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Haemorrhagic Telangiectasia : Case Study

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A B S T R A C T

Osler Weber Rendu also known as hereditary haemorrhagic telangiectasia (HHT) is an autosomal disorder affecting the multiorgan system, characterized by unprovoked nosebleed, mucocutaneous telangiectasia, abnormal blood vessels formation and arteriovenous malformations (AVM). Patient is suffering from an autosomal disorder, HHT, from last 19 years. She got admitted with chief complaints of bleeding from nose, tongue and syncope. The patient was anaemic with Hb of 4.5g/dl. After one unit PRBC and one unit FFP transfusion Hb raised to 5.5mg/dl. Oral tranexamic acid was given to control the unprovoked bleeding from nose and tongue. The prognosis of the HHT was good.

Keywords: Osler Weber Rendu disease, hereditary haemorrhagic telangiectasia, autosomal dominant disorder.

1. Introduction

Hereditary Haemorrhagic Telangiectasia (HHT) is an autosomal disorder affecting the multiorgan system, characterized by unprovoked nosebleed, mucocutaneous telangiectasia, abnormal blood vessels formation and arteriovenous malformations (AVM). It is also known as Osler Weber Rendu disease. Since it is a genetic disorder, one can inherit it from parents. Diagnosis is based on physical examination, health history and some imaging tests like, ultrasound imaging, magnetic resonance imaging.

2. CASE PRESENTATION

A patient aged 59 years old visited IGMC Shimla with the complaints of bleeding from nose, tongue and syncope. She was experiencing these conditions from last one week.

She is suffering from this disease from last 19 years.

Past Medical History

Patient was suffering from hypertension but currently patient's vitals are within normal range.

3. SPECIAL INVESTIGATION

According to the reported symptoms, patient's haemoglobin level was monitored. At that time patient's haemoglobin level was 4.5g/dl which was lower than the normal range of blood haemoglobin level.

4. TREATMENT

Inj. Tranostat 1g IV BD Tablet- IFA BD

One unit PRBC and one unit FFP

5. INTERVENTIONS

Drug should be taken after having meal.
Suggest patient to check haemoglobin level frequently.

6. CARE PLAN

Proper diet – green leafy vegetables

7. OUTCOME

Patient used suggested medicine and after taking the medicine bleeding was controlled. After blood transfusion the haemoglobin level was monitored. Hb- 5.5mg /dl.

8. CONCLUSION:

Patient is suffering from an autosomal disorder, HHT, from last 19 years. She got admitted in IGMC Shimla with chief complaints of bleeding from nose, tongue and syncope. The patient was anaemic with Hb of 4.5g/dl. After one unit PRBC and one unit FFP transfusion Hb raised to 5.5mg/dl. Oral tranexamic acid was given to control the unprovoked bleeding from nose and tongue. In this case a dramatic improvement of a patient with a high transfusion frequency due to severe recurrent anemia successively treated.

9. DISCUSSION:

Patient suffering from hereditary haemorrhagic telangiectasia (HHT). Diagnosis is based on physical examination, health history and some imaging tests like, ultrasound imaging, magnetic resonance imaging.. Tablet tranexamic acid is helpful for reducing the episodes of epistaxis. Other therapies need to add for more severe symptoms. In HHT patients with gastrointestinal bleeding, numerous endoscopic examinations or surgical resections are performed to prevent recurrent bleeding. However, these are also palliative approaches. In addition, these patients must receive erythrocyte concentrates and iron substitution. Gastrointestinal bleeding in HHT patients could be reduced by administration of an estrogen-progesterone combination. This therapy has the advantage that the gastrointestinal sections are treated, which cannot be examined endoscopically. In a study by van Cutsem et al. in several HHT patients with gastrointestinal bleeding the number of required blood transfusions could be reduced after hormone treatment.

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