

비강내 국소 에스트로겐으로 치료한 유전성 출혈성 모세혈관확장증 1예

송정환 · 김현준 · 윤용로 · 김운태

A Case of Hereditary Hemorrhagic Telangiectasia Managed with Intranasal Topical Estrogen

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ABSTRACT

Hereditary hemorrhagic telangiectasia is an autosomal dominant disorder affecting blood vessels of the skin, mucous membrane and viscera. It is characterized by a clinical triad of multiple telangiectasia, recurrent hemorrhages and familial occurrence. In 80% of patients, epistaxis is the first presenting symptom. It can have a massive impact on the quality of life of those affected. Otolaryngologists should be familiar with this disease because of its potential systemic complication. Recently, we have experienced a 69 year old female complaining of melena and recurrent epistaxis for 20 years. Epistaxis was successfully managed with topical estrogen ointment. (*Korean J Otolaryngol* 2006;49:447-50)

KEY WORDS : Hereditary hemorrhagic telangiectasia · Epistaxis · Estrogen.

가 , 20 69

가 1

1864 Sutton²⁾ 1901 Osler³⁾

가

4)5) 80% 69 2

96% 6) , , 20 1

10

가 2 가

, 2 가

(Fig. 1).

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E - mail : entkhj@ajou.ac.kr (Fig.

2A and B).

3.4 g/dl, 가 13.0% , (Fig. 3).
2 2

(shunt) Telangiectasia, HHT)
6.3 g/dl
28.2% 가 2
5
50 cm
9.5 g/dl 31.0% 10
11 가
(Estreva Gel®)
1 가 3

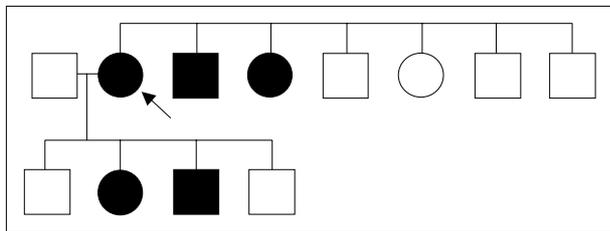


Fig. 1. Family pedigree shows two siblings and one daughter and one son had a history of recurrent epistaxis (Patient-Arrow).

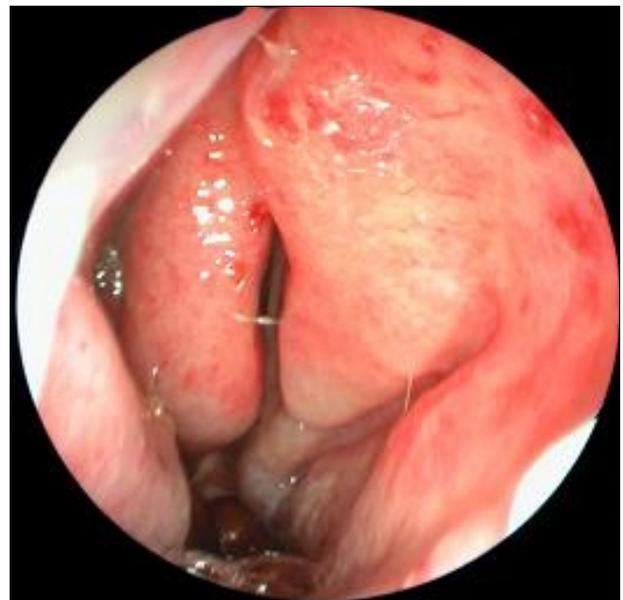


Fig. 3. Nasal endoscopic finding. Nasal mucosa after 3 months of topical estrogen treatment shows improvement of mucosal telangiectasia.

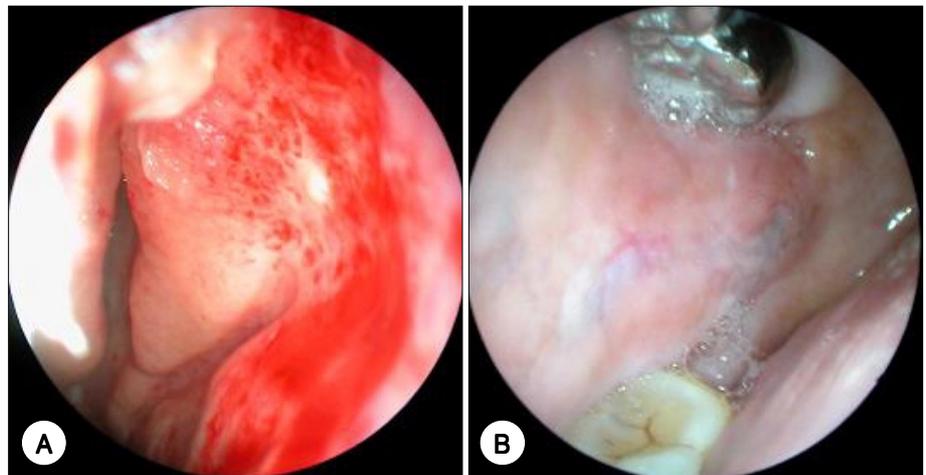


Fig. 2. A : Nasal endoscopic finding. Multiple telangiectasia was noted at lateral nasal wall of left nasal cavity. B : Oral cavity finding. Telangiectasia was noted at buccal mucosa.

가 .⁷⁾ 가 (Nasal vasculature embolization), (68~100%), (58~79%), (30~ (Laser Photocoagulation), 63%) .⁸⁾ (septodermoplasty) .
 , 96% 가 ,
 .⁶⁾ 가 ,
 , , .¹⁶⁾
 , , 9)
 .¹⁰⁾
 HHT 1909 Hanes 가 .
 , HHT 가 (septodermoplasty)
 가 .¹⁾ HHT
 Scientific Advisory 가 .⁴⁾⁵⁾ 1958
 Board of the HHT Foundation International, Inc. .¹⁷⁾
 Curacao Criteria가 (split skin
 1) , 2) , 3) (visceral graft)
 lesions), 4) 1 가 HHT 가 , 3가 27~64% ,
 HHT .¹¹⁾
 endoglin(9q33 - 34, HHT - 1) activin
 receptor - like kinase(ALK - 1, 12q, HHT - 2) .¹⁸⁾
¹²⁾
 가 (locus heteroge-
 neity) 가
 .¹⁹⁾
 .¹¹⁾ .²⁰⁾

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가 .¹³⁾
 (squamous metaplasia)
 .¹⁴⁾
 (gynecomastia)
 , (hepatic adenoma),
 (pulmonary embolism) 가 .¹³⁾¹⁵⁾
 가

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