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CENTRAL HYPOTHYROIDISM DUE TO RATHKE CLEFT CYST MANIFESTING AS VENTRICULAR BIGEMINY: A CASE REPORT

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ABSTRACT **BACKGROUND:** Central hypothyroidism is a disorder of pituitary, hypothalamus or hypophyseal portal circulation leading to thyroid dysfunction. Rathke's cleft cyst is a cystic sellar mass arising from the remnants of Rathke's pouch. It is one of the rare causes of central Hypothyroidism. Hypothyroidism prolongs the cardiac action potential which predisposes the patient to ventricular irritability manifesting as ventricular arrhythmias.

Case presentation: We present a rare case of central hypothyroidism in a 48 years old male manifesting with lethargy, facial puffiness, weight gain, exertional dyspnoea and atypical chest pain. Cardiac evaluation revealed premature ventricular contractions with ventricular bigeminy. Etiological evaluation revealed Rathke's cleft cyst on neuroimaging. Patient had complete resolution of cardiovascular symptoms and ventricular premature contractions with thyroxine replacement.

Conclusion: Rathke's cleft cyst is one of the rare causes of central hypothyroidism which may manifest with ventricular arrhythmias and there is complete reversal of cardiovascular manifestation with Thyroxine replacement as highlighted in this case.

KEYWORDS : Central Hypothyroidism, Rathke's cleft Cyst, Premature ventricular contractions, Ventricular Bigeminy.

BACKGROUND

Central hypothyroidism refers to thyroid hormone deficiency due to a disorder of the pituitary, hypothalamus, or hypothalamic-pituitary portal circulation, resulting in diminished thyroid-stimulating hormone (TSH), thyrotropin-releasing hormone (TRH) or both. It is one of the rare causes of hypothyroidism and its prevalence is documented in 1:20,000 to 1:80,000 in the general population [1]. The mass lesion of pituitary especially pituitary adenomas are the common cause of central hypothyroidism followed by acute hemorrhage, infarct or apoplexy of the pituitary gland [2]. Mutations in the TSH-beta subunit, multiple genes and pituitary transcription factors such as POU1F1 (PIT1), PROP1, HESX1, SOX3 and SOX2 [3,4]. Other mass lesions that can cause central hypothyroidism include cysts and abscesses, meningiomas and dysgerminomas, metastatic tumors like craniopharyngiomas which may enter the sella or remain suprasellar. Rathke's cleft cysts (RCCs) are benign, sellar and/or suprasellar lesions originating from the remnants of Rathke's pouch. Although a common finding in routine autopsies (12-33% of normal pituitary glands), symptomatic cases are rare and comprise 5-15% of all surgically resected sellar lesions [5]. Most of the RCCs are asymptomatic and rarely they may manifest by compressing adjacent structures and causing pressure effects such as headache, visual disturbance, or pituitary hormone deficits. In a study by Culver SA et al 76% cases had normal anterior pituitary hormone levels. Prolactin abnormality was commonest in 5.3% cases followed by Panhypopituitarism and Thyroid hormone abnormality in 4% cases and rarely other pituitary hormone abnormality[6]. Thyroid hormone abnormality due to RCCs is documented in the literature, but the clinical manifestation and that too predominantly cardiovascular manifestation is rarely reported. Hence we report a rare case of central hypothyroidism due Rathke's cleft cyst with cardiovascular manifestations.

CASE SUMMARY

A 48 years old male with no prior comorbidities presented with a history of easy fatigability in the form of difficulty in performing physically demanding activities for the past 10 months. He also complained of bilateral jaw pain and facial puffiness with no diurnal variation since 05 months. He had gained 10 Kg weight during the last 5 months. He gave history of spontaneous onset and spontaneous offset palpitations since 2-3 months which was not associated with exertion. He had one episode of atypical chest pain and was admitted for cardiac evaluation. There was no history suggestive of post palpitation diuresis or diaphoresis or syncope. There was no history suggestive of connective tissue disorder in the form of photosensitive rash, joint pain or swelling and oral ulcers. There was no history of headache, nausea, vomiting, dysuria, haematuria or frothy urine. On clinical evaluation he was afebrile, pulse was 64 bpm regularly irregular, blood pressure was 108/64 mm of Hg right arm supine with no thyromegaly. Other general physical examination and systemic examination findings were essentially within normal limits.

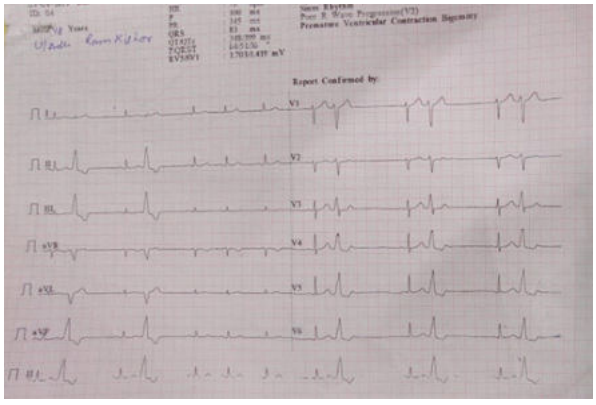

Image 1: Showing PVCs and Bigeminy on ECG at presentation to the hospital.

Initial ECG **(Image-1)** showed ventricular bigeminy with normal QT interval and no dynamic ST-T changes. 2D Echocardiography showed a structurally normal heart normal ventricular ejection fraction (LVEF 60%). 24 Hr Holter monitoring showed occasional VPCs with no abnormal pause, atrial fibrillation, atrial flutter or ventricular tachycardia. Frequent PVCs and Bigeminy seen in baseline ECG disappeared during TMT and there was no evidence of reversible myocardial ischaemia.

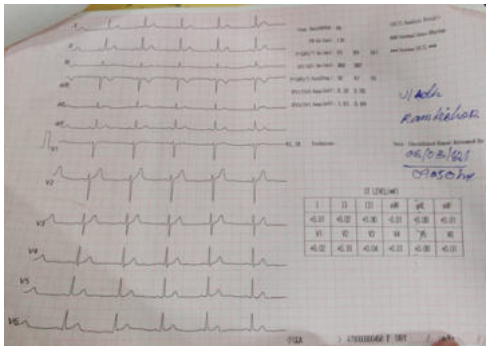

Image 2: Showing the rathke's cleft cyst on neuroimaging

Lab evaluation revealed normal complete blood count, liver function test, renal function test, blood sugars and lipid profile. Thyroid function test revealed suppressed early morning T3 and T4 levels with normal TSH level [T3: 75 (80-220); T4: 0.62 (5.10-14.10); TSH: 2.340 (0.27-4.20)]. Other anterior pituitary hormone levels were within normal range. His Anti-Thyroid Peroxidase Antibody was negative (<9.0) and early morning serum cortisol level was within normal limits [17.0 ug/ml (4.82 -19.90)]. There was no evidence of thyromegaly or increased vascularity on USG Neck. ANA and dsDNA were also negative, ruling out connective tissue disorder. Further to establish the cause of hypothyroidism **MRI Brain (02/03/021- Image 2)** was done which showed a 2 mm size non enhancing lesion in the central part of the anterior pituitary which appears hypo intense on on T1WI and Hyperintense on T2WI with no contour bulge in pituitary and normal para sellar areas suggestive of Rathke's Cleft Cyst.

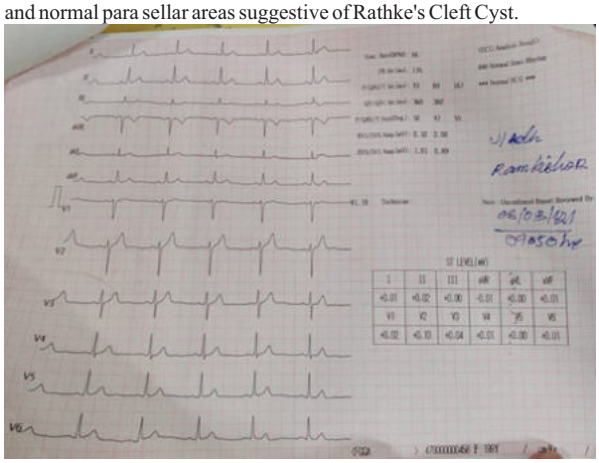

Image 3 showing the reversal of egg changes after 01 month of thyroxine replacement

Patient was managed as a case of central Hypothyroidism due to compression of the central portion of anterior pituitary by Rathke's cleft cyst with replacement of thyroxine hormone. On follow up after 01 month the patient is symptomatically better with resolution of presenting symptoms. Repeat ECG post thyroxine replacement did not reveal any bigeminy or VPCs **(Image 3)**.

DISCUSSION

Symptoms of cardiovascular dysfunction are not common or prominent in patients with hypothyroidism. It may manifest as exertional dyspnea, chest pain, exercise intolerance as in our case and edema. Findings on physical examination may include bradycardia, hypertension, non-pitting pedal edema, and pleural or pericardial effusion. Our patient presented with complaints of easy fatigability, weight gain, exertional dyspnoea and atypical chest pain requiring hospital admission and evaluation which revealed ventricular bigeminy. However there was no structural abnormality or inducible ischemia noted on detailed cardiovascular evaluation. He was found to have central hypothyroidism likely due to Rathke's cleft cyst and was managed with thyroxine replacement. The patient had complete resolution of the symptoms and reversal of ECG abnormality. In a study by Keating F R Jr et al it was reported that chest pain resolves only in 50% cases as in our case[7]. The conduction defects like Bigeminy, VPCs and bradycardia may either increase or decrease with T4 replacement. Only slight improvement in heart rate has been noticed with no decrease in PVCs. There may be occasional increase in VPCs with T4 replacement, but on the contrary our patient had complete resolution of VPCs.[8].

RCCs are the one of the rarest causes of central Hypothyroidism. Biochemical hypothyroidism has been reported in 4% cases of RCCs by Culver SA et al but the significant clinical manifestations are even rare[6]. Our patient on evaluation was found to have RCC but he did not have any complaints of headache, visual field defects or other pituitary hormone deficiency apart from thyroid hormone deficiency. Hence he was managed conservatively and is being closely followed up. In one of the largest follow up studies in patients with Rathke's cyst; out of 75 patients, in 57% cases there was no detectable increase in cyst size, only 28% of the cysts increased in size, whereas 15% decreased in size. Hence conservative treatment is recommended in these patients and a close follow up for onset of pituitary and optic chiasm compression leading to headache, visual field defect etc. as was done in our case[6].

CONCLUSION

This case highlights the rare cardiac manifestation of hypothyroidism and the importance of detailed evaluation of a case of hypothyroidism for central causes. Timely correct therapy can result in prompt and complete reversal of symptoms and signs

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