CASE REPORT

β – Thalassaemia: A case report of multiple pyogenic granulomas in thalassemic major patients

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Abstract:
Thalassaemia has become one of the common genetic disorder worldwide with high frequency of oral manifestations. It is an autosomal single gene recessive blood disease. The prominent Oral and maxillo facial features include frontal bossing which is due to bone marrow hyperplasia. Flaring of the nose is due to air hunger, prominence of malar bones, maxillary hyperplasia and mal-aligned and mobile teeth. In previous studies, no case of pyogenic granuloma was reported in thalassaemia patients.
We are reporting here a Beta thalassemia patient with recurrence of multiple pyogenic granulomas after 1 year of excision. Patient also had multiple mobile teeth due to bad oral hygiene.

Keywords: β – Thalassaemia, frontal bossing , prominence of malar bones, multiple pyogenic granulomas

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Introduction:
Thalassaemia is a blood disease inherited as autosomal single gene characterized by reduced or absent amount of hemoglobin chains.[3] It is considered to be the most common genetic disorder with defect in either alpha and beta chains which produce abnormal blood cells.[2] There are two main types of thalassaemia; Alpha thalassemia (reduced alpha chain synthesis) and Beta thalassaemia (reduced Beta chain synthesis).

Beta-thalassaemia: In homozygous type there is little or no beta chain production resulting in excess alpha chain leading to increased HBA2 and HBF. There are three main types:

A: Beta thalassemia minor: A symptomatic heterozygous carrier state in which anaemia is mild to absent with low MCV and MCH and iron stores and serum ferritin are normal.

B: Beta thalassemia intermedia: In this type of thalassemia there is moderate anemia (hemoglobin 7 to 10gm/dl) that does not require regular blood transfusion with splenomegaly, bone deformities and recurrent leg ulcers and gall stones present as other features.

C: Beta thalassemia major: This is homozygous beta thalassemia present in first year of life with severe anemia also known as Cooley's anemia. The affected patient failed to thrive with recurrent infections. Clinical features include bony abnormalities and enlarged maxilla and prominent frontal and parietal bones.

In beta thalassaemia of homozygous type, blood film shows hypochromic microcytic anemia with raised reticulocyte and nucleated red cells. This can be confirmed by hemoglobinelectrophoresis in which there is increase in HBF and absent or reduced Hb A.

Alpha-thalassemia: In this type there are four alpha globin chains in which there is mild anemia with microcytosis to a severe condition can be seen in the most severe formed there is complete absent of alpha globin chains (HB Bart's) infants are still borne (hydropsfetalis).

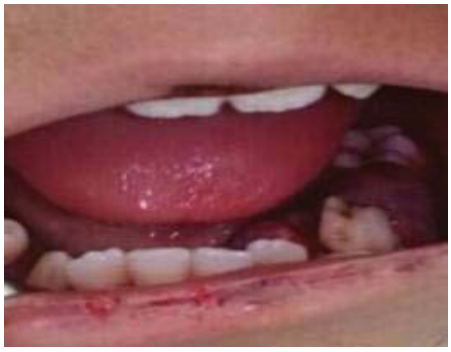

Figure-1: extra oral picture showing cracked lips and granuloma on the left posterior molar area

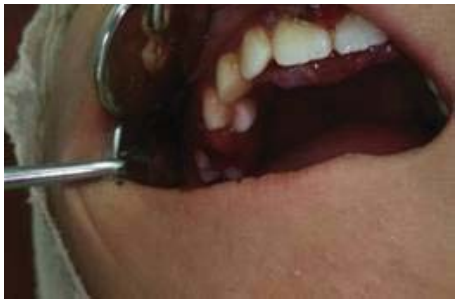

Figure-2: Intra oral picture showing swelling gums

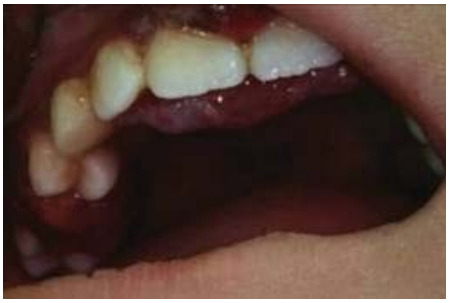

Figure-3: : Pyogenic granuloma on the palatal side and right upper posterior molar region

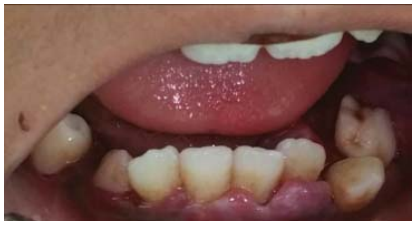

Figure-4: : intra oral picture of the patient showing mobile anterior teeth and generalized malalignment of teeth

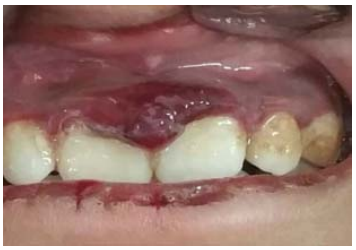

Figure-5: Classical picture of pyogenic granuloma on the upper anterior area

Case report:

A 10-years-old-female patient was referred to oral medicine department, Sindh Institute of Oral Health and Sciences, Jinnah Sindh Medical University, with a chief complaint of swelling in gums(figure-2) in relation to upper & lower anterior teeth and on posterior lower left and upper right region(figure-3) since15 to 20 days. It revealed that patient had similar swelling once before, occurring one year back which were excised. The swellings increased gradually with time. The swelling was firm in consistency, smooth lobulated exophytic lesion manifesting as small, redery the matous papule on pedunculated base. The growth cover 2/3rd of the crown. Bright red bleeding on probing was observed.

Patient was known case of thalassaemia major referred from JMPC OPD. Patient was lethargic, anemic, having fever with cracked lips which were easy to bleed (figure-1). Mobile lower left premolar tooth with swelling gums (figure-2) and mobile lower anterior teeth with pyogenic granuloma. Patient was advised an orthopantomograph and APTT, PT test prior to excision but because of known affordability, patient didn't come back for treatment.

Discussion:

Thalassaemia is one of the commonest blood diseases that can be easily transmitted through autosomal recessive genes. In thalassemic patients there is more likely history of inter-marriages.[3] Majority of thalassaemic minor patients have folic acid and iron deficiency anemia. The color of the skin changes to dark brown or muddy yellow[10] due to the iron deposition in sub-cutaneous tissue caused by excessive break down of abnormal erythrocytes, repeated transfusions, excessive iron absorption from the gut.

Bone marrow hyperplasia due to underlying anemia causes frontal bossing by enlarging the flat bones of skull including the frontal bone. Flaring of the nose is due to air hunger caused by chronic anemia. The forceful breathing gives rise to flaring of ala of nose accentuated by the depression of bridge of the nose. Prominence of malar bones was due to hyperplasia of bone marrow.[3]

Maxillary hyperplasia was due to over growth of its marrow.[3] Incompetency of the lips is due to proclined maxillary anterior incisors and prevents complete closure of lips leads to mouth breathing. Marginal gingivitis and recession could be due to maxillary hyperplasia.

Many of the features listed are seen in above case as patient was lethargic and pallor having fever with cracked lips which were easy to bleed on palpation with mobile and mal-aligned teeth. (figure-4).

Pyogenic granuloma is a soft tissue tumor arising as a result of inflammatory hyperplasia seen

in the oral cavity.[1] This lesion arises in response to various stimuli such as local irritation, traumatic injury or hormonal factors.[5] It occurs in the second decade of life[6] in young females possibly because of vascular effects of female hormones.[1] The possible site was the gingival (44%).[4] The other sites were lower lip, tongue, buccal mucosa, upper lip and palate.[5] Upper labial mucosa is the commonest site[4] (figure-5). In oral cavity, pyogenic granuloma with interdental papillae being the commonest site in 70% cases.[7] Clinically, pyogenic granuloma appears as a hemorrhagic smooth, exophytic growth having lobulated small red erythematous papules on a pedunculated base. The surface ranges from pink to red, depending upon the age of the lesion.[1]

All multiple pyogenic granulomas must be confirmed by taking biopsies to rule out more serious conditions. usually the treatment of pyogenic granuloma was conservative surgical excision with relatively high rate of recurrence(15%).[7] To avoid recurrence, lesion must be excised down with the periosteum and all the irritants must be removed.[5]

Multiple pyogenic granulomas were clinically diagnosed in upper and lower anterior teeth which had firm consistency with swelling and bleeding on probing. Scaling and excision of growth were planned. Tests were advised to check for the proneness of the patient to bleeding so that she could be managed at chair side. Patient didn't come for treatment because of non-affordability.

Conclusion:
Thalassaemia has become the commonest blood disease now-a-day. It was noted that many patients were as a result of inter-marriages between the carriers. They should be aware of the possible oral manifestations and complications that may arise as a result of disease process.

Current case is clinically diagnosed as pyogenic granuloma having thalassaemic major. In previous studies pyogenic granuloma was never reported in thalassemic patients.

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Role and contribution of authors:
Dr. Maria Naz, collected the data and references and wrote the initial write-up

Dr. Sanaa Ahmed, helped in collecting the data and references

Prof. Dr. Syed Mahmood Haider, critically review the article and made the final changes

Dr. Aisha Naureen, analysed the data and helped in discussion writing.

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