Case Report

A rare case of duodenal carcinoid presenting as ectopic Cushing’s syndrome

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ABSTRACT

ACTH-dependent Cushing syndrome (CS) due to an ectopic source is responsible for approximately 10-15% cases of Cushing’s syndrome. It is associated with various tumors such as small cell lung cancer and well-differentiated bronchial or gastrointestinal neuroendocrine tumors. Many a times ectopic ACTH production is difficult to manage, and identification of the source may take many years. Hormonal diagnostics include assessments in basic conditions as well as dynamic tests, such as the high-dose dexamethasone suppression test and corticotrophin releasing hormone (CRH) stimulation test. Treatment selection depends on the type of tumor and its extent. In the case of neuroendocrine tumors, the main treatments are surgery and administration of somatostatin analogues or bilateral adrenalectomy in refractory cases and if the source remains unidentified. Here, we report a case who presented with features of Cushing’s syndrome which eventually through workup led us to a diagnosis of duodenal carcinoid producing ectopic ACTH which is extremely rare and was successfully treated.

Keywords: ACTH secreting Tumors, Duodenal carcinoid, Ectopic Cushing’s syndrome

INTRODUCTION

Cushing’s syndrome is a state of hypercortisolism which results from chronic exposure to excess glucocorticoids from exogenous or endogenous source. It is associated with increased morbidity and mortality from musculoskeletal, metabolic, thrombotic, infectious and cardiovascular complications. Confirmation of hypercortisolism, identification of its cause and accomplishment of optimum treatment is still a challenging process. Manifestations range from subclinical, cyclical or mild to rapid-onset severe variants. Hypercortisolism due to ectopic ACTH syndrome is usually severe and of rapid onset. Ectopic ACTH producing Cushing’s syndrome is rare and constitutes only 15% of the total cases with most of them being attributed to bronchial carcinoid. Therefore, prompt clinical suspicion, early diagnostic work-up and management are necessary to avoid potential adverse outcomes. This is a rare case of a duodenal carcinoid producing ACTH causing Cushing’s syndrome.

CASE REPORT

This case was a 50-year-old female who had cushingoid features presented with history of irrelevant speech for two days, decreased urine output for 10 days and pain abdomen with vomiting for one day. She also had a history of weight gain which was around 12 kg over 6 months. She had a five-year history of rheumatoid arthritis.
arthrits and hypothyroidism, on treatment but was not treated anytime with prednisolone. She also had a history of diabetes and hypertension for one year which was poorly controlled. On general physical examination patient, was conscious but disoriented to time, place and person. She had positive findings of Facial puffiness, hirsutism, pallor and bilateral pitting pedal oedema with BMI of 33.2. Her Blood pressure was 170/110 mmHg. On Nervous system examination, Patient was moving all the four limbs with substantial decreased power in proximal part of bilateral lower limbs. Other system examination, Abdomen was soft, and no mass was palpable. Respiratory and Cardiovascular examination were also found to be normal.

In routine investigations, Complete blood count showed anaemia (7.5 g/dl) with elevated total WBC count (22,300/cumm) having neutrophilic predominance (78.4%). Renal function test comprising Urea (31 mg/dl) and creatinine (1.0 mg/dl) were normal. Amongst electrolytes, there was hyponatremia (118 mEq/l) with hypokalemia (2 mEq/l). Arterial blood gas analysis showed Metabolic Alkalosis (pH - 7.46, pCO2 - 32, pO2 - 68, HCO3- 29, SO2- 95%). Blood and Urine cultures were sterile. During her hospital stay, she developed high grade fever and psychosis. She was managed conservatively as sepsis with metabolic encephalopathy. Her hypertension was difficult to control even with three anti-hypertensives reaching up to their maximum doses. There was sustained hypokalemia throughout the hospital stay which was refractory to treatment.

Once clinically stable, 24-hour urine free cortisol was done, found to be elevated 2736ug/day (4.3-176). Serum ACTH was elevated 180.2 pg/ml (0-46) and High dose dexamethasone suppression test did not suppress serum Cortisol- 14.4ug/dl (Normal range- 3.7- 19.4). In order to localize the source of ACTH secretion, Radiological studies were done. CECT Thorax was normal. CECT Abdomen showed bilateral bulky adrenals which was followed with 68 Gallium DOTATOC PET CT. It showed two intramurals enhancing polypoidal lesions in 2nd and 3rd part of duodenum suggestive of somatostatin expressing tumour with Para duodenal and retro pancreatic lymph node spread (Figure 1 and 2).

Histopathology confirmed the diagnosis as an ACTH secreting well differentiated neuroendocrine tumour of grade 1 duodenal carcinoid. Post-surgery, her hypokalemia was treated with spironolactone and over next 6 months she was weaned off from all medications. Her potassium levels, Blood pressure and blood sugars came to normal limits without any medications. Her repeat ACTH after 3 months after surgery came to normal levels. Patient weight reduced by 10 kgs in 3 months follow up (Figure 4 and Figure 5).
DISCUSSION

Ectopic Cushing’s syndrome presents as a diagnostic challenge not least because of its myriad presenting features. They tend to present with short duration and rapid worsening of symptoms as in this patient. The source should be established because the excision of the tumour can be curative.

Ectopic Cushing’s syndrome results from inappropriately high levels of ACTH, secreted by various types of tumours. These tumours consist of NETs, islet cell tumours, small cell lung carcinomas, medullary thyroid cancers, pheochromocytomas, carcinomas of the thymus and pancreas tumours. Small cell lung carcinoma and lung carcinoids causes half of cases.

Carcinoid tumours represent 1.2%–1.5% of all gastrointestinal tract neoplasms. Within the gastrointestinal tract, carcinoid tumours are most commonly found in the appendix, followed by the ileum, rectum, and stomach.

Duodenal carcinoids are extremely rare, and their characteristics and biological behaviour have not been fully elucidated. Duodenal carcinoid tumours are most commonly found on the first part of the duodenum. These tumours are usually indolent, especially when they are small and limited to the submucosa, and the symptoms are generally nonspecific. The ideal treatment for duodenal carcinoids is an endoscopic or a radical surgical excision.

Most of duodenal carcinoids are known to produce serotonin including tachykinins, motilin and prostaglandins. ACTH secreting duodenal carcinoid is extremely rare as per our knowledge, only 3 cases have been reported in the literature so far.

CONCLUSION

A duodenal carcinoid producing ectopic ACTH is extremely rare and very few cases have been reported worldwide. A short history of symptoms with rapid progression must alert the clinician to a possibility of an ectopic source of Cushing’s syndrome. It is important to keep a wide perspective while evaluating when the commonly known sources of ectopic ACTH are not detected such as in this patient.

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