ORIGINAL RESEARCH PAPER

Orthopaedics

KEY WORDS: AVN: Avascular necrosis, SCD: sickle cell disease; HGP; Haemoglobinopathies, VOC: Vasculo occlusive crisis, RBC.Red blood Cells, THR; Total Hip Replacement

SICKLING IN CG,OUR EXPERIENCES

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ABSTRACT

Sickle cell disease(SCD) is a genetically transmitted multisystem disease7 which includes a group of disorders that differs in severity sign and symptoms, The disease is not uniformly seen everywhere but it has some topographical distribution. In India, it is frequently seen in Central India, in and around the vicinity of Chhattisgarh in some religions in caste like kurmis, satnami, mahar , other backward caste and some tribes, it has great pathological significance considering the high morbidity and mortality resulting from the disease process . we have studied the cases of SCD from 2001 to 2015 series of such patients ,since there is no cure of this disease, in regards to prevention of this genetic autosomal recessive disorder by marriage counseling ,the incidence can be significantly reduced by avoiding consanguineous marriages in the susceptible community .

INTRODUCTION :-

Sickle cell disease is an inherited blood disorder that affects red blood cells. People with sickle cell disease have red blood cells that contain mostly hemoglobin, an abnormal type of hemoglobin S. Sometimes these red blood cells become sickle-shaped (crescent shaped) and have difficulty passing through small blood vessels. When sickle-shaped cells block small blood vessels, less blood reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged. This is what causes the complications of sickle cell disease .Normal RBC are soft and round & contains Hb.A .Abnormal haemoglobins[13] Hb.S & HbC are stiff, distorted and do not live long. The exact etiopathology at both molecular and genetic level has been extensively studied and deciphered thoroughly, yet there is no cure or established treatment regimen that will arrest the disease.

There are several types of sickle cell disease. The most common are : Sickle Cell Anemia (SS), Sickle-Hemoglobin C Disease (SC).

Sickle Beta-Plus Thalassemia and Sickle Beta-Zero Thalassemia.

Sickle cell trait[5](AS) an inherited condition where both hemoglobin A & S are produced in RBC but people with this are healthy.

caste	% of total population in C.G.	Incidence of carrier in the caste
SAHU	12%	25.7%
KURMI	10%	20%
S.T.	18%	20%
S.C.	18%	17%
TOTAL	58%	82.7%

Signs and Symptoms :- Sickle-cell disease may lead to various acute and chronic complications, several of which have a high mortality rate.

(I) Sickle-cell crisis[9].
(ii) Vaso-occlusive crisis
(iii) Splenic sequestration crisis
(iv) Acute chest syndrome
(v) Aplastic crisis
(vi) Haemolytic crisis

Fig 1: Clinical photo of sickling patient with chronic osteomyelitis tibia with ulcer

Fig 2: Clinical photograph

Clinico-Radiological Diagnosis4 :

(I) Early changes :-
(a) Hyperplastic bone marrow.
(b) Osteoporosis14
© Thinning of cortex
(d) Jelly-like marrow

(ii) Late changes :-
(a) Thickening of cortex
(b) Osteoblastic hyperplasia
© Narrowing of medullary cavity
(d) Softening and changes in bone shape
(e) Bone infarcts and irregular bony thickening of cortex6

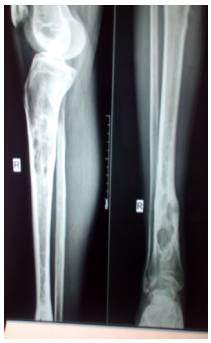

Fig. 3: X-ray leg of sickling pt. showing chronic osteomyelitis tibia

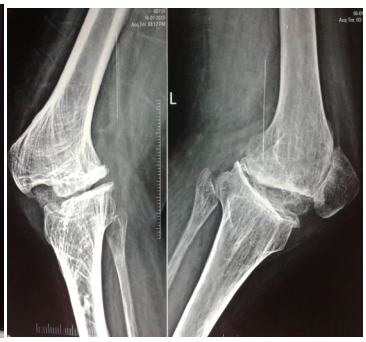

Fig. 4: X-ray of both knee with arthritis changes in sickling pt. with knock knee deformity

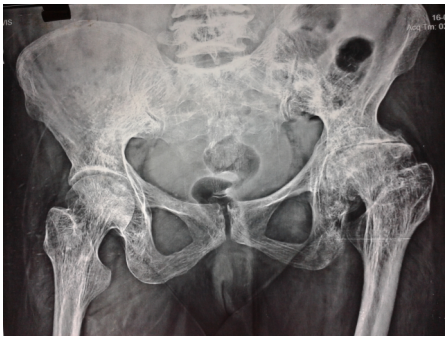

Fig. 5: x-ray pelvis,showing AVN in sickling pt.(preop.)

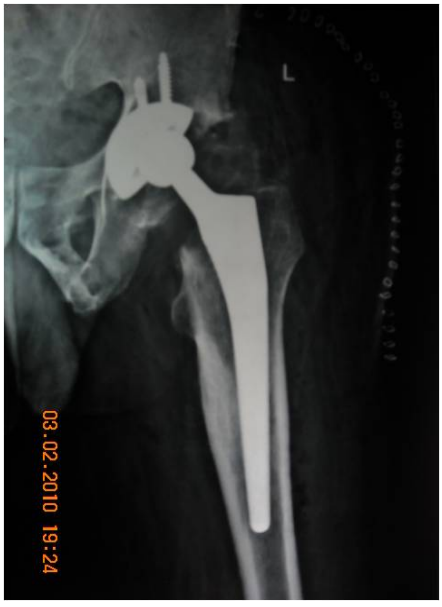

Fig. 6: Total hip replacement in AVN hip in sickling patient

Review of Literature :

(I) The condition was first described in the medical literature by the American physician **James B. Herrick in 1910**.According to **Cooley T.B. et al. 1927, Reynolds,** in his first description of the skeletal manifestations in erythroblastic anaemia, mentioned a case of Sickle cell anaemia which showed thickening and vertical striation of the occipital bone, similar to the cranial lesions of erythroblastic anaemia. The long bones in this case were normal.

(ii) Borzell f.F. 1933, also found changes in the skull and long bones similar to those of erythroblastic anaemia. **Grinnan** examined four cases of Sickle cell anaemia-4,5,7, and 11 years of age. The skulls in all cases showed lesions similar to those of erythroblastic anaemia.

(iii) Sickle cell anaemia with unusual bone changes, which occurred in a black male infant, was observed and described by **Danford E.A.** et al. in **1941.**

(iv) In 1943 Lins Pauling, noble price winner made it the first disease where the exact genetic & molecular defect was elucidated.

In our study sickling cases in Districts of Chhattisgarh

District of Chhattisgarh	Sickle Cell SS	Sickle Cell AS
Bilaspur	16	07
Raipur	19	10
Rajnandgaon	24	06
Bastar	22	09
Kabirdham,	24	08
Durg	26	10
Dhamtari	15	06

PATHOPHYSIOLOGY :

The loss of red blood cell elasticity is central to the pathophysiology of sickle-cell disease. Normal red blood cells are quite elastic, which allows the cells to deform to pass through capillaries. In sickle-cell disease, low-oxygen tension promotes red blood cell sickling and repeated episodes of sickling damage the cell membrane and decrease the cell's elasticity. These cells fail to return to normal shape when normal oxygen tension is restored. As a consequence, these rigid blood cells are unable to deform as they pass through narrow capillaries, leading to vessel occlusion and ischaemia.

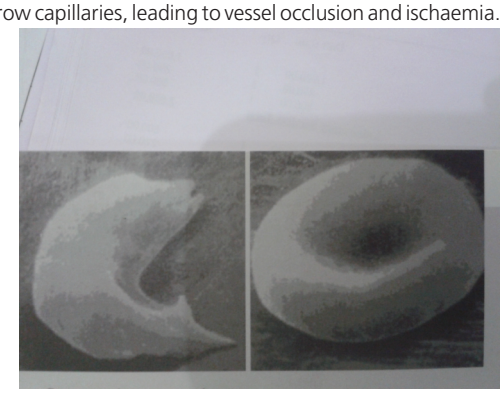

Fig. 7: Compare shape of RBC in sickling and normal patient.

The actual anaemia of the illness is caused by haemolysis, the destruction of the red cells, because of their shape. Although the bone marrow attempts to compensate by creating new red cells, it does not match the rate of destruction. Healthy red blood cells typically function for 90-120 days, but sickle cells live only 10-12 days.Looking at the complications and endresults, Govt of CG has now started giving such patients with permanent disability certificate just a compliment to help them somehow.

DIAGNOSIS :-

In Hb SS, the complete blood count reveals haemoglobin levels in the range of 6-8 g/dl with a high reticulocyte count (as the bone marrow compensates for the destruction of sickled cells by producing more red blood cells). In other forms of sickle-cell disease, Hb levels tend to be higher. A blood film may show features of hyposplenism (target cells and Howell-Jolly bodies).

Sickling of the red blood cells, on a blood film, can be induced by the addition of sodium metabisulfite. The presence of sickle haemoglobin can also be demonstrated with the **"sickle solubility test".** A mixture of haemoglobin (Hb S) in a reducing solution (such as sodium dithionite) gives a turbid appearance, whereas normal Hb gives a clear solution.

Abnormal haemoglobin forms can be detected on **Haemoglobin electrophoresis.**

COMPLICATIONS :-

- Increased risk of severe bacterial infections due to loss of functioning spleen tissue.
- Stroke, which can result from a progressive narrowing of blood vessels, prevents oxygen from reaching the brain.
- Hand foot syndrome
- Cholelithiasis (gallstones) and cholecystitis.
- Avascular necrosis (AVN)[1],aseptic bone necrosis of hip and other major joints.
- Decreased immune reaction due to hyposplenism.
- Priapism.
- Osteomyelitis[2] most commonly by Salmonella.
- Acute papillary necrosis in the kidneys.
- Leg ulcers.
- In eyes, retinopathy & vitreous haemorrhages, retinal detachments, leading to blindness.
- During pregnancy I.U.G.R. abortion and pre-eclampsia.
- Chronic Pain.
- Pulmonary hypertension.
- Chronic Kidney failures.

MANAGEMENT :-

Folic acid, IV Fluids,sodabicarb and higher antibiotics: - These patients take a 5mg dose of folic acid daily for life.

Hydroxyurea :- It reduces the number and severity of attacks.

Pain Management:- In Vaso-Occlusive crisis by NSAIDs diclofenac or naproxen and opioid administration at regular intervals.

Blood Transfusion therapy: - Used in management of sickle-cell disease in acute cases and to prevent complications by decreasing the number of R.B.C.

Bone marrow transplants :- Bone marrow transplants are the only known cure for SCD.

Surgeries; Replacement surgeries[3] , Core decompression, bone grafting & Various types of osteotomies[8].

DISCUSSION

Osteonecrosis femoral head(AVN) is common skeletal manifestation observed in SCD in our study with involvement of 65% articular surface femoral head requiring replacement[12] arthroplasty.Core decompression given.

In cases of early bone ischemia. Different intertrochantric osteotomies were attempted in younger children with necrosis of femoral head.

In cases of sickle cell disease, osteomyelitis[11] is fairly common manifestation were treated by sequestrectomy and saucerisation. pus culture with antibiotics and plaster cast/ fixator in pathological # cases.

In case of septic arthritis knee cases aspiration & antibiotic ,lavage done.

Other manifestation of SCD like hand foot syndrome & dactylitis[10] were treated only conservatively by supportive and symptomatically with good results Since SCD is inherited disease, parents should be taught about marriage consequence in between two SCD cases.

With regular medication(folic acid),avoiding dehydration & other precipitative causes of vascular crisis, regular follow-ups sickle cell disease patients can be well managed and live healthy life.

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