip pain and 2 patients from knee pain and swelling
- 5 patients complaining of old stroke
- 8 patients complaining of chest pain dyspnea and couph.
- 4 patients complaining of abdominal pain

Whole body MRI was performed to detect spectrum of abnormalities caused by multisystem affection. Spectrum of radiological abnormalities caused by sickle cell disease:

A- Musculoskeletal manifestation

1- Persistence of red marrow

2- Bone marrow expansion

3- Bone infarction

- May affect the medullary cavity of any marrow containing bone.

- Epiphyseal infarction

- Spine infarctions

4- Bone Infection (Osteomyelitis)

5- Joint Infection (Septic arthritis)

6- Muscles and soft tissue changes.

B- Extraskeletal manifestations

1- Brain

- Stroke
- Brain atrophy
- Cognitive impairment
- Aneurysm multiple in 57% of patients and may manifest with subarachnoid or intraparenchymal hemorrhage.

2- Lung
- Pneumonia
- Acute chest syndrome (ACS).
- Lung fibrosis, pleural adhesions & pulmonary hypertension

3- Spleen
- Infarction
- Sequestration syndrome

4- Liver
- Hepatitis from repeated transfusions.
- Ischemic necrosis, and cirrhosis.
- Pigmented gallstones.
- Intrahepatic biliary duct stenoses and cholestasis.
- Iron overload

5- Extramedullary hematopoiesis

**Technique**

A) Technique of Whole body MRI

MRI examination was performed using a super conducting 1.5 Tesla (T) magnet units.

The whole body was covered using both FSE-IR (Turbo-STIR) and T1-weighted FSE sequences in 4 coronal stations and 2 sagittal stations as follows:

1) Coils used:
- Body coils were used for the coronal stations.
- CTL (cervical, dorsal and lumber) phased array coil for sagittal stations.

2) Planes of examination:

- Body coverage was achieved using a maximum of four overlapping coronal body coil acquisitions. In each patient's coronal images were obliqued to the long axis of the spine.

- Position of the upper extremities was dictated by patient's habitus, in large patients, the arms were placed above the head, requiring an additional coronal acquisition.

- The spine was imaged in 2 overlapped sagittal stations parallel to the long axis of the spine in the coronal locator using the CTL coil. The 1st station included the cervical and upper dorsal vertebrae. The 2nd station included the lower dorsal and lumbosacral vertebrae.

3) Stations and parameters:

a) Locators:

A three-plan localizer scout view of the region of interest was performed for localization of the region to be scanned. This scout view was performed with spoiled gradient echo (fast multi planer spoiled gradient recalled), FMPSPGR.

b) Coronal Body Stations:

- 1st Station: was used to cover the head, neck, upper chest, proximal upper limb, and cervical and upper dorsal spine, using the following parameters:

<table>
<thead>
<tr>
<th>Sequence</th>
<th>TE/TI</th>
<th>Slice Spacing</th>
<th>FOV</th>
<th>Matrix</th>
<th>NEX</th>
<th>ETL</th>
<th>No. Of Slices</th>
<th>Time</th>
</tr>
</thead>
<tbody>
<tr>
<td>FSE-IR</td>
<td>5500</td>
<td>30/160</td>
<td>9.5mm1mm</td>
<td>48X48 256X192</td>
<td>8-12</td>
<td>22</td>
<td>4:24</td>
<td></td>
</tr>
<tr>
<td>T1 FSE</td>
<td>500</td>
<td>17.5/eff</td>
<td>9.5mm1mm</td>
<td>48X48 256X192</td>
<td>4-8</td>
<td>22</td>
<td>1:40</td>
<td></td>
</tr>
</tbody>
</table>

- 2nd Station: used to examine - 2nd Station: was used to cover the lower chest, abdomen, upper pelvis, distal upper limb, lower dorsal and lumbosacral spine, using the same parameters as the 1st station.

- 3rd Station: used to examine the lower pelvis and the thigh using the following parameters
**c) Sagittal Spine Stations:**

- **3rd Station:** used to examine the sternum, cervical and upper dorsal spine, using the following parameters:

<table>
<thead>
<tr>
<th>Sequence</th>
<th>TR</th>
<th>TE</th>
<th>TI</th>
<th>Slice Thickness</th>
<th>Spacing</th>
<th>FOV</th>
<th>Matrix NEX</th>
<th>ETL</th>
<th>No. Of Slices</th>
<th>Time</th>
</tr>
</thead>
<tbody>
<tr>
<td>FSE-IR</td>
<td>4000</td>
<td>30/eff</td>
<td>150</td>
<td>4mm</td>
<td>0.2</td>
<td>40X40</td>
<td>256X192</td>
<td>14-18</td>
<td>12</td>
<td>2:48</td>
</tr>
<tr>
<td>T1 FSE</td>
<td>525</td>
<td>17.5/eff</td>
<td>-</td>
<td>4mm</td>
<td>0.2mm</td>
<td>40X40</td>
<td>256X192</td>
<td>14-18</td>
<td>12</td>
<td>1:17</td>
</tr>
</tbody>
</table>

- **2nd Station:** it is used -2nd Station: was used to examine the lower dorsal, and lumbosacral spine. Using the same parameters as those used in the first sagittal station.

**B) Interpretation:**

Each case of Whole body MRI was analyzed in consensus by experienced radiologist searching for spectrum of abnormalities caused by sickle cell disease as previously described.
### Results

<table>
<thead>
<tr>
<th>Radiological abnormalities</th>
<th>NO of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persistence of red marrow</td>
<td>27</td>
</tr>
<tr>
<td>Bone marrow expansion</td>
<td>27</td>
</tr>
<tr>
<td>Bone infarction</td>
<td>27</td>
</tr>
<tr>
<td>Bilateral proximal femoral head infarction</td>
<td>9</td>
</tr>
<tr>
<td>Spine infarctions</td>
<td>23</td>
</tr>
<tr>
<td>Osteomyelitis</td>
<td>6</td>
</tr>
<tr>
<td>Septic arthritis</td>
<td>2</td>
</tr>
<tr>
<td>Old brain infarcts</td>
<td>13</td>
</tr>
<tr>
<td>Brain atrophic changes</td>
<td>8</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>5</td>
</tr>
<tr>
<td>Sequestration syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Iron deposition in the liver</td>
<td>21</td>
</tr>
</tbody>
</table>

Images for this section:
Fig. 1: Multiple bone infarcts in a 20 year old patient with SCD. (a, b, c,d,e) whole body coronal T1 IR weighted MR images showing multiple irregular areas of high signal seen scattered at dorsolumbar vertebrae, sacrum, left femur, ribs, both iliac bone, and left tibia.
**Fig. 2:** Fig 2. Acute bone infarct with extra osseous soft tissue signal abnormality in 17 year old patient with SCD. Whole body MRI, coronal T1 (a), IR (b, c) WIs show abnormal narrow signal intensity of the right femoral shaft with surrounding soft tissue, abnormal high signal during IR WIs (white arrow in b&c).
Fig. 3: Osteonecrosis of right femoral head in a 18-year old patient. Coronal STIR WIs of the pelvis showing STAGE IV avascular necrosis of the right femoral head and multiple bone infarcts of both iliac bones, sacrum, right femoral neck and proximal shaft
Fig. 4: Osteomyelitis in an 11 year old boy with SCD. Coronal STIR MR images show abnormal marrow signal with associated soft tissue oedema and minimal surrounding fluid collections.
Fig. 5: Septic arthritis in a 17 years old patient with left-sided knee pain and fever. (a) plain x ray of the left knee revealed erosive changes of the central tibial articular surface. (b&c) coronal STIR MRI revealed heterogenous texture of the distal femur and proximal tibia, erosive changes and fluid collection of the central proximal tibial articular surface and surrounding soft tissue edema.
Fig. 6: Acute chest syndrome in a 27 years old female patient presenting with severe dyspnea, fever, chest pain, and cough. (a, b & c) Whole body coronal T1, T2, and T2 IR WIs showing posteriorly located mild right sided pleural effusion with RT basal lung consolidation.
Fig. 7: Silent white matter infarct in an 18-year-old woman with SCA. (a) Axial T2-weighted MR image of the brain shows two irregular but well-defined areas of high signal intensity that represent chronic infarcts in left parietal region.
**Fig. 8:** Fig 8. Multiple vertebral infarctions in a 12 year old patient. Sagittal MR T1(a), sagittal MRI T2 WIs(b&c) showing heterogenous bone marrow signal intensity of the cervical, dorsal and lumbar vertebrae consistent with vertebral infarcts.
Conclusion

Whole body MRI can help identifying multisystem affection in sickle cell disease in a single session. MRI is a useful imaging tool in distinguishing acute osteomyelitis and bone infarct. Knowledge of the range of imaging findings is crucial in order to accurately depict the complication and initiate appropriate therapy.

References


Personal Information

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