

Case Report on Management and outcomes of Avascular Necrosis in sickle cell Anemia

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ABSTRACT:

Introduction: Components of the bone's cellular death induced by a blood supply blockage is known as avascular necrosis (AVN). Pain, loss of joint function, and long-term joint damage result from the collapse of bone structures. Osteonecrosis, aseptic necrosis, and ischemic bone necrosis are all terms used to describe AVN. **Present complaints and investigation:** The Male patient 22 year old who was admitted in AVBRH on date 07/07/2021 with chief complaint of Discomfort in both hips (left and right) was caused by distension of the afflicted joint, which could cause hip pain in the groin, thigh, or buttock. Aside The hip, shoulder, knee, hand, and foot, as well as both hips and knees, are all susceptible to injury. Following a test of X-rays, computed tomography scans (often known as CT scans), magnetic resonance imaging (MRI), radionuclide bone scans, and biopsies are all used in the physical examination. Bone function is assessed. **Past history:** 6 month ago patient admitted in rural hospital Yawatmal chief complaint on back pain and leg pain, loss of appetites, fever after the she diagnosis X-ray of took (MRI) the treatment for that. **The main diagnosis, therapeutic intervention and outcomes:** After physical examination and investigation doctor diagnose. Despite the fact that she never required urgent RBC exchange transfusions for ACS therapy, she had basic RBC transfusions frequently during her hospitalizations. With the following liver iron content, she had increasing transfusion hepatic iron overload. She was started on calcium and vitamin D supplementation. **Conclusion :-** Hi was response to all medication as well as doctor treatment and her recovery was good.

Keywords: Bone, Infection, Joint Pain, Hips, Necrosis, Sickle.

Introduction:

Components of the bone's cellular death induced by a blood supply blockage is known as avascular necrosis (AVN). Pain, loss of joint function, and long-term joint damage result from the collapse of bone structures. Osteonecrosis, aseptic necrosis, and ischemic bone necrosis are all terms used to describe AVN. It's critical not to ignore different genetic diagnoses. The symptoms and treatment of a patient with Warden burg syndrome type 4 (Wardenburg-Shah syndrome), an incredibly rare ailment, and homozygous sickle cell disease, a frequent disease in the Caribbean, are described in this case report.

This case is unusual in that both diseases may have been passed down the generations for the first time. Combination disorders are likely under-reported because to the prevalence of sickle cell and related hemoglobinopathies. Importantly, discussing this case will add to the medical literature by expanding knowledge of the phenotypic aspects of the condition.¹

Patient information:

The 22 year old patient was admitted in Acharya Vinoba Bhave Rural Hospital with the complaints of Discomfort in both hips (left and right) was caused by distension of the afflicted joint, which could cause hip pain in the groin, thigh, or buttock. Aside The hip, shoulder, knee, hand, and foot, as well as both hips and knees, are all susceptible to injury. Following a test of X-rays, computed tomography scans (often known as CT scans), magnetic resonance imaging (MRI), radionuclide bone scans, and biopsies are all used in the physical examination. Bone function is assessed.

Primary concern and symptoms of patient :- Present case visited AVBR Hospital in OPD base on date with complaints of Discomfort in both hips (left and right) was caused by distension of the afflicted joint, which could cause hip pain in the groin, thigh, or buttock. Aside The hip, shoulder, knee, hand, and foot, as well as both hips and knees, are all susceptible to injury before 1month .

Medical family and psychosocial history: Patient had medical history of Avascular necrosis (AVN) before 6 month. And history of splenomegaly with pemphigus vagaries. Hi took treatment for that but not cure . She belongs to nuclear family. There are five members in his family. All family members are healthy except the patient. patient look anxious, depressed and confused.

Relevant past intervention with outcome: history of cirrhosis of Avascular necrosis (AVN) 6 month back for which she was hospitalized for 20 days. After X-ray And (MRI)was observed she took treatment for that. And her outcome was not good. Physical examination and clinical finding

General examination – State of health was unhealthy, thin body built, but distension of necrosis the height of patient is 152 cm and weight is 65kg. her vital parameters are normal.

Timeline: 6 month ago she was admitted in the hospital for 20 days for the treatment of necrosis. Hi was also hospitalized for the treatment of Avascular necrosis (AVN) Currently she was admitted for the treatment of Avascular necrosis (AVN). Following a test of X-rays, computed tomography scans (often known as CT scans), magnetic resonance imaging (MRI), radionuclide bone scans, and biopsies are all used in the physical examination. Bone function is assessed. And medication, she had basic RBC transfusions frequently during her hospitalizations. With the following liver iron content, she had increasing transfusion hepatic iron overload. She was started on calcium and vitamin D supplementation. ²

Diagnostic assessment :-They can detect abnormalities in the bone that occur later in the course of avascular necrosis. X-rays are frequently normal in the early stages of the illness. MRI and CT scan are two types of imaging tests. RBC 3.66m/cu mm, Platelets count 1.19. Peripheral blood examination RBC 3.66m/cu mm , Platelets count 1.19. These tests yield high-resolution pictures that can reveal early abnormalities in bone that may suggest avascular necrosis. Following a test of X-rays, computed tomography scans (often known as CT scans), magnetic resonance imaging (MRI), radionuclide bone scans, and biopsies are all used in the physical examination. Bone function is assessed.

Diagnostic assessment:

Diagnostic challenging: No any challenging during diagnostic evaluation.

Diagnosis: After physical examination and investigation doctor diagnose a case of Avascular necrosis (AVN) .

Therapeutic intervention:

The patient was given medical care and an interferon alpha 2b solution for 10 days to improve immune function. Nonsteroidal anti-inflammatory drugs (NSAIDs) include ibuprofen (Advil, Motrin IB, and other brands) and naproxen sodium (Aleve).With the following liver iron She was becoming increasingly iron-overloaded as a result of transfusions. Calcium and vitamin D supplements were started for her.

Hi received all treatment and had a positive outcome. Her signs and symptoms had subsided, and she was able to resume her normal activities. There has been no change in the therapeutic intervention.³

Follow-up and Outcomes: Clinical and patient assessment outcomes: patient condition was improved. Important follow-up diagnostic and other test results: to preventing the progression of disease and trying to reserve any sign and symptoms that have appeared because of reduced of pane. a Doctor advised follow up after 1 month and X-ray and (MRI), blood investigation and blood pressure examination to know the further disease progression.

Intervention adherence and tolerability: patient took all prescribed medications regularly. but sometime she was refused to take medication. Hi also followed the dietician advised. Dietician was advised rich in calcium and rich in protein supplementation . Her interventional adherence was satisfactory.⁴

Adverse and unanticipated events: no any.

Discussion:

Osteonecrosis is a degenerative bone disease caused by a disruption in the subchondral blood supply, which causes the death of cellular components of bone. Avascular necrosis is a condition that affects the epiphysis of long bones at weight-bearing joints. Advanced illness can lead to subchondral collapse, which puts the joint's survival in jeopardy. As a result, early diagnosis and treatment of osteonecrosis are critical. The aetiology and pathogenesis of the illness, as well as the presentation and treatment choices for the most common kinds of osteonecrosis, are discussed.

Early management may be a way to slow down this process and prevent or delay the need for total joint arthroplasty. Synthetic grafting with a calcium sulphate and calcium phosphate composition that can be injected . The gradual revascularization and re-ossification of the region is enabled by the removal of necrotic bone and the interposition of a structurally robust osteoconductive substrate. While decompression and grafting will not change the underlying sickle cell disease process, we anticipate that they will improve sickle cell patients' quality of life. ⁵ Different studies on necrosis related to sickle cell disease were reported by Agrawal et.al.⁶ and Taksande et. al.⁷. Bhagat et. al. reported on poor health related quality of life among patients of sickle cell disease⁸. Borle et. al. reported a case of sickle cell osteomyelitis of the maxilla⁹. Other related studies on sickle cell disease were reviewed¹⁰⁻¹⁵.

Conclusion:

The patient was admitted to hospital with chief complaints of distension of Avascular necrosis (AVN) After all investigations patient was diagnosed with a case of Avascular necrosis (AVN). In our case stresses the need for good clinical assessment, good nursing care by trained nurses and the use of effective forensic studies is compulsory to secure patients from such a vital health condition.

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