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A Coat Button Lesion in Duodenum

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INTRODUCTION

Most duodenal neuroendocrine tumors (NETs) are detected incidentally and therefore are recognized at an early stage. Duodenal NETs are well differentiated, less than 10 mm and limited to the mucosa/submucosa. The management of duodenal NETs 1-2 cms needs an interdisciplinary discussion before management and those more than 2 cms need surgery. Endoscopic ultrasound is the method of choice to determine tumor size and depth of infiltration. Surgery is recommended for any localized ileal NET. Advanced small intestinal NETs with a carcinoid syndrome are treated with long-acting somatostatin analogs. This treatment significantly improves survival in patients with metastatic NETs. For optimal NET management, tumor biology, type, localization and stage of the neoplasm, as well as the patient's individual circumstances have to be taken into account.

CASE REPORT

38 year old female with history of melena, two weeks back (not endoscoped then), admitted with two episodes of black tarry stools in early morning on the day of admission. Her clinical examination was unremarkable except for mild pallor. CBC showed Hypochromic microcytic anemia. All other baseline investigations were normal. Upper GI endoscopy (Figure 1) showed approximately 1 cm^2 well circumscribed, ulcerative lesion in second part of duodenum. Biopsy showed well differentiated neuroendocrine tumor [Fig 2]. CECT abdomen showed normal Study. Biopsy for Immune-histochemistry was Positive for Chromogranin and Synaptophysin. Urinary 5-HIAA was 5.8ng/ 24 hours (N= 1.8-6.0 ng/24 Hrs.). Serum Chromogranin A was 356 ng/ml (N<100ng/dl) this patient was subjected to duodenotomy and resection of the lesion. Surgical biopsy confirmed the diagnosis. Extension of

involvement was up to serosa. MIB- labeling index was less than 1.

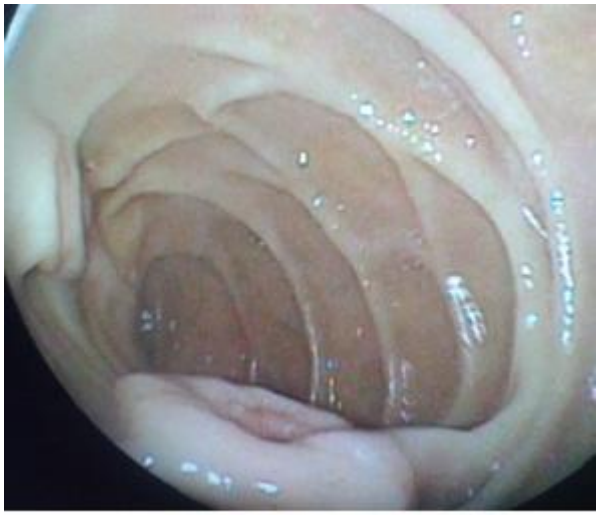

Figure 1

Figure 1. Endoscopic image: well circumscribed, elevated, flat topped lesion with ulcer over it (Coat button lesion) in D2.

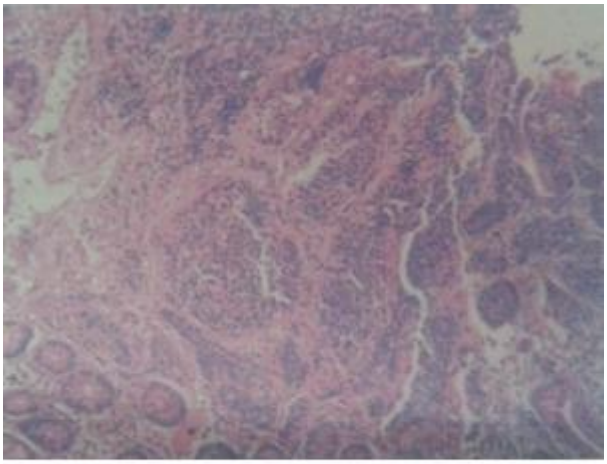

Figure 2

Figure 2. HPE- Well differentiated Neuroendocrine tumour (Carcinoid)

DISCUSSION

Lubarsch first described what are now recognized to be Neuroendocrine tumors (NETs) when he described multiple ileal tumors on autopsy[1]. The other name "carcinoid" was introduced by Oberndorfer in 1907 when he described a class of less malignant tumors than adenocarcinoma[2]. The traditional classification based on the anatomical location is now being abandoned [3], and is now being replaced by classification based on tumor biology introduced by WHO in 2000. Its revision and amplification by Europian Neuroendocrine Tumor Society (TNM classification) is going to be standard classification in future[4]. NETs arise from cells of diffuse neuroendocrine system, so can arise anywhere in GIT and are related to Medullary carcinoma thyriod, pheochromocytoma and pancreatic neuroendocrine tumors.

Gastrointestinal NETs synthesize bioactive amines and peptides like 5-hydroxytryptamine or serotonon, 5-hydroxytryptophan (5-HTP), chromogranin A, pancreatic polypeptide, calcitonin and various growth factors. Gastrointestinal NETs can be sporadic or part of familial syndromes. Gastrointestinal NETs comprise 0.5% of all malignancies and their incidence has increased over last few decades[5]. These tumors are most frequently found in GI tract (67%) followed by bronchopulmonary system (25%); less common locations are Ovaries, Testes and Hepatobiliary system[6]. Most common site in GI tract is ileum (17%), while as duodenal carcinoids comprise 4% of all GI NETs. Most GI NETs are asymptomatic. These tumors can present as pain abdomen, intestinal obstruction, abdominal angina and carcinoid syndrome[7, 8]. Most common histopathological types of gastrointestinal NETs is the Enterochromoffin (EC) cell carcinoid, which are composed of medium sized tumor cells arranged in arganoid

pattern showing only mild to moderate atypia and characterized by argentaffin staining, serotonin production, typical pleomorphic secretory granules. Necrosis is absent and mitotic rate is < 2/10 HPF. The other morphological types are Somatostatin cell tumors (Somatostatinomas) and gangliocytic paragangliomas. Duodenal carcinoids account for 11% of all intestinal NETs. Their incidence increased from 2.6% in 1973 to 16% in 2002. The annual incidence of duodenal carcinoids is 0.07/100,000 population. It is high in males and blacks and whites[5,6].

Duodenal carcinoids present at a mean age of 48-62yrs. Most duodenal carcinoids are symptomatic because of their local growth, as such presenting with obstruction, jaundice, abdominal pain and GI bleeding. Less than 10% present with symptoms of hormone overproduction like Zollinger Ellison syndrome (ZES), Carcinoid syndrome, Cushing's syndrome and Acromegaly. Duodenal carcinoids are mostly located in first and second part of duodenum. A high percentage(25%) is seen in peri ampullary area. The average duodenal carcinoid size is 0.1-4cm. Gastrin cell tumors represent most frequent histopathological type of duodenal NETs (50-60%). These tumors can be non functional or functional (ZES). Somatostatinoma (15-27%) and gangliocytic paraganglioma (6-9%) are other histopathological types[7,8].

The overall 5yr survival of duodenal carcinoids is 84%[5]. However it varies with histopathological type, extent of disease and presence of hormonal syndrome. Surgery is the only form of curative therapy for carcinoid tumors, however most symptomatic patients are not candidates for curative surgery and may require medical management after tumor debulking[9]. The type of surgery depends upon the location of the tumor and depth of involvement ranging from endoscopic resection to local surgical resection to extensive surgical procedures like hemicolectomy, Abdomino-perineal resection (APR) and Whipple's procedure.

REFERENCES

1. Lubarsch O: Ueber den primaren Krebs des Ileum, nebst Bemerkungen ?ber das gleichzeitige Vorkommen von Krebs und Tuberkuolose. *Virchow Archiv Pathol Anatom Physiol Klin Med* 1867; 111:280-317.
2. Oberndorfer S: Karzenoide Tumoren des D?nndarms. *Frankf Zschr Path* 1907; 1:426-30.
3. Williams ED, Sandler M: The classification of carcinoid tumours. *Lancet* 1963; 1:238-9.
4. Solcia E, Kloppel G, Sobin L: *Histological typing of endocrine tumours*. In: Verlag S, ed. *World Health Organization histological classification of tumors*, 2nd ed. New York: Springer; 2000:38.
5. Kloppel G, Perren A, Heitz PU: The gastroenteropancreatic neuroendocrine cell system and its tumors: The WHO classification. *Ann N Y Acad Sci* 2004; 1014:13-27.
6. Kloppel G, Perren A, Heitz PU: The gastroenteropancreatic neuroendocrine cell

system and its tumors: The WHO classification. *Ann N Y Acad Sci* 2004; 1014:13-27.\

7. Burke AP, Sobin LH, Shekitka KM, et al: Somatostatin-producing duodenal carcinoids in patients with von Recklinghausen's neurofibromatosis. A predilection for black patients. *Cancer* 1990; 65:1591-5.
8. Burke AP, Helwig EB: Gangliocytic paraganglioma. *Am J Clin Pathol* 1989; 92:1-9.
9. Akerstrom G, Hellman P: Surgery on neuroendocrine tumours. *Best Pract Res Clin Endocrinol Metab* 2007; 21:87-109.