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SM Journal of Clinical Pathology

Case Report

Primary Mesenteric Neuroendocrine Tumor: A Case Report and Review of the Literature

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Article Information

Received date: Nov 19, 2018
Accepted date: Nov 27, 2018
Published date: Nov 29, 2018

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Keywords Primary, mesenteric, neuroendocrine, tumor

Abstract

Mesenteric primary neuroendocrine tumors (NET) are extremely rare, while mesenteric secondary NET is classical, reported that 40% to 80%. And the distant metastasis rate reported that 80% to 90%, when the size is larger than 2 cm [1]. We present a rare case mesenteric neuroendocrine primary tumor Benin showing the character despite his size greater than 2 cm. The patient visited our hospital, with increasing palpable abdominal mass. At laparotomy, a resulting mass mesentery well encapsulated near the ligament of Treitz was found without any extension to adjacent organs or distant metastasis. The mass was measured 55x54 mm and histology plan was classified Grade1 (G1) classification of WHO 2017. At 11 months follow-up, the patient had no recurrence.

Introduction

Neuroendocrine tumours (NETs) are tumours that commonly involve the lungs and gastrointestinal system, especially the small intestine, appendix, rectum and pancreas. They have also been occasionally reported in ovaries, the prostate,thymus, lymph nodes and the cervix. Mesenteric NETs are extremely rare and are often secondary to another primary tumour located elsewhere. They can go unrecognized due to nonspecific symptoms.

Epidemiology Mesenteric masses are predominantly lymphatic origin. The incidence of cystic masses of the mesentery is estimated at 1/100 000 admissions in the United States, 50% of cystic lymphangioma. Lymphoma (especially non-Hodgkin phenotype b) is the most common solid tumor mesenteric. Neuroendocrine tumors of the small intestine are rare, but the mesenteric lymph node metastases are present in 80-90% of cases. 8% of desmoid tumors are localized in the mesentery. Other mesentery tumors are rare. Some lesions are classically described in other anatomical locations, and rarely in the mesentery (liposarcoma, pseudotumor inflammatory, Castleman's disease, actinomycosis) [2].

Case report

A 56-year-old woman presented with history of lower abdominal pain for 4 months, gradually progressive, type cramps, lumbar irradiation with low-grade fever term, constipation and asthenia. No significant medical history. Physical examination revealed a round and fixed mass with tenderness in the periumbilical area . Computed tomography (CT) scan demonstrated an 5.5 × 5.4cm sized well-defined mass with complex density located in the mesentery (Figure 1).

Computed tomography scan shows well-defined and complex density mass in mesentery.

At laparotomy, a well-encapsulated mass was found to be arising from the small bowel mesentery near the ligament of Treitz. We dissected and removed the mass and part of the small bowel compared to the mass.

Macroscopically, the mass was encapsulated measuring 7x6x3cm, gray-white appearance and firm consistency (Figure 2).

Histological examination showed tumor cells composed of homogenous small cells arranged in a trabecular and pseudo acinar pattern with the nucleus showing a round shape, rarely nucleolus, and coarsely granular chromatin pattern. There was no mitosis (Figures 3, 4A and 4B). Immunohistochemistry showed the tumor to be positive for neuroendocrine markers including chromogranin A (Figure 5A), synaptophysin (Figure 5B), and less than 2% of the tumor cells express Ki67 (Figure 4C). Histopathologic examination confirmed the mass as neuroendocrine tumors well differentiated grade 1 according to the WHO classification 2010 (called before carcinoid tumor).

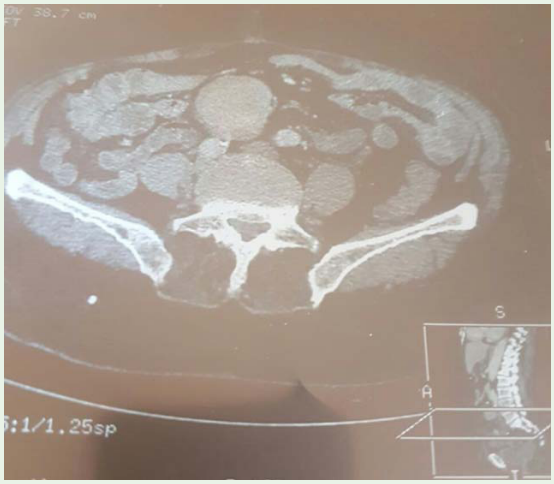

Figure 1: Computed tomography scan.

Figure 2: Encapsulated solid lesion focally adherent to the small bowel.

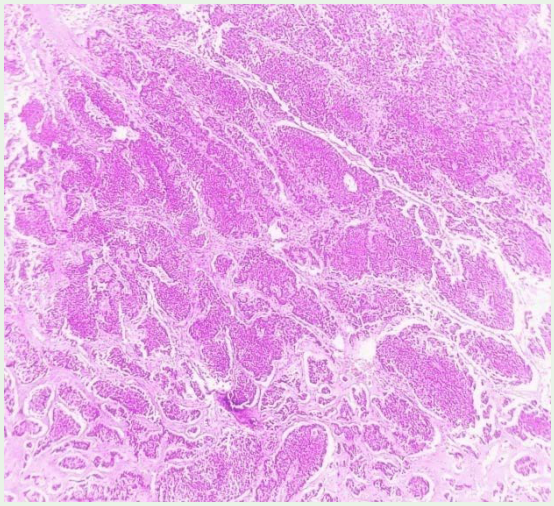

Figure 3: Photomicrograph showing tumor cells in trabecular pattern (H and E, ×100).

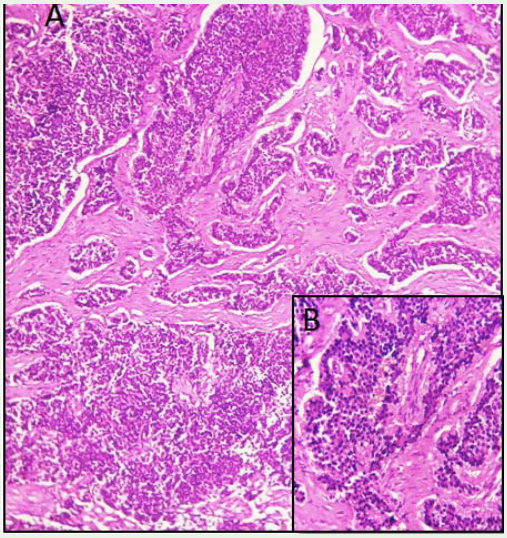

Figure 4: (A) Cells composing tumor are arranged in trabecular pattern (H&E,×200). (B) Cellular nucleus has coarsely granular chromatin pattern without mitosis (H&E,×400).

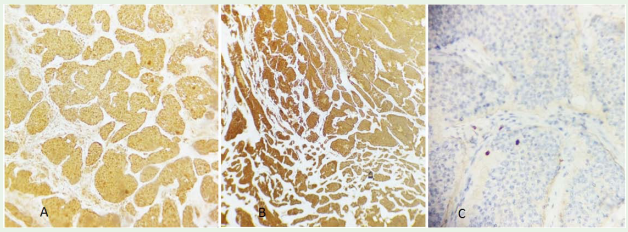

Figure 5: Tumor expresses strong positivity in immunohisochemical stain with Chromogranin (A), synaptophysin (B) and less than 2% of cells express ki67 (C) (×200).

Discussion

Carcinoid tumor is a rare, slow-growing, and NET with about 90% of the lesions arising in the gastro intestinal track (GIT). GIT carcinoid tumors are classified by the embryologic origin as foregut, midgut and hindgut. 46% to 64% of GIT carcinoid tumors arise in the midgut and most midgut carcinoid tumors originate in the terminal ileum. However, primary carcinoid tumors of the mesentery are very rare [3]. And carcinoid tumors arising in the mesentery are usually metastatic. Midgut carcinoid tumors commonly spread to the mesentery, reported as occurring in 40% to 80% of cases in various series [4].

NETs originate from neuroendocrine cells, which are widely distributed throughout the body. They secrete various substances and hormones including ACTH. These substances result in diverse clinical presentations. NETs most commonly involve the lungs and gastrointestinal system. They have also been reported in other sites such as the ovaries, prostate, lymph nodes and cervix [5-7]. Gastrointestinal NETs usually involve the small bowel, rectum, appendix and pancreas. Primary mesenteric NETs are extremely rare and very few cases of primary mesenteric involvement have been reported worldwide [5-8].

Citation: Essaoudi MA, Aboulfeth EM, Albouzidi A, Aitali A and Oukablii M. Primary Mesenteric Neuroendocrine Tumor: A Case Report and Review of the Literature. SM J Clin Pathol. 2018; 3(2): 1018.

The clinical presentation of these tumours depends on their location, and the types of hormones and substances they secrete.

On CT scan, mesenteric carcinoid tumors exhibit varying degrees of fibrosis, calcification, focal or diffuse neurovascular bundle invasion by the tumor or both mechanisms [4].

Microscopically, typical NETs have one of five growth patterns: insular, trabecular, glandular, undifferentiated, or mixed [9]. Most midgut carcinoids show a mixed insular and glandular growth pattern [10]. NETs have specific immunohistochemical characteristics. Synaptophysin, chromogranin, cytokeratin and neuron-specific enolase are usually positive. [1] In our case, synaptophysin, chromogranin A were positive, which confirmed the diagnosis of NET. To make the diagnosis of mesenteric NET, you must first rule out other primary sites by the use of CT colonoscopy, the series of the small intestine and scintigraphy [11]. In our case the scanner objectified hepatic hydatid cyst calcified segment I without tumor lesion of other organs. Colonoscopy, small bowel series and abdominal surgical exploration can confirm the diagnosis of mesenteric NET [11].

Surgical excision is a mainstay of treatment for carcionid tumor. Larger tumors are usually associated with locally advanced or distant metastasis [9]. Also, approximately half of midgut carcinoid patients present with liver metastasis [4].

Generally, for tumors smaller than 2 cm without lymph node involvement, local segmental resection is adequate [9,10]. And tumors larger than 2 cm with regional mesentery metastasis and lymph node involvement, wide excision of the bowel and mesentery with lymph node dissection is needed because tumors larger than 2 cm are associated with 80% to 90% incidence of metastasis [9-12]. Also, surgical excision of local or distant metastatic carcinoid tumors is recommended, both for relief of symptoms and prolonged survival [9-12].

Conclusion

Primary mesenteric NETs are very rare entities and very few cases have been reported worldwide. After a Thorough Assessment eliminating other primary sites, it should be kept in mind that the mesentery can also be a primary site for NETs. Hence, use of specific radiologic imaging techniques is obligatory in the diagnostic of neuroendocrine tumors in rare locations as mesentery, stomach and rectum. Timely identification of these usually undetected tumors prevents diagnostic delays and development of carcinoid syndrome surgical excision is is the cornerstone of treatment for NETs tumors with minimum rate of complications and mortality.

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