Online Journal of Health and Allied Sciences
Peer Reviewed, Open Access, Free Online Journal
Published Quarterly : Mangalore, South India : ISSN 0972-5997
Volume 12, Issue 1; Jan-Mar 2013

Case Report:

Pseudohypoparathyroidism Presenting as Congestive Cardiac Failure.

Authors
Madan Lal Kaushik, Department of Medicine,
Sanjay Mahajan, Department of Medicine,
Rajiv Raina, Department of Medicine,
Bhupender Kumar, Department of Medicine,
Ashwani Tomar, Department of Radiodiagnosis,
Rajeev Marwah, Department of Cardiology,
IG Medical College, Shimla-171 001, Himachal Pradesh, India.

Address for Correspondence
Dr. Madan Lal Kaushik,
Assistant Professor,
Department of Medicine,
IG Medical College,
Shimla-171 001,
Himachal Pradesh, India.
E-mail: madankaushik2002@yahoo.com

Citation
Kaushik ML, Mahajan S, Raina R, Kumar B, Tomar A, Marwah R. Pseudohypoparathyroidism Presenting as Congestive Cardiac Failure. *Online J Health Allied Scs.* 2013;12(1):7. Available at URL: http://www.ojhas.org/issue45/2013-1-7.html

Open Access Archives
http://cogprints.org/view/subjects/OJHAS.html
http://openmed.nic.in/view/subjects/ojhas.html

Submitted: Jan 5, 2013; Accepted: Apr 3, 2013; Published: Apr 15, 2013

Abstract: Hypocalcaemia can have a variety of manifestations including cardiovascular changes. Hypocalcemic cardiomyopathy has been reported in hypoparathyroidism and vitamin D deficiency but association of hypocalcemic cardiomyopathy in pseudohypo-parathyroidism has been reported scarcely in literature. We describe here a case of PHP presenting as congestive cardiac failure.

Key Words: Pseudohypoparathyroidism; Hypocalcaemia; Cardiomyopathy; Vitamin D

Introduction:

Plasma calcium level may be decreased due to insufficient Parathormone production by the parathyroids (hypoparathyroidism), or a resistance against its action in target tissues (pseudohypoparathyroidism). In both cases, there are significant reduced levels of plasma calcium associated with hyperphosphatemia. Hypocalcaemia can have a variety of manifestations including cardiovascular changes.[1]

We describe here a case of Pseudohypoparathyroidism (PHP) presenting as congestive cardiac failure.

Case Report

A 24 years old female presented with insidious and gradually progressive shortness of breath for 28 days with orthopnea and paroxysmal nocturnal dyspnoea for last few days and breathlessness even at rest for 1 day. She was taking carbamazepine for seizure control for last eight years. She had developmental delay with poor scholastic performance at school. She was married and had a 2 years male healthy child. Her both parents and younger sister were healthy but her younger brother was a diagnosed patient of pseudohypoparathyroidism.

On general physical examination her respiratory rate was 30/min., pulse rate was 100/min., BP 100/70 mm Hg. and she had cyanosis. She developed severe carpopedal spasm during examination (Figure 1) and attendant told that she was taking carbamazepine for these episodes. Chvostek's sign was also present. There was LV S3 gallop and bilateral basal crepitations. Her Mini mental score was 15/30 and deep tendons jerks were brisk. On investigation, X-ray chest showed bilateral non homogenous opacity in middle and lower zones. Echocardiography showed LV enlargement with severe LV systolic dysfunction (EF = 22 %) (Figure 2). Her serum Ca^{++} was 4.40 mg/dl (Normal range 8.7–10.2 mg/dL), Phosphorus 5.6 mg/dl (Normal range 2.5–4.3 mg/dL with marked elevation of serum PTH 201.30 pg/mL (Normal range 15.00 – 68.00 pg/ml). Her serum albumin was 4.3 g/dl. Alkaline phosphatase was 78 U/L (38 -94 U/L), 25 hydroxy Vit. D was 20.12 nmol/L (< 25 nmol /L deficient). Renal and liver function tests were normal. ECG revealed a prolonged QT interval (QTc 620 ms) (Figure 3). CT scan head showed calcification of basal ganglia bilaterally (Figure 4).

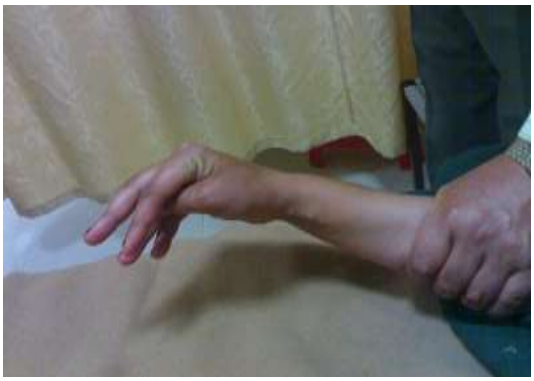

Figure 1: Photograph showing carpal spasm on compression of forearm.

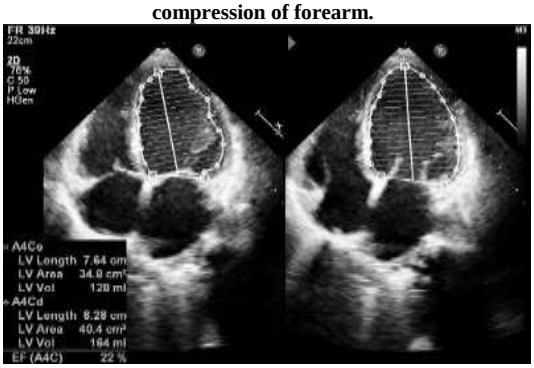

Figure 2: Echocardiography showing LV Dysfunction in hypocalcemia.

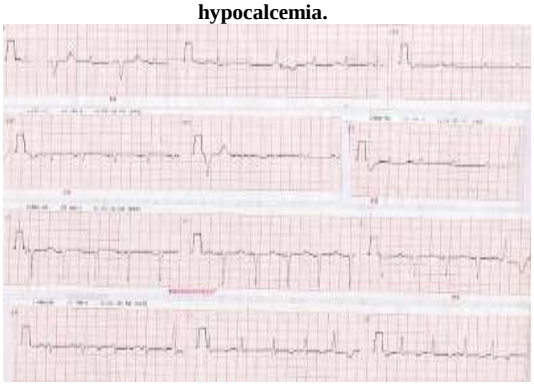

Figure 3: ECG showing prolonged QTc Interval.

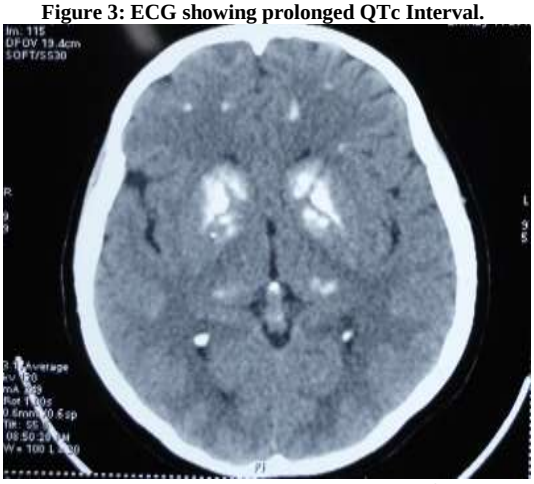

Figure 4: CT Scan head showing bilateral calcification in basal ganglia.

Figure 5: Echocardiography after correction of hypocalcemia.

Diagnosis of Pseudohypoparathyroidism and Vitamin D deficiency was considered. She was managed with calcium gluconate infusion for 24 hours and injection cholecalciferol 6 lakh IU intramuscular was given. After that oral calcium and 1, 25(OH) 2D supplements were started. Carbamazepine was tapered off. After 2 weeks her serum Ca^{++} was 8 mg/dl and Phosphorus was 4.6 mg/dl and Echocardiography parameters of LV function along with LV dimensions showed improvement and left ventricular ejection fraction was 25%. Chest X ray showed clearing of the opacities. QTc improved to 472 milliseconds. Patient came for follow up after 4month. She was asymptomatic and her serum calcium was 9.4 mg/dl and phosphate was 4.5mg/dl. she never developed any seizure or tetany after treatment. Her echo was normal and ejection fraction was 60% (Figure 5). She was advised to continue the same treatment and to follow up after 6 months.

Discussion

Hypocalcaemia characteristically causes prolongation of the QT interval in the electrocardiogram.[2] Torsades de pointes can potentially be triggered by hypocalcaemia but is much less common than with hypokalemia or hypomagnesemia. In addition, decreased myocardial performance and even congestive heart failure have been reported.[3-7] Myocardial dysfunction is reversible with calcium repletion.[5,8] Hypocalcemic cardiomyopathy in PHP has been reported scarcely in literature.[9]

Our patient had PHP along with vitamin D deficiency causing hypocalcemia leading to QTc prolongation along with congestive cardiac failure which improved with restoration of calcium level and calcitriol supplementation. In this patient, vitamin D deficiency can be due to prolonged carbamazepine use and moreover deficiency of Vitamin D is very common in India.[10] Furthermore, conversion of 25-hydroxy vitamin D to 1, $25(OH)_2D$ is impaired in PHP due to parathormone resistance.

This case demonstrates that hypocalcemic cardiomyopathy should be considered in the differential diagnosis of congestive cardiac failure in PHP who do not respond adequately to the usual treatment. Recognition of the condition is important as myocardial impairment is reversible after administration of calcium.

References

1. Maeda SS, Fortes EM, Oliveira UM et al. Hypoparathyroidism and Pseudohypoparathyroidism: a review. *Arq Bras Endocrinol Metab*2006;50/4:664-673.
2. Benoit SR, Mendelsohn AB, Nourjah P, et al. Risk factors for prolonged QTc among US adults: Third National Health and Nutrition Examination Survey. *Eur J Cardiovasc Prev Rehabil* 2005;12:363.

3. Kazmi AS, Wall BM. Reversible congestive heart failure related to profound hypocalcemia secondary to hypoparathyroidism. *Am J Med Sci* 2007;333:226.
4. Levine SN, Rheams CN. Hypocalcemic heart failure. *Am J Med* 1985;78:1033.
5. Wong CK, Lau CP, Cheng CH, *et al*. Hypocalcemic myocardial dysfunction: short- and long-term improvement with calcium replacement. *Am Heart J*. 1990;120:38.
6. Kudoh C, Tanaka S, Marusaki S, et al. Hypocalcemic cardiomyopathy in a patient with idiopathic hypoparathyroidism. *Intern Med* 1992;31:561.
7. Brunvand L, Haga P, Tangsrud SE, Haug E. Congestive heart failure caused by vitamin D deficiency? *Acta Paediatr* 1995;84:106.
8. Rimailho A, Bouchard P, Schaison G, et al. Improvement of hypocalcemic cardiomyopathy by correction of serum calcium level. *Am Heart J*. 1985;109:611.
9. Mönig H, Föh KP, Schulte HM, Simon R. Hypocalcemic cardiomyopathy as the cause of severe left heart failure. *Dtsch Med Wochenschr*. 1994 Sep 23;119(38):1270-1275.
10. Harinarayan CV, Joshi SR. Vitamin D status in India-Its implications and Remedial Measures. *J Assoc Physicians India*. 2009;57:40-48.