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Primary Neuroendocrine Tumor of Mesentry-A Rare Case Report

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Abstract **Case Report**

Primary neuroendocrine tumor of the mesentery is very rare. More than 90% of gastrointestinal neuroendocrine tumors are located in the appendix, small intestine and rectum. We report a case of 70yr old male with very rare primary neuroendocrine tumor of the mesentery without any evidence of any primary tumor site. The only symptoms were persistent pain of the abdomen lasting for one year. The mass was radically excised during laparotomy. CT and MRI were useful to clarify the site of origin of the tumor but final diagnosis was based on histological findings.

Keywords: Primary neuroendocrine tumor, appendix, CT and MRI, diagnosis.

INTRODUCTION

Neuroendocrine neoplasms (NENs) encompass diverse types of tumors arising from neuroendocrine (enterochromaffin) cells found throughout the body. Well-differentiated NENs, also known as neuroendocrine tumors (NETs), are generally indolent and are often found incidentally, while poorly differentiated tumors are more aggressive. Most of the neuroendocrine tumors (NET) of the gastrointestinal tract are traditionally termed "carcinoid tumors." More than 90% of gastrointestinal carcinoids are located in the appendix, small intestine and rectum [1]. Carcinoid tumor is the most frequent primary malignant tumor of the small intestine beyond the ligament of Treitz and most frequently arises in the terminal ileum [2, 3]. Secondary mesenteric involvement of small bowel carcinoid tumors is common, reported as occurring in 40% to 80% of cases [3]. However, primary mesenteric carcinoid tumors are very rare and most solid tumors arising in the mesentery are usually metastatic tumors [2].

Broadly, NENs are classified based on tumor differentiation into well-differentiated (low and intermediate grade) neuroendocrine tumors (NETs) and poorly differentiated (high grade) neuroendocrine carcinomas [4]. Grading addresses aggressiveness of the tumor, with grade 1, 2, and 3 representing low, intermediate and poorly differentiated tumors, respectively [5]. Well-differentiated NETs are generally rare [4]. Carcinoid tumors are well-differentiated NETs arising commonly from the digestive tract [6]. Carcinoid tumors account for about 2% of all gastrointestinal tumors [7]. 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.

CASE REPORT

A 70-year-old man presented with abdominal pain and palpable abdominal mass from the last one year. On physical examination, patient was afebrile, pilse 90/min regular and blood pressure 126/88mm Hg. Per abdominal examination revealed an huge round and fixed mass with mild tenderness in the periumbilical area noted. Computed tomography (CT) scan demonstrated an 25×26x34mm sized well-encapsulated enhancing mass located in the mesentery. At laparotomy, a huge mass was found with bowel loops around it forming a ball like structure, which was completely excised and sent to pathology department for histopathological examination. There were no abnormal findings in the other organs including the small bowel and colon.

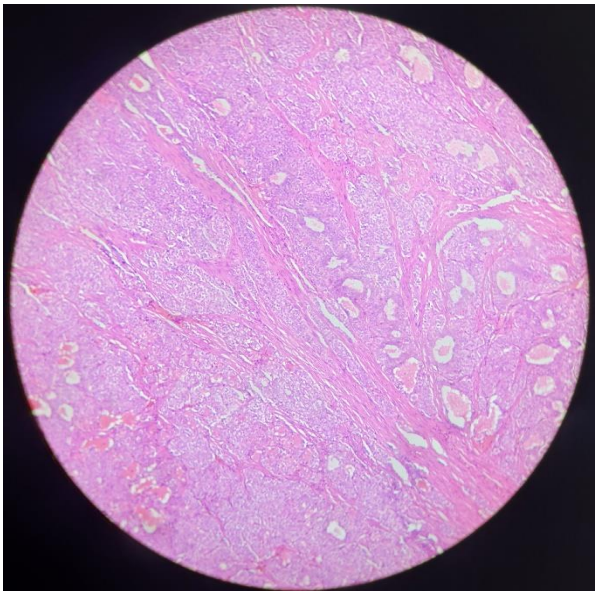
Gross Examination

Segment of intestine measuring 80cm in length with a grey white encapsulated growth at the mesenteric border measuring approximately 4x3x2cm in size. Cut section- grey white. The intestinal loop was unremarkable. On examination of the mesentery, no lymph nodes were dissected.

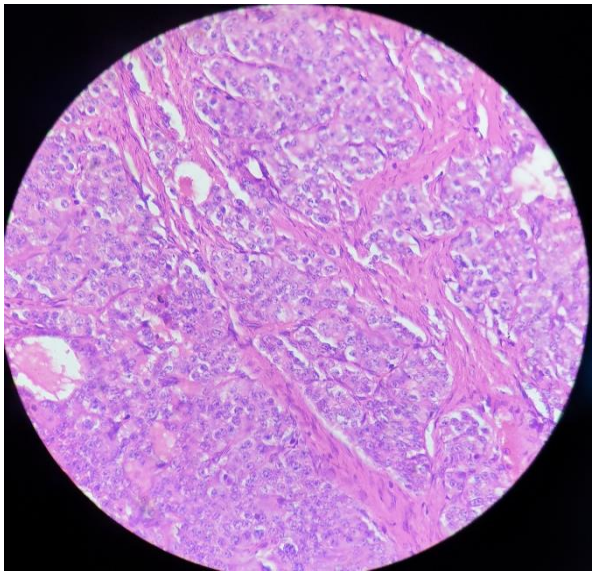
Well-capsulated mass in the mesentry (Left upper corner)

Microscopic examination

Histological examination showed tumor cells composed of homogenous small cells arranged in a trabecular pattern with the nucleus showing a round to oval shape, indistinct nucleoli, and coarsely granular chromatin pattern. There was no mitosis. No invasion was seen into the adjacent mesentery and intestinal loop. Mitotic activity -1mitoses/ 10 hpf. Immunohistochemistry showed the tumor to be positive for neuroendocrine marker synaptophysin. Histopathologic and immunohistochemical examination confirmed the mass as a well differentiated Neuroendocrine tumor, Grade 1.



Tumor arranged in trabecular pattern and nests (H&E, ×40)



Cellular nucleus has salt and pepper chromatin pattern without mitosis (H&E, ×400)

DISCUSSION

Carcinoid tumor is a rare, slow-growing, neuroendocrine tumor with about 90% of the lesions arising in the GIT [11]. 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 [9].However, primary carcinoid tumors of the mesentery are very rare [8]. 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 [8]. It was shown in a series conducted by mayo clinic that in small bowel even small carcinoids can metastasize [12]. So, to rule out a possible primary small intestinal tumor, a meticulous pathological examination of the resected adjacent bowel is advised. A CT or 5 HIAA levels alone are not confirmatory. In 1996, Kimchi *et al*., [13] reported a case of primary mesenteric neuroendocrine tumor arising in the mesocolon. They did not find another tumor on abdominal CT or during laparotomy. Furthermore, the pathologists did not find a primary lesion in the resected colon; thus, helping in the diagnosis of a primary mesenteric carcinoid. In our case, we conducted a thorough histopathological examination of the resected bowel to rule out the presence of possible primary tumor in the small bowel loop. The occurrence of carcinoid tumor in the mesentery can be explained by the presence of neural crest cells in small amounts in unconventional sites such as interatrial septum of the heart, the liver hilus, and mesenteric vessels due to the dispersed migratory properties of the neural crest [14, 15].

On CT scan, mesenteric carcinoid tumors exhibit varying degrees of fibrosis, calcification, focal or diffuse neurovascular bundle invasion by the tumor or both mechanisms [8].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 [8]. 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 [9-11]. Also, surgical debulking of local or distant metastatic carcinoid tumors is recommended, both for relief of symptoms and prolonged survival, despite randomized evaluation not being done [9-11]. Microscopically, typical carcinoid tumors 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]

CONCLUSION

The primary mesenteric carcinoid tumor is a rare entity. A thorough histopathological examination of the resected bowel is required to rule out the presence of possible primary tumor in the adjacent bowel. Carcinoid tumors often have advanced malignant potential depending on location, size, nature. But our case shows a rare large primary mesenteric carcinoid tumor with characteristics of uncommon position and benign nature

REFERENCES

1. Kvols, L. K. (1998). Gastrointestinal carcinoid tumors and the malignant carcinoid syndrome. In: Sleisenger, M. H., Sleisenger, M. H., Fordtran, J. S., Feldman, M., & Scharschmidt, B., editors. Sleisenger & Fordtran's gastrointestinal and liver disease: pathophysiology, diagnosis, management. 6th ed. Philadelphia, PA: Saunders; p. 1831–1833.
2. Karahan, O. I., Kahriman, G., Yikilmaz, A., Ozkan, M., & Bayram, F. (2006). Gastrointestinal carcinoid tumors in rare locations: imaging findings. *Clinical imaging*, *30*(4), 278-282.
3. Woodside, K. J., Townsend Jr, C. M., & Evers, B. M. (2004). Current management of gastrointestinal carcinoid tumors. *Journal of gastrointestinal surgery*, *8*(6), 742-756.
4. Klimstra, D. S., Modlin, I. R., Coppola, D., Lloyd, R. V., & Suster, S. (2010). The pathologic classification of neuroendocrine tumors: a review of nomenclature, grading, and staging systems. *Pancreas*, *39*(6), 707-712.
5. Rindi, G., Klöppel, G., Couvelard, A., Komminoth, P., Körner, M., Lopes, J. M., ... & Wiedenmann, B. (2007). TNM staging of midgut and hindgut (neuro) endocrine tumors: a consensus proposal including a grading system. *Virchows Archiv*, *451*(4), 757-762.
6. Modlin, I. M., Lye, K. D., & Kidd, M. (2003). A 5-decade analysis of 13,715 carcinoid tumors. *Cancer: Interdisciplinary International Journal of the American Cancer Society*, *97*(4), 934-959.
7. Horton, K. M., Kamel, I., Hofmann, L., & Fishman, E. K. (2004). Carcinoid tumors of the small bowel: a multitechnique imaging approach. *American Journal of Roentgenology*, *182*(3), 559-567.
8. Karahan, O. I., Kahriman, G., Yikilmaz, A., Ozkan, M., & Bayram, F. (2006). Gastrointestinal carcinoid tumors in rare locations: imaging findings. *Clinical imaging*, *30*(4), 278-282.
9. Woodside, K. J., Townsend Jr, C. M., & Evers, B. M. (2004). Current management of gastrointestinal carcinoid tumors. *Journal of gastrointestinal surgery*, *8*(6), 742-756.
10. Åkerström, G., Hellman, P., Hessman, O., & Osmak, L. (2005). Management of midgut carcinoids. *Journal of surgical oncology*, *89*(3), 161-169.
11. De Vries, H., Verschueren, R. C. J., Willemse, P. H. B., Kema, I. P., & De Vries, E. G. E. (2002). Diagnostic, surgical and medical aspect of the midgut carcinoids. *Cancer treatment reviews*, *28*(1), 11-25.
12. Kvols, L. K. (1998). Gastrointestinal carcinoid tumors and the malignant carcinoid syndrome. In: Feldman, M., Scharschmidt, B. F., & Sleisenger, M. H., editors. Sleisenger and Fordtran's Gastro-Intestinal and Liver Disease. 6th ed. Philadelphia: W.B. Saunders; p. 1831-1843.
13. Kimchi, N. A. (2001). Primary neuroendocrine tumor (carcinoid) of the mesocolon. *Isr Med Assoc J*, *3*, 288-289.
14. Kudoh, A., Tokuhisa, Y., Morita, K., Hiraki, S., Fukuda, S., Eguchi, N., & Iwata, T. (2005). Mesenteric paraganglioma: report of a case. *Surgery today*, *35*(7), 594-597.
15. Karahan, O. I., Kahriman, G., Yikilmaz, A., Ozkan, M., & Bayram, F. (2006). Gastrointestinal carcinoid tumors in rare locations: imaging findings. *Clinical imaging*, *30*(4), 278-282.