International Journal of
Science Inventions Today
www.ijsit.com
ISSN 2319-5436
Case Report

AN UNUSUAL PRESENTATION OF PANCREATIC VASOACTIVE INTESTINAL PEPTIDE TUMOUR (VIP OMAS) WITH GASTROINTESTINAL STROMAL TUMOUR (GIST) WITHOUT HYPOKALEMIA AND HPERCALCEMIA: A CASE REPORT WITH REVIEW ARTICLES

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ABSTRACT

Pancreatic VIPoma are rare neuroendocrine tumour of gastrointestinal tract. These tumours secret excessive amount of vasoactive intestinal peptides, which simultaneously leads to chronic diarrhea wth electrolytes disturbances. The annual incidence of tumour is one in 10000000 individual in general population. Most commonly presented with diarrhea associated with electrolyte disturbances. Here we are presenting a case of 65 years female patient, complaining diarrhea for more than three months and surprisingly laboratory reports shows there is normal serum potassium and calcium level. Thus this is unusual presentation which is significant for both literature and scientific purpose.

Key words: *Pancreatic VIPoma, Hypokalemia, Gastric GIST, GIP-NET (Gastrointestinal-pancreatic neuroendocrine tumours)*

INTRODUCTION

In 1958 Verner and Morrison [1] describes two cases of severe watery diarrhea non-associated with insulin secreting islet cell adenoma of pancreas. This association was previously known as VERNER MORRISON SYNDROME or also known as WHDA (Watery, Diarrhea, Hypokalemia, Achlorhydria) or cholera of pancreas. Other features are cutaneous flushing, hypokalemia associated renal failure, reduced or absent gastric secretion, Diabetes Mellitus and Hypercalcemia[2,3].

An association between ganglioneuroblastoma and diarrhea are usually seen in children[4]. Vasoactive intestinal peptide is a basic 28 amino acid peptide present in central and peripheral nervous system. Its role is as a putative neurotransmitter, which is supported by neurophysiological experiments [5].

In 1973 raised plasma and tumour concentration of VIP was demonstrated in patient with ganglioneuroblastoma and secretory diarrhea [6]. We present single case of pancreatic VIPoma without hypokalemia and hypercalcemia.

CASE REPORT

A 65 years old female patient, farmer by occupation, presented with three months history of watery diarrhea varying three to four episodes per day. The diarrhea was not-associated with pain, no abdominal distension, no blood in stool. In this period patient was afebrile.

The diarrhea is also not-associated with nausea or vomiting. Upper gastrointestinal endoscopy shows 2cm^{2}small nodular mass in body of stomach and CT scan shows Probable GIST. Later ultrasonography andCT scan shows small cystic lesion in tail of pancreas. Her investigation are as follow:

Vitals

Blood pressure 130/75 mmHg

Temperature 36.50c

Pulse 78/min

Respiratory Rates 20/ min

Biochemical findings

K^+ 3.5 mmol/l

Albumen 271.20 mg/l

Na^+ 142.3 mmol/l

Amylase 187U/l

Cl^- 108.0 mmol/l

Lipase 264U/l

Ca^{++} 2mom/l

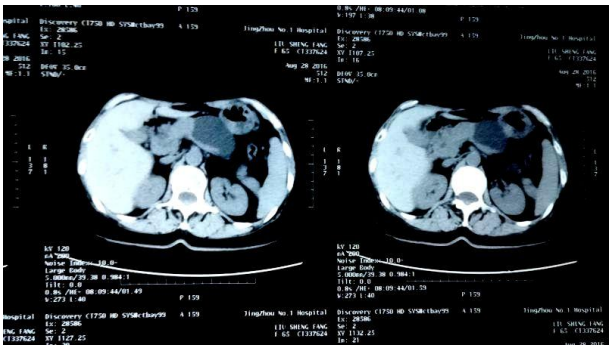

Figure 1

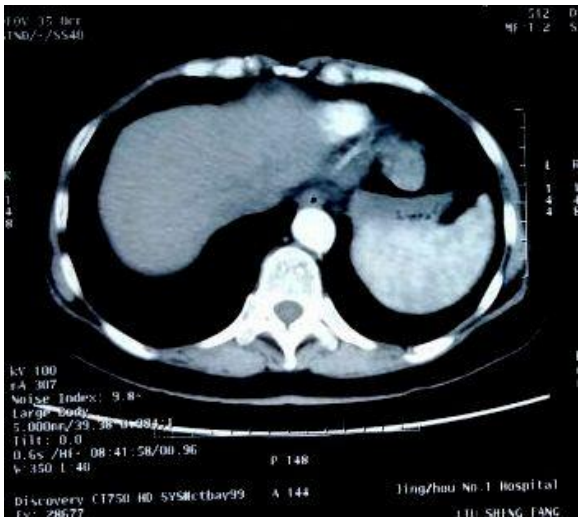

Figure 2

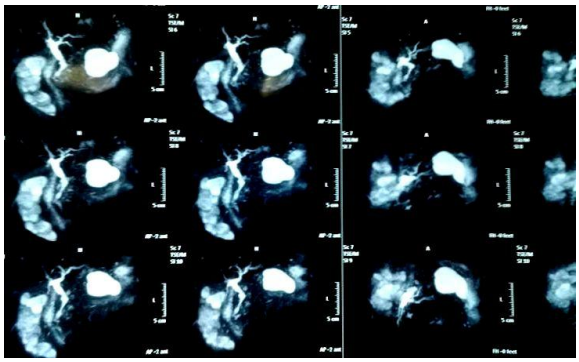

Figure 3

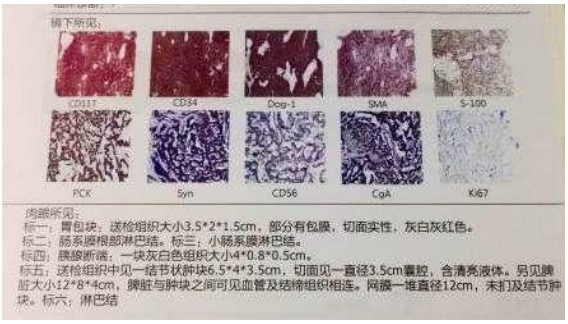

Figure 4

DISCUSSION

The neuroendocrine tumour of pancreatic islet cells can be functional tumour often shows malignant courses (Table -1) [7-9]

Tumor (Penetrance %)	**Sites**	**Malignancy (%)**
Non-functioning (60–100)	Pancreas	64-92
Insulinoma (21)	Pancreas	12-20
Gastrinoma (50)	Pancres, duodenum (>80%)	60
Glucagonoma (3)	Pancreas	35
Somatostatinoma (1)	Pancreas, duodenum/ jejunum (44%)	70
VIP-oma (1)	Pancreas, duodenum (10%)	40
GHRH-oma (1)	Pancreas	30

Table 1

VIPoma syndromes also known as VERNER-MORRISON SYNDROME is usually present with WDHA syndrome [10]. Pancreatic tumour are not sole origin of VIP secretion [11,12]. It may also produced by neuroblastoma, bronchogenic carcinoma, ganglioneuroblastoma or pheochromocytoma [13, 14]. As VIPoma present with watery diarrhea with hypokalemia our case is atypical with clinical features of VIPoma.

In our case these is not-association of hypokalemia or hypercalcemia or achlorhydria. These features make difficult for the initial diagnosis of disease. Other features of pancreatic VIPomas are consistent with our caase, as study shows peak incidence in 4th or 5th decade [15,16]

The case is also associated with GIST which makes it rarest [8-10]. The VIPoma in our case is present in tail of pancreas (figure 1-figure 4) as the typical presentation where 75% of tumour presents in tail or body of pancreas [17]. Surgical resection and pathological study confirms our diagnosis.

CONCLUSION

Pancreatic vipoma with gastric GIST are neuroendocrine tumours with not nerely present with WDHA syndromes. It can also present without hypokalemia or hypercalcemia.

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