



Tamoxifen: An Anti-Estrogen Effectively Controlling Osler-Weber-Rendu Disorder



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Introduction

- Osler-Weber-Rendu disease (OWRD) is an uncommon autosomal dominant disorder affecting the blood vessels throughout the body leading to vascular dysplasia. Patients have a high tendency for bleeding.
- The prognosis is overall good, as long as bleeding is adequately controlled.

Case Report

- We report the case of a 48 year-old male patient who was referred to our institution for blood transfusion incompatibility.
- History goes back to two years ago when he started complaining of severe periodic epistaxis and gingival bleeding. He received more than 20 units of packed red blood cells (PRBCs) over the past 2 years.
- Physical examination was pertinent for telangiectasia overlying the nostrils and the lips.

Diagnosis

- While taking a further detailed history, he mentioned that his sister was diagnosed with a liver hematoma secondary to an arterio-venous malformation (AVM) which was treated with embolization.
- Laboratory work-up didn't reveal any abnormality in primary nor secondary hemostasis (normal platelet count, no von Willebrand factor deficiency, normal PT/PTT/fibrinogen/factor XIII/bleeding time).
- Total body CT angiography was negative for any AVM. Consequently, the patient was diagnosed with OWRD based on epistaxis, telangiectasias and family history of a first-degree relative with OWRD. Genetic testing was not available.



Telangiectasia overlying the lips

Treatment

- He received moisturizing topical therapies to humidify the nasal mucosa. Our main aim was to control the bleeding episodes in order to defer blood transfusions.
- Thus, he was started on oral tranexamic acid (TXA) 1 gram three times per day and tamoxifen 20 mg per day. He also received regular intravenous iron.
- The bleeding episodes became less frequent. It has been 1 year without any blood transfusion.
- He is continuing TXA, tamoxifen and intravenous iron replacement regularly.

Conclusion

- Clinicians must be aware of this rare genetic disorder. Early diagnosis leads to improvement of the patients' quality of life by initiating a medical and supportive treatment.
- Tamoxifen appears to be an effective agent in minimizing the bleeding episodes due to Osler-Weber-Rendu disease.

Suggested therapeutic options for OWRD

Topical and ablative therapies for epistaxis, gastro-intestinal AVMs

Oral tranexamic acid

Systemic antiangiogenic therapies *

Tamoxifen and Raloxifene

Intravenous iron replacement

Specific therapies for pulmonary and hepatic AVMs

* Bevacizumab; Immunomodulatory imide drugs (thalidomide, lenalidomide)

References

Claire L Shovlin; Hereditary hemorrhagic telangiectasia (HHT): Evaluation and therapy for specific vascular lesions – UpToDate 2021