Otorhinological Manifestation in Patients with Thalassemia Major

Qays Jaafar Khalaf[1], Omar Malik Bargas[2]

Department of Otolaryngology, College of Medicine, [1]Diyala University, [2]University of Anbar, Iraq.

Original Article

ABSTRACT

Aim: This study was designed and intended to reveal the incidence of certain otorhinological presentations in patients with thalassemia major and the relation of desferrioxamine with it.

Patients and Methods: In our cross-sectional study we blindly select 100 patient complaining of thalassemia major whom attending Baquba teaching hospital for blood transfusion as part of their regular treatment. Data collection was done in the otolaryngology outpatient clinic of the hospital.

In those patients, thalassemia major was documented by hemoglobin electrophoresis. History and examination in those patients were concentrated on nasal problems like nasal airway obstruction, temporomandibular joint complaints like pain in TMJ region, ear problems like deafness, tinnitus and morphological abnormalities like saddle nose. Clinical examination and pure tone audiometry were done.

Results: The relative frequency of certain otorhinological manifestations is high in patients with thalassemia major.

Conclusion: Thalassemia major may be associated with certain otorhinological problems which can be early diagnosed and treated.

Key Words: Desferrioxamine, hearing loss, saddle nose, thalassemia major.

Received: 10 December 2019, **Accepted:** 19 April 2020.

Corresponding Author: Qays Jaafar Khalaf, MSc, Department of Otolaryngology Head And Neck Surgery, University of Diyala, Iraq, **Tel.:** 009647707720200, **E-mail:** qaise2014@yahoo.com

ISSN: 2090-0740, November 2020 Vol.21, No.3

INTRODUCTION

Thalassemia is an autosomal recessive inherited disorder in which there is reduction in production of the normal globin chain (alpha and beta) which may be manifested by a different grades of hypochromic microcytic anemia.the affected children may be asymptomatic at birth with exception of those with thalassemia major who develop severe anemia and transfused early in their life. Two main variants are present alpha and beta-thalassemia according to globin chain defect. Signs and symptoms of thalassemia usually start after the age of 6 months when there is a shift in HbF to HbA and the patients become complaining of anemia, jaundice, hepatosplenomegaly, failure to thrive and bony deformities. because the marrow hyperplasia secondary to anemia facial bone expansion especially cheek one leads to obliterate the nose base and upper teeth exposure with malocclusion. Thickening of cranial bones may lead to frontal bossing, pneumatization of sinuses is delayed especially with maxillary expansion. Bleeding tendency secondary to splenomegaly may be represented by epistaxis. Extramedullary marrow growth in middle ear bones is reported. High-frequency SNHL is detected in a high percentage in those patients receiving desferrioxamine. All these changes may be reduced or prevented during transfusion therapy where a reduction in desferrioxamine dose may be of great benefit for complaining patients.

PATIENTS AND METHODS:

In our cross-sectional study, 100 patients attend Baquba teaching hospital every month for blood transfusion.

In those patients, thalassemia was documented by hemoglobin electrophoresis. History and examination were concentrated on nasal airway obstruction, temporomandibular joint complains, deafness, tinnitus and saddle nose. Pure tone audiometry also was done.

Data Analysis:

Data was collected after questionnaire completion and checkup for any error or discrepancy. Data was entered on Microsoft excel worksheet and exported to statistical software and analyzed using statistical tests by using statistical packages.

Ethical considerations:

Approval was obtained from the patients by taking consent. The approval was on the agreement that patient

 DOI: 10.21608/ejentas.2020.17290.1151

namelessness should be maintained, smart laboratory follows, internal control ensured, which each finding would be treated with most confidentiality and for this analysis solely.

RESULTS:

We took 100 patients with thalassemia major with age between 10 and 40 years, 52% female and 48% male.

Table 1: Distribution of the patients with thalassemia major according to age and sex:

Age	Number	Male	Female	Percentage
10-20 years	74	32	40	74%
20-30years	18	12	6	18%
30-40years	8	4	6	8%
	100%	48%	52%	100%

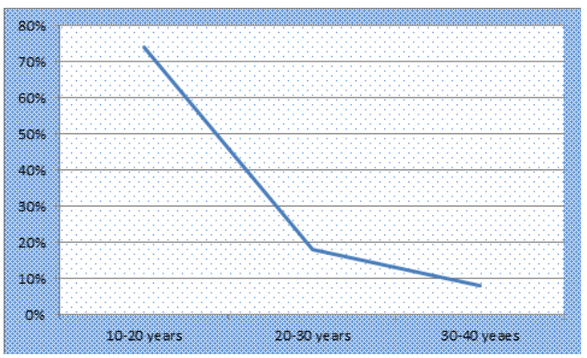

Chart 1: Distribution of patients with thalassemia major according to age:

Table 2: The relative frequency of ear problems and PTA in thalassemia patients:

symptoms	Number	Percentage
Tinnitus	40	40%
Temporo-mandibular joint pain	18	18%
Hearing loss (PTA)	8	8%

Table 3: The relative frequency of nose problems in thalassemia major patients

complain	Number	Percentage
Saddle nose	40	40%
Nasal obstruction	32	32%

DISCUSSION

Thalassemia is an inherited disorder in which there is a decrease in the production of normal globin chains. Desferrioxamine (DFO) is a commonly used Iron chelator which is important in reducing Iron overload in thalassemic patients, but not with adverse effects, DFO is implicated in most of the ear symptoms like sensory neural hearing loss and tinnitus and most these side effects can be avoided by drug monitoring. The amount of DFO and the therapeutic index are important factors in determining the toxic effects. The accepted dose is less than 40 mg/kg/day, and TI less than 0.025. In this study, (52%) of the patients were females and 48% of patients were males with age range (10-40) years (Table 1) and this prevalence is very close to the values reported in the literature, also the incidence of TMJ pain occurs in 18% of patients with thalassemia major and increases with aging, (Table 2) this agrees with (Berjis N)[1]. Who found 19.1% of patients had TMJ pain. This occurs due to growth disturbance and bony deformities (deformed facial bones, pathological fractures). Incidence of tinnitus occurs in 40% of patients with thalassemia major (Table 2). No previous identical study explains the relationship between the tinnitus and thalassemia major but the temporomandibular disorders (TMD) which include pain, impaired jaw function, malocclusion that occurs in thalassemia patients may explain the tinnitus also the SNHL which occur in thalassemia may explain the tinnitus. (Womack)[2] found 42% of patients with TMD reported tinnitus, also (Tuz HH)[3] show 59.1%, of patients with TMD, reported tinnitus. The percentage Hearing loss in our study was 8% This agrees with (Kontzoglou)[4] who show 20% of patients had hearing loss. but disagréé with (Karimi)[5] who found 44% of patients exhibit hearing loss.

The type of hearing loss in our study was high-frequency SNHL (Figure 1) which agrees with Kontzoglou and karimi result. Also, this finding of the type of hearing loss is compatible with what (V Wong)[6] and (H Kanno)[7] found in their study. SNHL is highly linked to DFO treatment and the thromboembolic complication of thalassemia no attacks of vertigo were well explained in our patients and no attacks of otorrhea seems to be related to the thalassemia itself.

The incidence of saddle nose in our study 40% (Table 3) this agree relatively with (Hattab)[8] who take a sample consisted of 54 patients with thalassemia major and found More than half of the patients exhibited frontal bossing and saddle nose.

Regarding Nasal obstruction, it occurs in 32% of patients with thalassemia major (Table 3) which may differ from the findings of (Ragab)[9] who found 70% of patients exhibit nasal obstructions this difference in findings may be due to variation in numbers of patient and parameter used in these studies. Nasal obstruction in our study may be explained by the maxillofacial deformities especially saddle nose in which lose of septal support will affect the airflow through the internal nasal valve in addition to the collapse in the upper and lower lateral cartilage as a result of long term damage to the nasal septal support one other things identified which may be implicated in the sense of nasal obstruction is the dryness of nose which is identified in most of those patients this dryness mostly related to the turbulence of the airflow at the entrance to the nasal cavity as a result of the bony deformities or due to the paranasal sinus poor pneumatization which is a well defined finding in thalassemia as documented by (Smithson)[10]. Regarding other nasal complaints in those patients they deny any smell problems or Annoying snoring with no recorded attacks of a truly diagnosed sinusitis, the majority (90%) of those patients not reporting attacks of epistaxis needs medical care with only 10 patients reporting blood-stained nasal discharge or blood streaks on their handkerchief which is inconsistent with (Eldor A)[11] who mention in his study that there are frequent attacks of epistaxis in thalassemic patients without documentation of the exact percentage. Bleeding tendency may be explained by hypersplenism in which there is pooling of platelet in the spleen resulting in thrombocytopenia or due to decrease in platelet aggregation to ADP, collagen, ristocetin, and epinephrine as (Eldor A) mention in his study and due to lack of the availability of special laboratories to measure the platelet aggregation and due to normal bleeding time test in those patients no further discussion or explanation is done in this study which in turn may require further study in future. Most of the patients refuse to do a CT scan of nose and paranasal sinuses due to being afraid of radiation exposure especially no one of them thinking of doing elective nasal surgery. For those who agree to do CT scan the findings was only mucosal thickening in paranasal sinuses with no significant hypopneumatization in PNS which may be due to either small number of patients who agree to do CT or due to absence of a strict parameters that diagnose it due to normal variation. (Figure 2).

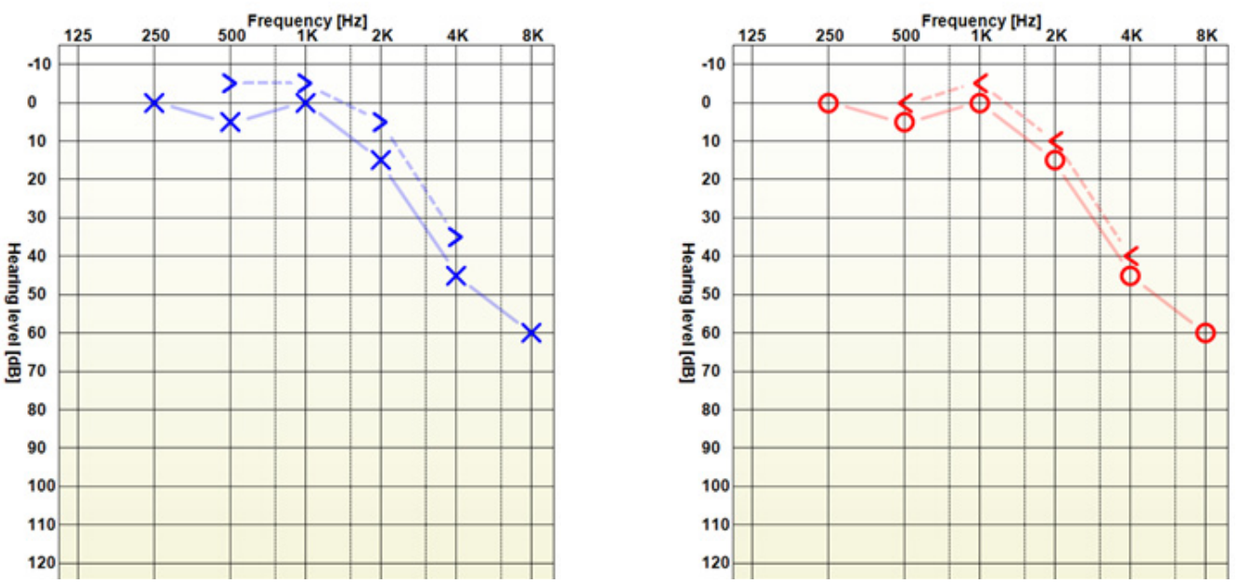

Fig. 1: Audiogram of 22- years old male with bilateral high frequency SNHL on daily dose DFO 45 mg /kg x 5 days / week.

Fig. 2: coronal, axial and sagittal section CT scan of nose and paranasal sinuses in 6 years old boy with thalassemia showing mucosal thickening in sphenoid sinus and Rt. maxillary sinus.

CONCLUSION

From the previous numbers, we can see that certain otorhinological manifestations are high in thalassemic patients like tinnitus, saddle nose and nasal obstruction these findings can be prevented or at least ameliorated if desferrioxamine dose can be closely monitored and followed. Further studies are advised to try to explain the exact underlying pathophysiology of certain complaints like tinnitus.

CONFLICT OF INTEREST

There are no conflicts of interest.

REFERENCES

1. Berjis N, Sonbolestan SM, Nemati S, Mokhtarinejad F, Danesh Z, Abdeyazdan Z. Otorhinolaryngologic manifestations in thalassemia major patients. 2007;
2. Womack A, SMT C, Osteopractic D, Butts R. Somatosensory Tinnitus: Neurophysiological Considerations & Treatment Options.
3. Tuz HH, Onder EM, Kisnisci RS. Prevalence of otologic complaints in patients with temporomandibular disorder. Am J Orthod Dentofac Orthop. 2003;123(6):620–3.
4. Kontzoglou G, Koussi A, Economou M, Tsatra I, Perifanis V, Noussios G, *et al*. Long term audiological evaluation of beta-thalassemic patients. Acta Otorhinolaryngol Belg. 2004;58(2):113–7.
5. Karimi M, Asadi-Pooya AA, Khademi B, Asadi-Pooya K, Yarmohammadi H. Evaluation of the incidence of sensorineural hearing loss in beta-thalassemia major patients under regular chelation therapy with desferrioxamine. Acta Haematol. 2002;108(2):79–83.
6. Wong V, Li A, Lee ACW. Neurophysiologic study of β-thalassemia patients. J Child Neurol. 1993;8(4): 330–5.
7. Kanno H, Yamanobe S, Rybak LP. The ototoxicity of deferoxamine mesylate. Am J Otolaryngol. 1995;16(3):148–52.
8. Hattab FN. Periodontal condition and orofacial changes in patients with thalassemia major: a clinical and radiographic overview. J Clin Pediatr Dent. 2012;36(3):301.
9. Ragab A, Ragab SM, Shawki M. Impact of beta-thalassemia on maxillary sinuses and sino-nasal passages: A case-control study. Int J Pediatr Otorhinolaryngol. 2015;79(12):2253–9.

10. Smithson L V, Lipper MH, Hall JA. Paranasal sinus involvement in thalassemia major: CT demonstration. Am J Neuroradiol. 1987;8(3):564–5.

11. Eldor A. Hemorrhagic Tendency in β-Thalassemia Major. In: Pathophysiology of Blood Disorders. Karger Publishers; 1979. p. 32–4.