Institute of Haematology & Oncology

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Haematology and Haemato-Oncology Newsletter

Many thanks for visiting our new LOTUS Institute of Haematology, Oncology and Bone Marrow Transplantation on 6th March 2011. Those of who could not come, I extend my personal invitation for a visit.

Cases 1,2 and 3 will be discussed together as they all had Sickle cell anaemia.

Case 1: Referred by Dr Pravin Supe/Dr Manoj Chopda

Case 2: Dr Sameer Bhide Case 3: Dr Kailash Rathi.

As you know we have3 prominent sickle cell belts in Nashik, Dhulia and Nandurbar Districts.

Case 1 : He is a 14 year old male with recent diagnosis of sickle cell anaemia which was diagnosed when he came with typical vaso-occlusive crises. He came to his physician with breathlessness and fever and was shifted to ICU because of drop in oxygen saturation. He was conscious with no seizures. His CXR showed bilateral shadowing suggestive of infiltrates. His oxygen levels dropped and he was put on BIPAP. Investigations: Hb: 6.4, wbc: 19,200 (neutrophilia). Raised S.Bilirubin and enzymes. S. creatinine was normal.

Case 2 : He is a 12 year old male with history typical of sickle cell anaemia and was diagnosed during admission. He had persistent fever which settled with second line antibiotics. There was no sign of osteomyelitis(Salmonella osteomyelitis is common in sickle cell anaemia and can cause PUO). 6 days into his admission he started to complain of chest discomfort and after 12 hours dropped his saturation to 76%. CXR was suggestive of chest infiltrates. Investigations: Hb: 5.8. wbc: 22,100. Bilirubin was raised. Enzymes were normal. He was put on high flow oxygen.

Case 3 : She is a 17 year old female who presented with sudden onset unconsciousness to her GP and was then transferred to ICU. No focal neurological signs. All routine investigations were normal except Hb which was 6.3 wbc: 29,100(82% neutrophils). Platelets were normal. I was asked to review the PBS for malaria, but I found sickle cells and then we diagnosed her to have sickle cell anaemia.

CT scan was normal. MRI Brain revealed Posterior Reversible Encephalopathy Syndrome (PRES)(reported by Dr Rohan Kashyape) which can be seen in variety of conditions including sickle cell vasculopathy. We diagnosed her as PRES secondary to sickle cell vasculopathy.

Management (of all 3 cases) :

All of our three patients had life threatening complications due to sickle cell anaemia. First 2 patients had "ACUTE CHEST SYNDROME". They happened due to increased percentage of sickle cell in them which caused vascular occlusion. The only curative and life saving option for them in EXCHANGE TRANSFUSION.

What is exchange transfusion ?

Quite simply it is replacing patients sickle rich blood with non-sickle cell normal blood, rather packed cells!

How do we do it ?

We insert 2 cannulas in forearm or a central line. We first remove 1 unit of blood from one arm and then give packed cells from other arm. We carry out these cycles 4-6 times depending on the patients weight.

Normally sickle percentage in such patients is between 70-85%. We aim to bring it down to 30%. Once this happens the sickling process stops and the patient dramatically improves.

Clinical outcome :

All 3 patients improved and were discharged in good condition. We have started Cap. Hydroxyurea in all of them. The mortality if not treated and recognized early is almost 100%.

Take home message :

1. Early diagnosis of sickle cell disease is important as we can start them on medications which can reduce their vaso-occlusive crises.
2. Acute Chest Syndrome can closely simulate bronchopneumonia, ARDS and if not recognized early can be fatal.
3. Exchange transfusion is a life saving , simple measure which has to be started early in the course of the illness.
4. Always think of sickle cell anaemia in unusual presentations and simple PBS looked upon carefully clinches the diagnosis.
5. Sickle cell anaemia can be missed throughout the childhood and they can present late to adult physicians as well! We have diagnosed sickle cell anaemia in a 34 years pregnant female who presented with heart failure in third trimester of pregnancy.

Laboratory news :

1. We will soon install platelet aggregometer which will help me in diagnosing cases who come with excessive bleeding, but in whom all routine coagulation tests are normal.
2. We have installed 5 part differential cell counter, where neutrophils, lymphocytes, eosinophils, monocytes and basophils are separated and we get more accurate reports.

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