Difference Between Sickle Cell Disease and Sickle Cell Anemia

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Key Difference - Sickle Cell Disease vs Sickle Cell Anemia

Sickle cell disease is a common hereditary hemoglobinopathy caused by a point mutation in beta globin that promotes the polymerization of deoxygenated hemoglobin, leading to red cell distortion, hemolytic anemia, micro vascular obstruction and ischemic tissue damage. Sickle cell anemia is a severe hereditary form of anemia that arises as a result of the sickle cell disease where the mutated form of hemoglobin distorts the red blood cells into a crescent shape at low oxygen levels. **Sickle cell disease has a group of pathological manifestations** while **sickle cell anemia is one such pathological manifestation of the sickle cell disease.** This stands out as the key difference between sickle cell disease and sickle cell anemia.

What is Sickle Cell Disease?

Sickle cell disease is a common hereditary hemoglobinopathy caused by a point mutation in beta globin that promotes polymerization of deoxygenated hemoglobin leading to red cell distortion, hemolytic anemia, micro vascular obstruction and ischemic tissue damage.

Hemoglobin has a tetrameric structure which is made up of two pairs of alpha and beta chains. Normal adult red cells have HbA ($\alpha_2\ \beta_2$) as their dominant form of hemoglobin. In sickle cell disease, the glutamate residue in the sixth codon of the beta globin gene is replaced by valin. This substitution leads to various structural and functional changes in the hemoglobin molecule. In addition to HbA, people suffering from the sickle cell disease have a special type of hemoglobin in their red cells called the sickle hemoglobin (HbS).

Pathogenesis

Freely flowing cytosol of the red blood cells changes into a viscous gel when the partial pressure of oxygen drops below a certain critical level. With continued deoxygenation, HbS molecules polymerize into long fibers inside the red cells distorting them into a crescent shape. This is the pathological basis for the major

manifestations such as chronic hemolysis, micro vascular occlusion, and tissue damage.

As the HbS polymers grow, they start to herniate through the red cell membrane. This structural modification of the red blood cells induces an influx of Ca^{2+}.Increased intracellular calcium level then induces the cross linking of the intracellular proteins, resulting in an efflux of K^{+} and water. Repetition of this process dehydrates the red blood cells, making them rigid and dense. Ultimately they become irreversibly sickled cells which are rapidly removed from the circulation by extravascular hemolysis.

There are several notions about the pathological basis of the micro vascular occlusions, but the exact mechanism is not clearly understood.

Figure 01: Sickle cell disease is inherited in the autosomal recessive pattern.

Clinical Features

Sickle cell disease has a wide spectrum of clinical manifestations. Some of the affected individuals can be crippled while some can have only mild symptoms.

(Both sickle cell disease and sickle cell anemia have many common clinical manifestations which are discussed under the heading “clinical features of sickle cell anemia”)

Diagnosis

- Hemoglobin electrophoresis to demonstrate the presence of HbS
- Dithionate test
- Prenatal diagnosis is possible by the analysis of fetal DNA obtained by amniocentesis.

What is Sickle Cell Anemia?

The severe hereditary form of anemia that arises as a result of the sickle cell disease where the mutated form of hemoglobin distorts the red blood cells into a crescent shape at low oxygen levels is called the sickle cell anemia.

(Pathogenesis of sickle cell anemia has been discussed under the heading “pathogenesis of sickle cell disease”.)

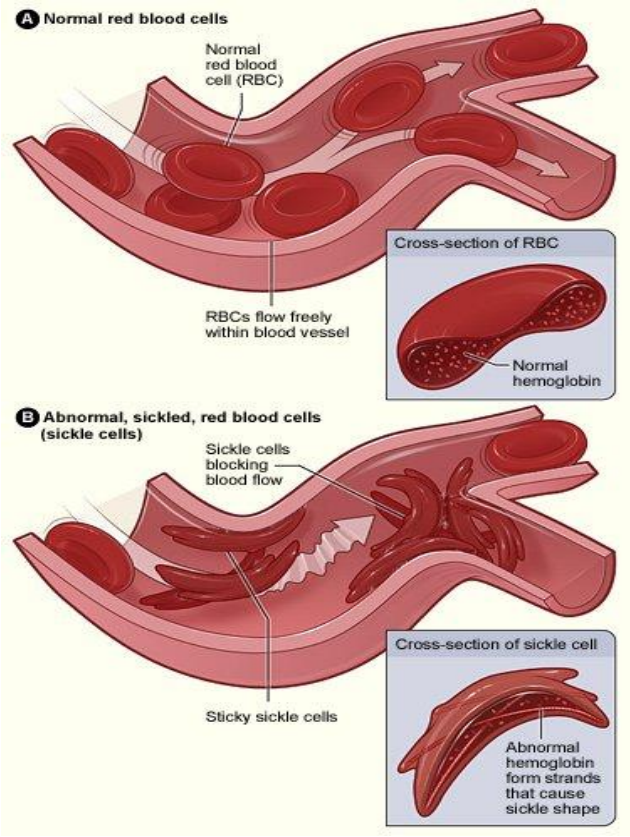

Figure 02: Sickle Cells

Clinical Features

Clinical features are of a severe hemolytic anemia punctuated by crises. There can be four main types of crises.

Vaso Occlusive Crises

Vaso occlusive crises are more frequent than the others and are predisposed by factors such as infections, acidosis, dehydration and deoxygenation. Because of the blockage of the blood vessels, blood supply to certain areas of the body, especially to the extremities, is compromised. Consequently, infarcts appear in these regions, giving rise to an intense pain. There is a condition called the hand foot syndrome where the patient complains of severe pain in the extremities. This happens because of the infarcts in the small bones of the limbs.

Visceral Sequestration Crises

These crises occur as a result of the sickling and pooling of blood inside the organs. Anemia exacerbates to a severe level during a visceral sequestration crisis. The acute chest syndrome is the most dangerous complication of this crisis. Patients suffer from a chest pain and dyspnea. A chest x-ray will show the presence of pulmonary infiltrates.

Aplastic Crises

These occur following a parvo virus infection and sometimes because of folate deficiency also. Aplastic crises are characterized by the sudden drop of hemoglobin level often requiring transfusion.

Hemolytic Anemia

Other Clinical Features

- Ulcers in the lower limb.
- Spleen is enlarged in infancy but is gradually reduced in size because of infarcts (autosplenectomy).
- Pulmonary hypertension.

Laboratory Diagnosis

- Hemoglobin level is usually 6-9g/dL.
- Presence of sickle cells and target cells in the blood film.
- Screening tests for sickling with chemicals such as dithionate are positive when the blood is deoxygenated.
- In HPLC, HbSS is detected as the dominant form of hemoglobin and HbA is not detected.

Treatment

- Avoiding the factors known to precipitate the crises.
- Folic acid.
- Good nutrition and hygiene.
- Pneumococcal, *Haemophilus* and meningococcal vaccination.
- Crises should be treated according to the condition, age and drug compliance of the patient.

What are the similarities between Sickle Cell Disease and Sickle Cell Anemia?

Both sickle cell disease and sickle cell anemia are caused by the same genetic mutation which affects the beta globin chains and thus the structure and function of hemoglobin. Since sickle cell anemia is one pathological manifestation of the sickle cell disease, they also share common clinical features.

What is the difference between Sickle Cell Disease and Sickle Cell Anemia?

Sickle Cell Disease vs Sickle Cell Anemia	
Sickle cell disease is a common hereditary hemoglobinopathy caused by a point mutation in beta-globin that promotes the polymerization of deoxygenated hemoglobin, leading to red cell distortion, hemolytic anemia, micro vascular obstruction and ischemic tissue damage	Sickle cell anemia is a severe hereditary form of anemia that arises as a result of the sickle cell disease where the mutated form of hemoglobin distorts the red blood cells into a crescent shape at low oxygen levels.
Pathological Manifestations	
Sickle cell disease has several pathological manifestations.	Sickle cell anemia is one such pathological manifestation of the sickle cell disease.

Summary - Sickle Cell Disease vs Sickle Cell Anemia

Both sickle cell disease and sickle cell anemia are common hereditary conditions and proper treatments can be helpful in elevating the standard of living of the patient. Sickle cell disease has a group of pathological manifestations while sickle cell anemia is one such pathological manifestation of the sickle cell disease. This is the main difference between sickle cell disease and sickle cell anemia.

References:

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