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內文：

Osler-Weber-Rendu syndrome (OWRS) (hereditary hemorrhagic telangiectasia)

History:

1. Described by Sutton in 1864 and Babington in 1865 as a hereditary epistaxis disease.
2. In 1896, **Rendu** described the disease as a pseudo hemophilia related to hereditary epistaxis.
3. In 1901, **Osler** described the clinical symptoms of the syndrome and emphasized its hereditary occurrence.
4. **Weber** (1907) recognized OWRS as a clinical entity distinct from hereditary hemophilia
5. Hanes (1909) named the syndrome hereditary hemorrhagic telangiectasia.

Characteristics:

1. An uncommon autosomal dominant disorder
2. An angiodysplasia in the presence of **telangiectasia of the skin and oral mucosa**
3. **Arteriovenous malformations** in the brain, lung, liver and gastrointestinal tract.
4. incidence is 1 in 5,000–10,000
5. OWRS manifests itself in 2 forms:
 - **hereditary hemorrhagic telangiectasia type 1 (HHT1)**
where there is mutation of the endoglin gene on chromosome 9 with pulmonary involvement
 - **hereditary hemorrhagic telangiectasia type 2 (HHT2)**
with a mutation in the activin receptor-like kinase-1 (ALK-1) gene. HHT2 is the milder form and its onset is later.
6. The proteins produced by the involved genes may play an important role in the integrity of the vessel wall.

Clinical characteristics:

1. epistaxis, telangiectasia of the skin and oral mucosa
2. visceral lesions (lungs, gastrointestinal tract, liver and brain)
3. Family history
4. Dentist play an important role in diagnosis of OWRS, as its first signs often appear in the oral mucosa

D.D

1. Benign liver disease, benign hereditary telangiectasia
2. CREST syndrome (calcinosis, Raynaud phenomenon, sclerodactyly and telangiectasia)
3. Ataxia-telangiectasia

Case report

1. General data: 74 year old female
2. Medical history: congestive heart failure, chronic renal failure, H/T, hypothyroidism, and rheumatism. Right lower limb had been surgically amputated because of vascular disorders
3. P.I: Frequent nosebleeds and a family history of telangiectasias and epistaxis
4. LAB test: significant changes in her red blood cell count—hypochromic anemia with anisocytosis and a high level of liver enzymes, with no changes in coagulation (Table 1). **Bleeding episodes may occur due to capillary fragility rather than disturbances in coagulation.**
5. Extraoral and intraoral clinical examination : telangiectasia were found on the skin, especially on the face (Fig. 1) and upper limbs, and were highly visible on the oral mucosa in the regions of the tongue (Fig. 2), hard palate and in the vermillion of the lip. Also present were periodontal disease and caries.

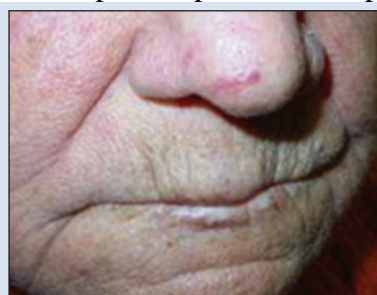


Figure 1: Telangiectasias on the face.

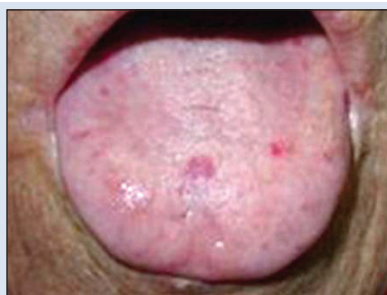


Figure 2: Telangiectasias on the tongue.

6. Tx : referred to an internal medicine specialist
Before dental treatment, **antibiotic prophylaxis** (500 mg amoxicillin) was administered every 8 hours, starting 12 hours before the procedure and continuing for 7 days after, to avoid the risk of cerebral abscesses or pulmonary infections due to the arteriovenous malformations
7. Rivero-Garvia reported the case of a 41-year-old patient with OWRS who had teeth extracted without antibiotic prophylaxis and, after a few days, developed a brain abscess.
8. use of a **vertical dental chair position** to reduce the risk of lung and nasal bleeding, measurement of blood pressure before and after the procedure, request for an up-to-date laboratory evaluation and assessment of her clinical condition at the time of treatment, because of the potential for renal failure and liver disease.

Discussion

1. Curaçao criteria:
 - I. telangiectasia on the face, hands and oral cavity
 - II. recurrent epistaxis
 - III. arteriovenous malformations with visceral involvement
 - IV. family history. (at least 3)

2. Histologically, they appear as a superficial collection of dilated blood vessels with a layer of endothelial cells in the lamina propria.
3. Electron microscope studies show a lack of perivascular elastic fibres and smooth muscle.
4. Pulmonary arteriovenous malformations occur in more than a third of patients with the disease and can cause various complications, such as hypoxia, pulmonary hemorrhage and cerebral embolism.
5. Dental professionals must be aware of these complications, keep the dental chair in a vertical position during dental treatment and be prepared to administer oxygen.
6. Cerebral abscess(5-9%): a situation that requires special care during invasive dental procedures, such as antibiotic prophylaxis, especially in infected areas.
7. patients with OWRS with severe anemia (hemoglobin level <10 mg/dL) should avoid certain routine procedures, as invasive procedures can exacerbate anemia, depending on the amount of blood that is lost.
8. Some measures, including iron supplements, blood transfusions and laser therapy, have met with varying degrees of success; sclerosing techniques have been used to control epistaxis. In patients with recurrent episodes of epistaxis, surgery of the nasal septum may be indicated.
9. A study in Italy reported excellent hemostatic results using Nd-Yag laser treatment in 8 patients with OWRS for the control of epistaxis and oral bleeding
10. The prognosis associated with OWRS is good, but the morbidity is significant. Moreover, a mortality rate of 1%–2% is reported due to complications related to epistaxis, and it rises to 10% in patients with cerebral abscess.

題號	題目
1	Hereditary hemorrhagic telangiectasia(HHT)依突變的基因不同分成兩種HHT1及HHT2,以下哪項不是HHT1的特徵呢? (A) HHT1 tend to have more pulmonary involvement (B) HHT1 generally have milder disease of later onset (C) With HHT1,numerous vascular hamartomas develop, affecting the skin and mucosa (D) Other vascular problems, like arteriovenous fistulas may also be seen
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題號	題目
2	下列關於OWRS在牙科治療方面,何者錯誤? (A) Prophylactic antibiotics before and after dental treatment are suggested. (B) use of a horizontal dental chair position to reduce the risk of lung and nasal bleeding, (C) request for an up-to-date laboratory evaluation and assessment of clinical condition at the time of treatment, because of the potential for renal failure and liver disease. (D) patients with OWRS with severe anemia (hemoglobin level <10 mg/dL) should avoid certain routine procedures, as invasive procedures can exacerbate anemia.
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