

Carcinoid tumor of unknown origin with liver metastasis associated with carcinoid syndrome

R. Umadevi, M. S. Darsana, S. Gopalakrishnan*, G. L. Murugavel

ABSTRACT

Background: Carcinoids or neuroendocrine tumors (NETs) are derived from diffuse neuroendocrine system, which is made up of peptide and amine-producing cells with different hormone profiles depending on their site of origin. **Methodology:** These are metastasizing; highly aggravating tumors are associated with carcinoid syndrome. They are also named after the hormones they produce. **Results:** A 73-year-old female patient reported with abdominal pain weight loss and vomiting for about 1½ months. **Conclusion:** On further investigations showed focal lesions in liver, indicating metastasis of a NET origin.

KEY WORDS: Carcinoid syndrome, Neuroendocrine tumors, Radioimmunoassay

INTRODUCTION

Carcinoid tumors are common neuroendocrine tumors (NETs). Their primary localization is on pancreas, intestine and bronchus.^[1] They remain asymptomatic until they compress pancreatic duct, intestinal obstruction, or bronchoconstriction or they metastasize. Majorly, they metastasize to liver and usually present with carcinoid syndrome (CS). Major symptoms of CS are diarrhea, flushing, abdominal pain, weight loss, and valvular heart disease palpitations. The tumors are named after the common hormones they produce. Endoscopies, positron emission tomography scan, computed tomography, etc., are used for diagnosis. Five-year survival rate of all 8305 patients reported in Surveillance, Epidemiology, and End Results Program of the U.S. was 80%. The highest survival rate was reported in appendiceal carcinoid – 86–100%.^[2]

MATERIALS AND METHODS

Case Discussion

NETs or carcinoid tumors predominantly can be gastric carcinoids, pancreatic carcinoids, or the site of origin can be intestine, bronchial or can be of unknown origin too.^[3] Age of presentation is between

the 5th and 8th decades of life and there is no any sex predilection.^[4] Among the carcinoid syndrome, the incidence of gastric carcinoids and rectal carcinoids accounts for 9.6% and 16% respectively from 2000-2007.^[1] The treatment and type of carcinoids are classified predominantly with its primary location. Metastasizing sites can be liver, bone lymph nodes, etc., and their incidence in patient irrespective of their primary site is 45%. Prognostic value depends on the site of primary tumor and metastasizing sites. However, the diagnosis is done mostly after metastasis to the liver as they are highly symptomatic than the primary tumor.^[2] Diagnosis is carried out with the help of urinary release of 5HT, 5HTP, 5HIAA, histamine, and others. Chromogranin-A levels are increased in about 56–100% of patients, indicating that they are highly specific. Chromogranin-A levels are increased in about 56–100% of patients, indicating that they are highly specific, while plasma enolase is less specific as they increase only up to 17–47%.^[4]

Highly aggressive tumors show CS which occurs in about 4%. CS occurs due to interactions of secretions such as 5-HIAA, kallikrein, dopamine, histamine, and prostaglandin.^[4] CS can be typical-classic more common or atypical syndrome. Typical syndrome shows flushing, diarrhea, abdominal pain, valvular heart disease, palpitations, wheezing, and weight loss. In our patient, abdominal pain, weight loss, and vomiting are present.

Access this article online

Website: jprsolutions.info

ISSN: 0975-7619

Department of Community Medicine, Sree Balaji Medical College and Hospital, Bharath Institute of Higher Education and Research, Chennai, Tamil Nadu, India

*Corresponding author: Dr. S. Gopalakrishnan, Department of Community Medicine, Sree Balaji Medical College and Hospital, Bharath Institute of Higher Education and Research, Chennai, Tamil Nadu, India. Phone: +91-9941984498. E-mail: drsgopal@gmail.com

Received on: 17-08-2019; Revised on: 19-08-2019; Accepted on: 23-09-2019

In contrast, atypical syndrome consists of flush, headache, lacrimation, cutaneous edema, and bronchoconstriction. In our patient, carcinoid tumor of unknown origin liver metastasis is present.^[3] The differential diagnosis includes inflammatory bowel disease gut or pancreatic neoplasia, irritable bowel syndrome, and diverticular disease. Differential diagnosis of appendiceal carcinoids is mainly appendicitis.^[4]

Treatment can be mainly surgery in non-metastasizing carcinoids. In metastasizing carcinoids, chemotherapy is the mainstream treatment management. Complications of carcinoid tumors include mainly CS and their associated symptoms – severe diarrhea, flushing, pain, asthma and wheezing, pellagra, and carcinoid heart disease. Furthermore, the complications can be related to the site of metastasis and the hormone produced.^[3]

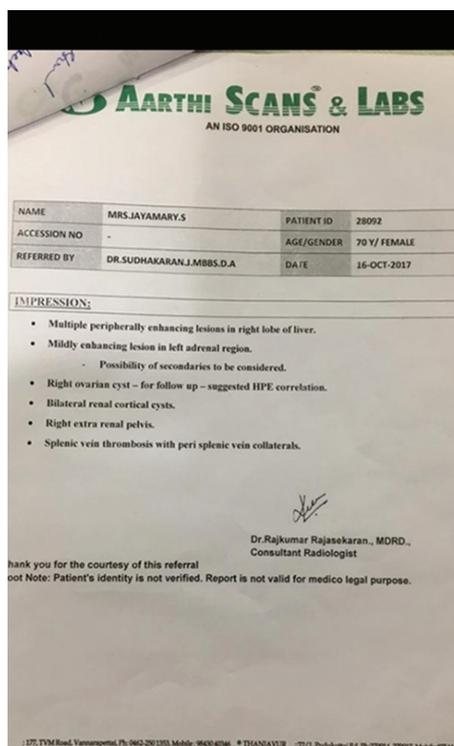
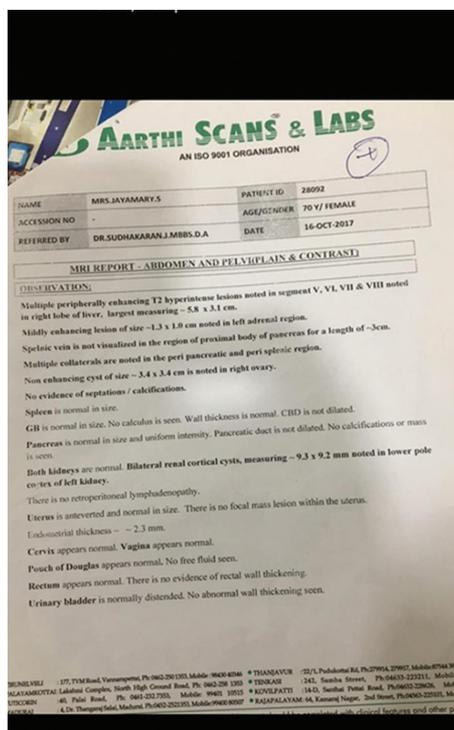
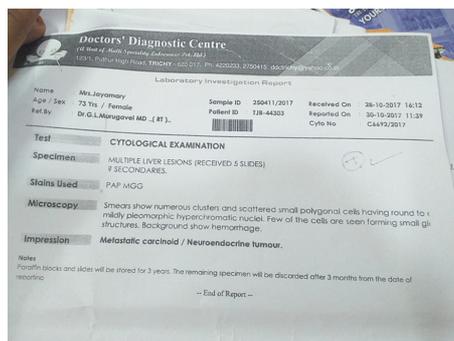
RESULTS

Case Report

A 73-year-old female patient presented with a history of abdominal pain, vomiting, and weight loss for 1½ months. The patient was conscious oriented. There was no pallor, icterus, lymphadenopathy and edema. Mild flushing of face was present without permanent discoloration left. The examination showed the following result: Pulse 90/min, blood pressure 110/80 mm/Hg, respiratory rate 18/min. No other abnormalities in systemic examination were found. Blood investigations show hemoglobin of 12.5 g/dl and raised erythrocyte sedimentation rate was reported. Lymphocyte rate was also slightly raised. Slight increase in blood glucose level was also found. Traces of glucose were found in urine too. First, ultrasonogram taken on October 2017 showed cystic mass lesion in the right iliac region measuring 3.5 cm × 2.7 cm no evidence of internal septations or internal vascularity. Magnetic resonance imaging report indicates multiple peripherally enhancing T2 hypertensive lesions in the right lobe of liver, indicating metastases. There was no evidence for calcification or necrosis. Cytological smear examination of multiple liver lesions shows numerous clusters and scattered small polygonal cells having round to oval mildly pleomorphic hyperchromatic nuclei. Background shows hemorrhage. This indicates an impression of metastatic carcinoid or NET.

CONCLUSION

Radioimmunoassay test indicates the presence of a tumor marker chromogranin A – an acidic human protein found in number of normal and neoplastic endocrine tissues and elevated plasma concentration, this indicates tumors of neuroendocrine origin.



The patient was not medically fit for surgery so chemotherapy and symptomatic treatment were given and her symptoms were relieved. The patient was followed up for about a year and she is well now.

REFERENCES

1. Devita VT, Lawrence TS, Rosenberg SA. Cancer Principles and Practice of Oncology. 8th ed., Vol. 2, Sec. 6. Philadelphia, PA: Wolters Kluwer/Lippincott Williams & Wilkins©; 2008.

2. Jameson JL, Kasper D, Hauser S, Dan Longo, Fauci A, Loscalzo J. Harrison, Principle of Internal Medicine. 19th ed., Vol. 2, Ch. 113. New York: McGraw Hill Education; 2015.
3. Kaltsas GA, Besser GM, Grossman AB. The diagnosis and medical management of advanced neuroendocrine tumors. *Endocr Rev* 2004;25:458-511.
4. Morgan JG, Marks C, Hearn D. Carcinoid tumors of the gastrointestinal tract. *Ann Surg* 1974;180:720-7.

Source of support: Nil; Conflicts of interest: None Declared