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RESEARCH ARTICLE

HYPOPARATHYROIDISM – A RARE ENDOCRINOPATHY IN BETA THALASSEMIA MAJOR

Ajay Raj Mallela, *Hariprasad Shetty, Rohini Koya, Ganesh Shetty and Shastry Ba

Department of Medicine, Kasturba Medical College, Manipal, Karnataka, India

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ABSTRACT

Hypoparathyroidism is rare atypical clinical consequence of iron overload in patients of Beta thalassemia major (BTM) who are on regular blood transfusions. We report a case of 22 yr old male who presented with generalized tonic clonic seizures. Laboratory investigations showed anemia, hypocalcemia, hypophosphatemia with very low serum parathyroid hormone (PTH) level. Patient was started on calcium and vitamin D supplementation.Serum calcium levels were optimized. The patient gradually improved with no further episodes of seizure. Rarity of this endocrinopathy in beta thalasemia major made us report this case.

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INTRODUCTION

Beta thalassemia major (BTM) is hereditary hemoglobin disorder due to deficiency of beta globin synthesis. In homozygous state it is known to cause severe anemia. Regular blood transfusions and chelation therapy prolong the survival in thalassemia patients. Endocrine abnormalities such as hypogonadotrophic hypogonadism, diabetes mellitus, hypothyroidism and are known to occur despite chelation therapy (Angelopoulos *et al*., 2006). The parathyroid gland involvement is very rare in thalassemic patients. Hypoparathyroidism (HPT) is not usually monitored due to unawareness of this rare clinical sequelae. The intention to report this case was to elucidate the significance of monitoring serum calcium levels and early detection of hypoparathyroidism among thalassemia patients.

Case presentation

A 22 year old male patient diagnosed case of BTM on regular chelation therapy came with recurrent episodes of generalized tonic clonic seizures. On examination the patient had prominent malar eminences with frontal bossing (Figure 1), significant pallor and was hemodynamically stable. Central nervous system examination showed worsening of sensorium with Glasgow coma score of 11/15 with pupils bilaterally equal and reacting to light, intact cranial nerves, no obvious motor deficits and normal deep tendon reflexes with bilateral flexor plantar response. The abdomen examination showed a splenectomy scar and gross hepatomegaly with a liver span of 26cm. Other system examination was within normal limits. Diagnosis of BTM was confirmed at the age of 6 months by high performance liquid chromatography (HPLC) of hemoglobin which showed absent HbA and HbF being 90%. He received more than 140 units of packed red blood cells transfusion since the time of the diagnosis. Complete blood picture showed anemia with blood hemoglobin level of 8.0gm/dl (13-17gm/dl), Biochemical investigations including blood sugars, renal function and liver function tests were within normal limits. Structural causes were ruled out with computed tomography (CT) of brain which was essentially normal. The patient was found to have hypocalcemia with serum calcium of 5.2 mg/dl (8.9 – 10.3mg/dl), hyperphosphatemia with serum phosphorus of 7.5mg/dl (2.4- 4.7mg/dl).Serum parathyroid hormone level (PTH) was 4.21pg/ml (15-65pg/ml), serum ferritin level was 2034 ng/dl (23.9-336ng/dl).Other metabolic and infective workup for seizures was unremarkable. The patient was hence diagnosed to have hypocalcemia secondary to hypoparathyroidism. Once the diagnosis was sought the patient was immediately started on intravenous calcium gluconate 30ml (elemental calcium of

Corresponding author: Hariprasad Shetty,
Department of Medicine, Kasturba Medical College, Manipal, Karnataka, India.

300mg) with 100ml of 5% dextrose over 10 minutes, calcium gluconate infusion of 1mg/kg/dl was continued. ECG monitoring was done during calcium infusion and also started on oral calcium and vitamin D supplementation. The patient gradually improved with no further episodes of seizures.

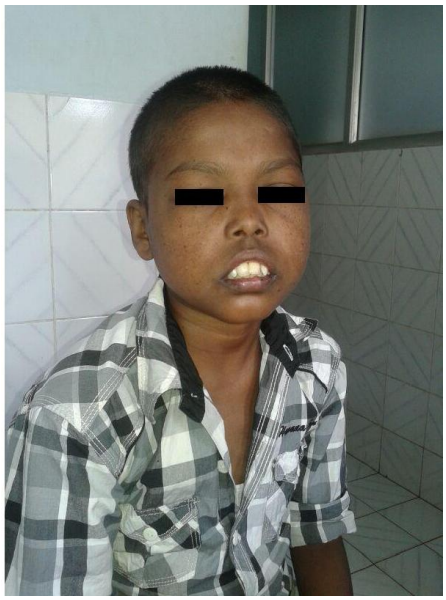

Figure 1. Facial features of beta thalassemia showing prominent malar eminences with frontal bossing

DISCUSSION

In Transfusion dependent patients of BTM, adverse sequelae of iron overload are hypogonadism (35-55%), hypothyroidism (9-11%), diabetes (6-10%), liver fibrosis and heart dysfunction (33%) (Cunningham *et al*., 2004; Borgna-Pignatti *et al*., 2004). HPT is an extremely rare complication which is reported to be less than 4 % among the thalassemic patients (De Sanctis *et al*., 1992). Most of them are asymptomatic and only few of them manifest with neurological symptoms like seizures, carpopedal spasms and paraesthesias (De Sanctis *et al*., 1992; Zafeiriou *et al*., 2001). The degree of iron overload, as reflected by serum ferritin levels is not associated with development of endocrine complications (Angelopoulos *et al*., 2006; Cario *et al*., 2003). Development of endocrine complications depends on long term iron status rather than present iron status. The two factors resulting to HPT in BTM are increased iron deposition in parathyroid gland leading to its dysfunction and other one being, suppression of PTH production by bone resorption secondary to expansion of marrow due to chronic hemolytic anemia (Zafeiriou *et al*., 2001; Shamshirsaz *et al*., 2003; Dhouib *et al*., 2011). Nicholas G *et al* and Shamshirsaz AA *et al* reported that development of HPT does not depend upon age of initiation of blood transfusion therapy or chelation therapy and as well as the delay in chelation therapy (Angelopoulos *et al*., 2006; Shamshirsaz *et al*., 2003). Hypocalcemia is also associated with impaired glucose tolerance as calcium is related with release of insulin from pancreas (Ismail and Namala, 2000). Several studies showed that hypocalcemia can impair myocardial performance and most of them present with diastolic heart failure (Altunbas *et al*., 2003). The milder form of hypocalcemia is managed with calcitriol, however patient manifesting with severe hypocalcemia requires intravenous calcium adminstration, followed by oral calcium and vitamin D supplementation. Role of early chelation therapy is doubtful however it might have delayed the onset of endocrine complications. So further studies are required to understand the etiology of endocrinopathies and better treatment options for improved outcomes.

Conclusion

We report a case of recurrent seizures due to hypocalcemia secondary to hypoparathyroidism. This report highlights the importance of monitoring serum calcium and serum PTH levels in all patients of BTM as serum ferritin is considered as inaccurate marker of iron overload. Initiation of chelation therapy at an early age might be helpful in patients who are on regular blood transfusions however it's not proven.

References

Altunbas, H., Balci, M.K., Yazicioglu, G., Semiz, E., Ozbilim, G., Karayalcin, 2003. Hypocalcemic cardiomyopathy due to untreated hypoparathyroidism. *Horm Res.,* 59:201–204.

Angelopoulos, N.G., Goula, A., Rombopoulos, G. *et al*. 2006. Hypoparathyroidism in Transfusion- dependent Patients with Beta-Thalassemia. *J Bone Miner Metab.,* 24(2): 138-45.

Borgna-Pignatti, C., Rugolotto, S., De Stefano, P., Zhao, H., Cappellini, M.D., Del Vecchio, G.C., Romeo, M.A., Forni, G.L., Gamberini, M.R., Ghilardi, R., Piga, A. and Cnaan, A. 2004. Survival and complications in patients with thalassemia major treated with transfusion and deferoxamine. *Haematologica*, 89:1187-1193.

Cario, H., Holl, R., Debatin, K. and Kohne, E. 2003. Insulin sensitivity and betacell secretion in thalassaemia major with secondary haemochromatosis: assessment by oral glucose tolerance test. *Eur J Pediatr.,* 162:139–46.

Cunningham, M.J., Macklin, E.A., Neufeld, E.J., Cohen, A.R. 2004. Thalassemia Clinical Research Network. Complications of beta-thalassemia major in North America. *Blood*, 104:34-39.

De Sanctis, V., Vullo, C., Bagni, B. *et al.* 1992. Hypoparathyroidism in beta-Thalassemia Major. Clinical and Laboratory Observations in 24 Patients. *Acta Haematol*., 88(2-3): 105-8.

Dhouib, N., Turki, Z., Mellouli, F., Ouederni, M., Yahiaoui, S., Nagi, S., Kouki, R., Ben Slama, C. and Bejaoui, M. 2011. Hypocalcaemia due to hypoparathyroidism in β-thalassemia major. A study of a new case. *Tunis Med.* 2011 Mar;89 (3):302-4.

Ismail, A. and Namala, R. 2000. Impaired glucose tolerance in vitaminD deficiency can be corrected by calcium. *J Nutr Biochem*., 11:170–175.

Shamshirsaz, A.A., Bekheirnia, M.R., Kamgar, M. *et al.* 2003. Metabolic and Endocrine complications in beta-Thalassemia Major: a Multicenter Study in Tehran. *BMC Endocr Disord.,* Aug 12; 3(1): 4.

Zafeiriou, D.I., Athanasiou, M., Katzos, G. *et al.* 2001. Hypoparathyroidism and intracranial calcifications in ß-thalassemia major, *J Pediatr.,* 138:411.

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