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Case report on neuroendocrine tumor of ileocaecal junction

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Abstract

Neuroendocrine tumors (NETs) are unique and rare neoplasms that arise from cells of the endocrine and nervous systems. These tumors have been found in almost every organ such as gastrointestinal tract, bronchopulmonary, pancreas, uterine cervix, urinary bladder, and salivary gland, but primary sites are gastrointestinal tract and lungs. We studied a rare case of neuroendocrine tumor grade II located in the ileocaecal junction. Tumor cells show strong synaptophysin positivity. Contrast Enhanced Computed Tomography (CECT) scan, Colonoscopy, Ultrasound Sonography (USG) abdomen, histopathological and hematological examination confirmed tumour nature. Treatment includes surgery as well as adjuvant chemotherapy (Etoposide and Cisplatin). Surgical details include post limited resection of ileocaecum, ascending colon and end to end ileotransverse colon anastomosis. Six cycles of chemotherapy are planned for the patient. Early detection and surgical removal of the tumor increase the survival rates. But, these are asymptomatic and are discovered upon surgery for unrelated causes.

Keywords

neuroendocrine tumour; etoposide; cisplatin

Abbreviations

NET: Neuroendocrine Tumor; CECT: Contrast Enhanced Computed Tomography; USG: Ultrasound Sonography; MRI: Magnetic resonance imaging**;** CEA: Carcinoembryonic Antigen; GEP-NET: Gastroenteropancreatic Neuroendocrine Tumors; AP: Anterior–Posterior; MEN: Multiple Endocrine Neoplasia; VHL: Von Hippel-Lindau; RET: Ret Proto-Oncogene; TSC: Tuberous Sclerosis Complex

Introduction

Neuroendocrine tumors are thought to arise from various neuroendocrine cells throughout the diffuse endocrine system. Neuroendocrine cells are present not only in endocrine glands throughout the body that produce hormones but are found in all body tissues. They comprise a broad family of tumors, the most common of which are carcinoid and pancreatic neuroendocrine tumors. Other neuroendocrine tumors include those arising in the parathyroid, adrenal, and pituitary glands, and in calcitonin-producing cells of the thyroid [1]. The crude incidence has significantly increased over the last years and is now estimated to be 5.25/100,000 person-year. The prevalence has recently been calculated to 35/100,000 person-year. The incidence of the small intestinal neuroendocrine tumor (NETs) (carcinoids) is estimated to be from 0.32/100,000 person-year (England) to 1.12/100,000 person-year

(Sweden). The incidence of rectal tumors is 0.86/100,000 person-year, pancreatic 0.32/100,000 person-year, and gastric NETs 0.30/100,000 person-year. There is a slight overall higher incidence of NETs for males (5.35) compared with females (4.76) [2].

A plethora of genes are known to be involved in neuroendocrine tumorigenesis, including MEN1, RET, VHL, TSC1 and TSC2 with mutations in MEN1 remaining the most common form of genetic predisposition to neuroendocrine tumors. Neuroendocrine tumors form a diverse group of conditions with a large spectrum of clinical syndromes depending on the presence of secreted hormones. Their management is challenging since morbidity is quite often related to the hypersecretion syndrome rather than tumor bulk. The advent of genetic screening has had a significant effect on patient management and counseling, and it is hoped that new technologies will allow for more precise patient risk stratification. The frequent expression of somatostatin receptors by these tumors allows for the highly beneficial use of somatostatin analogues, and most recently the introduction of targeted therapy. This is a very exciting development in the field, and further results are currently awaited [3].

Case Presentation

A 47 year old male was presented with abdominal pain for 1 month, haematochezia for 2 years, diarrhoea/mucus discharge with stools. Pain is not associated with food intake. The patient had the history of dyspeptic symptoms. He had no similar episodes in past and urinary urgency for 1 month. No h/o hesitancy or dysuria. The local investigation revealed a mass 5 x 4 cm palpable in right iliac fossa extending to the right lumbar region, with well defined rounded borders was identified.

Routine hematological investigation and the biochemical investigation was performed and found to be normal. Carcinoembryonic antigen (CEA) was found to be slightly increased. The patient was a smoker and stopped since 2 years. In smokers, the carcinoembryonic antigen normal level is slightly above the normal individuals.

After the routine hematological and biochemical investigation Colonoscopy, USG, CECT was done. Colonoscopy shows mass lesion at caecum. USG abdomen shows Right Iliac Fossa: 5.5 X 3.8 cm mass consisting of aperistaltic bowel loops with thickened wall containing echogenic fluid (15-20 cc), two enlarged lymph nodes measuring 10-12 mm in AP diameter and edematous fat. CECT sections of the abdomen show asymmetrical wall thickening involving the medial wall of caecum terminal ileum with pericolic fat stranding with adjacent lymph node enlargement. And suggest the possibility of intestinal tuberculosis.

Surgery was the primary treatment objective. Based on the investigation report post limited resection of ileo-caecum + ascending colon and end to end ileotransverse colon anastomosis was done. The dissected portion sends for further pathological analysis. The result shows “Neuroendocrine tumor grade II”. The tumor is located in the ileocaecum junction 3.8 X 3.5cm. Tumor extends into serosal fat and serosal lymph nodes 2/3 show tumor infiltration and stage found to be P T4a N1 M0.

So adjuvant chemotherapy is started to the patients besides the surgery. 6 cycles of chemotherapy were planned. The choice of drugs was Inj. Etoposide 150 mg (days 1, 2 and 3) and Inj. Cisplatin 55mg (days 1 and 2) along with premedication.

Discussion

NETs are arising from various neuroendocrine cells found throughout the body that produces hormones. The normal function of neuroendocrine cells is to serve at the neuroendocrine interface [4]. There are 2 main types of NETs: gastroenteropancreatic neuroendocrine tumors (GEP-NET) and carcinoid tumors. Carcinoid tumors are most commonly affect the small bowel, particularly the ileum, and are the common malignancy of the appendix. The majority of carcinoids are asymptomatic and are discovered by coincidence (eg. surgery for an unrelated cause) [5]. NETs sometimes exhibit symptoms due to the secreted hormone [6,11].

Here, the patient presented with an abnormal mass at right iliac fossa with dyspeptic symptoms. Routine biochemical and hematological investigation reveal the slight excess in the levels of CEA. CEAs are a set of glycoproteins involved in cell adhesion. It is normally produced in gastrointestinal tissue during fetal development, but the production stops before birth. Therefore, CEA is usually present only at very low levels in the blood of healthy adults. However, the elevated serum level shows the presence of some cancers, which means that it can be used as a tumor marker in clinical tests. Serum levels can also be elevated in smokers [7]. From initial diagnosis concluded that elevated CEAs may be due to the presence of some carcinoid tumor or smoking habit of the patient.

Generally, CT-scans, MRIs, sonography (ultrasound), and endoscopy (including endoscopic ultrasound) are common diagnostic tools. CT-scans using contrast medium can detect 95 percent of tumors over 3 cm in size, but not tumors under 1 cm [8]. Here special investigations such as Colonoscopy, CECT and USG abdomen confirm the actual size, location, and nature of the mass.

Appropriate treatment of neuroendocrine tumors depends on its location, invasiveness, hormone secretion, and metastasis. The goal of therapy may be the cure of disease or relieving symptoms [9]. Surgery, chemotherapy, somatostatin analogues in secretory tumors for symptomatic relief, radionuclide therapy, radiofrequency ablation etc are the common treatment strategies [10]. Here also the primary option includes surgical removal of a part of ileum + caecum+ ascending colon and end to end anastomosis of ileum + transverse colon. The surgically removed portion further send for pathological examination confirmed the presence of neuroendocrine tumor grade II (P T4a N1 M0), which extends to the serosal fat and serosal lymph nodes. So besides surgery adjuvant chemotherapy is also started. Most gastrointestinal carcinoid tumors are not responding to chemotherapy agents, showing 10 to 20% of response rates that are typically less than 6 months. Combining chemotherapy medications has not usually been of significant improvement showing 25 to 35% response rates that are typically less than 9 months. Exceptionally for poorly differentiated (high-grade) metastatic disease cisplatin with etoposide can be used [10]. Here also the patient treated with cisplatin and etoposide combination chemotherapy.

Conclusion

Neuroendocrine tumors are rare neoplasms arising from the neuroendocrine cells. Early detection and treatment of these tumors increase the survival rates. Because of the poorer diagnosis, the mortality rate is increasing. Surgical removal of affected area is preferable in almost cases. In some cases, symptomatic relief treatments, chemotherapy or combination of these can be used.