Case Report

ORAL MANIFESTATIONS OF HEREDITARY HEMORRHAGIC TELANGIECTASIA (OSLER - WEBER - RENDU DISEASE)

* Drs. Mala S. Daxit ** Prabha Jadhav

Hereditary hemorrhagic telangiectasia is an unusual bleeding disease which is clinically characterised by numerous angiomatous, lesions (telangiectasis) hereditary incidence and hemorrhagic diathesis. This Syndrome is not a true intrinsic bleeding disorder since the hemostatic factors usually appear to be normal. Although this syndrome is not commonly diagnosed in dental practice, its recognition is important since haemorrhage from the oral mucosa can be a significant aspect of the disease and is often the first manifestation.[2]

The first documented clinical report which viewd chronic epistaxis as a disease or as a prime symptom of a disease state is that of Sutton which appeared in 1864. Babington also reported a case in 1865. Rendu, in 1896, named the disease pseudohaemophilia and believed it to be related to familial epistaxis.

Later, Osler gave a clinical description of the condition and emphasized the familial occurance. It was in 1907 that Weber recognized hereditary hemorrhagic telangiectasia, (HHT) as a distinct clinical entity and made a clear distinction between it and hereditary haemophila. The notable detailed case description of Rendu, Osler and Weber are responsible for the alternative designation of the syndrome as Osler Weber-Rendu disease.[2]

CLINICAL FINDINGS :

The lesions generally involve the skin or mucous membranes (or both) and tend to bleed spontaneously after slight trumma. Overt lesions may be found on the lips, tongue, buccal mucosa, nasal mucosa, less common locations include ears, nail-beds, scalp rare sites are the mucosa of the palate, the gingiva and the remaining oral mucosa. Other lesions may occur in the gastrointestinal tract, including the liver and in the spleen, respiratory system, brain, meninges, spinal cord and bones.[1]

Usually the telangiectasis appear during the 2nd and 3rd decade of life they are increasing frequency in the 4th decade. Rarely are they seen before puberty or in blacks. The syndrome is inherited through an autosomal dominant gene but at least 20% of the affected subjects are not aware of a familial diathesis. The disease tends to be milder in successive generation but according to F/tz-Hugh (1923) ativistic skipping is not uncommon.[4]

DIAGNOSIS

The diagnosis is essentially clinical based on the triad of telangiectatic lesions, hereditary incidence and hemorrhagic diathesis.

The lesion may be an angiomatous skin or mocous membrane lesion. Lesions pathognomic of this malady fall into 3 categories : Punctiform, spider like and nodular, nodular tumour like. It may very in colour from bright red to purple and in size from a few millimeters in diameter to 2-3- cms.

Bleeding, clotting and prothrombin times are usually within normal limits, even though one report noted a low adhesive platelet count and another study reported an increase in plasminoger activator.[3] The haemorrhagic episodes are generally short but occasionally may be massive and prolonged. The diagnosis is often easily made, but in some cases the clinical picture may be complicated because of episodes of hematemesis and meltena associated with chronic secondary anemia. In these cases, a differential with gastroduodenal ulcer or cancer of the G.I. tract must be considered. Sometimes, if the disease involves the respiratory system, the symptomatology may include hemoptysis dyspnea, cyanosis, polycythesia and finger clubbing.

In other cases, where hepatic or splenic enlargement is present or when neurological symptoms of an intra cerebtal hemorhage are manifest, the diagnosis is purely ocasional. Not uncommonly, telangiectases may appear in several other diseases and syndromes including cirrhosis of liver ataxia telangiectasia - Calcinosis - Raynand's - Sclerodactyl. Telangiectasia - syndrome and angiokeratoma Corporis diffusum Universale (fabry's disease). But in all of these cases the diagnosis is not difficult because of the unique symptoms such as the cerebellar ataxia of the ataxia telangiectasia, the systemic signs of the scleroderma typical of the calcinosis-Raynand's- Superodactyly Telangiectasia syndrome, the inherited disorder in the glycolipid metabolism of the fabry's disease or the typical multiple spider-nevi the neurological signs and the liver enlargement usually associated with cirrhosis of the liver and chronic alcoholism.

HISTOPATHOLOGY

Hanes remains the forgotten man among the chronicles of the disorder, although in 1909 he was the first to describe the histopathology of the lesion and proposed the name herditary hemorrhagic telangiectasia.

Light microscopy reveals dilated blood vessels in the upper part of the lamina propria. These vessels present a single layer of endothelial cells these may be enlaraged capillaries, venules or arteriovenular pre-capillary shunts. Such vessels studied with EM show a lack of perivascular plastic fibers and smooth muscles. Pericytes, which characterize capillaries are not present and therefore these vessels were identified as small venules. These venules present defects in the interdigitation between adjoining endothelial cells. Hashimoto found the presence of microthrombin in the gaps of the endothelial junctions and explained them as necessary for closing the endothelial junction defects. There is marked edema in the perivascular tissues with an increase in amorphous and filamentous material with fibrils showing abnormally large dimensions and peculiar banding pattern.

The perivascular connective tissue changes found around the small venules in the skin and mucous membrane of patient affected by HHT, seem to reflect the pathogenic mementum

* Lecturer in Periodontology Govt. Dental College and Hospital Bombay
** Lecturer in Community Dentistry Govt. Dental College and Hospital Bombay

of the disease. Any trauma, even if very small can easily breakdown the interendothelial junctions of the small venules and initate the formation of telangiectses.

Hereditary Hemorrhagic Telangiectasia is a degenerative disease affecting the vessel walls and not a coagulation deficiency. The disease in carried genetically and may have wide distribution in the body.

REPORT OF A CASE

This 32 year old man with good oral hygiene came with the complain of bleeding and enlarged gums in 3/45 Region. No other region was involved in the oral cavity. All areas of the skin not covered with clothing were examined and found free of lesions. The patient gave no history of oral bleeding except from the lesion, however a review of the medical history revealed episodes of epistaxis 10-12 yrs. prior to date. Patient also gave history of epistaxis in the family; his brother and mother used to occasionally get epistaxsis.

On oral examination an ulcerated granular lesion was seen surrounded by an erythematous halo on the bucal mucosa and attached gingiva. The patient had first noted this lesion as a small swelling which gradually and asymptomatically enlarged to involve 31/45 region to a size of 13 x 3 x 3 mm in the last 9 months. The laboratory investigation of the patient's blood were all within the normal range. A gingival biopsy was taken to be examined under light microscrope, where it was diagnosed as Telangiectatic granuloma.

An intra oral periapical radiographic picture of the region showed severe angular bone loss with only apical 3rd bone support left. The teeth showed two degree mobility and 43 was distally migrated probably due to pressure from the enlargement. The occlusion was traumatic in this region.

Not associated with various of the large veins or with hemorrhages or symptoms it had gradually extended over the span of months. It had presisted indefinitely, showed no spontaneous resolution, did not affect the health, nor reflected any known internal disease.

Our ignorance of the pathogenesis of this vascular change has been essentially matched by our ignorance of what to do for it. Indeed, there has been a striking therapeutic silence in the literature on this unusual problem.

Some authors believed that, the excessive bleeding noted on mild trauma was due to increase in plasmogen Activator which causes fibrinolysis. Hence inhibitors of fibrinoloysis like aminocaprioic acid was used as local application. Also chemical agents viz. chromic acid, trichloroacetic acid and solid carben dioxide was used with little success.

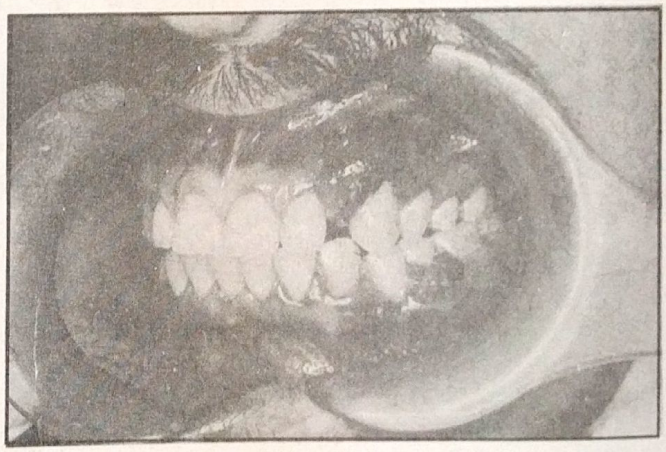
Through scaling and root planning was performed with subgingival curettage under antibiotic cover. It was advised tetrarcycline 250 mg. Q.I.D. dose for 3 weeks. This line of treatment was based on the treatment given to tetracycline responsive rosacea type of telangiectasia due to its vacular resembalance.[7]

The mechanism of action is obsure since tetracycline may not be achieving its effect as a wide-spectrum antibiotic under these circumstances, but rather as a pharmacodynamic agent. In this regard Mill has pointed out that tetracycline do inhibit oxidative phosphorylyation. Certainly the dramatic action of tetratycline in cleaning the lesion has been felt to be other than simple reduction of bacterial flora of the pocket.[7]

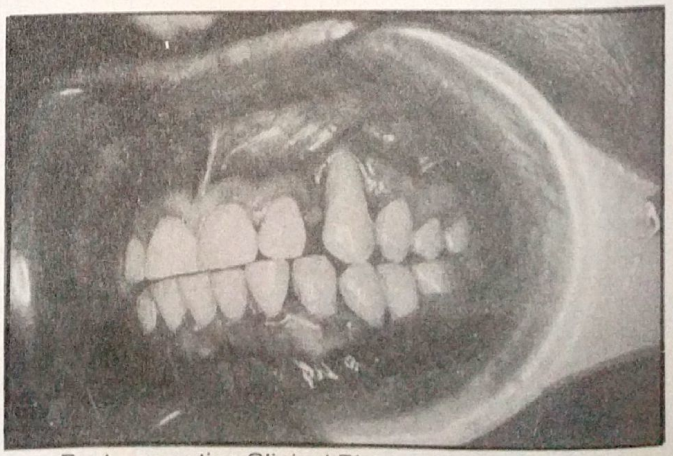
It is evident from this and other reports that tetracycline deserves a trial in treating obscure disease of the microvasculature.

BIBLOGRAPHY

1. Dorocher R.T Morris Ac & Burket L.W : Oral manifestation of HHT. Oral Surg. 14, 550, 1961.
2. Enrico G. Bartolucci, Richard. H. Swan, William C. Hort Oral manifestation of HHT (osler, weber, Rendu Disease Review and case reports. J. Periodontol 53 : 165, 1982.
3. Everlt F.G. & Hahn C.R. : HHT with gingival lesion. Review and case report J. Periodontol 47 : 295, 1976.
4. Killey H.C. Kay L.W : HHT Brit. J. Oral Surgery 7:161, 1970.
5. Scopp I.W & Quart A : HHT involving the oral cavity Oral Surg 11 : 1138 1958.
6. Soudah H.P. & Tilson H.B. HHT J. Oral. Surg. 79 : 225, 1971.
7. Walter B. Shelley. Essential progressive Telangiatasia Successful treatment with tetracycline J. Am. Med. Ass 216 1343, 1971.



Pre - Operative Clinical Picture of the Lesion



Post - operative Clinical Picture after one week.