A 16 years old boy with neuroendocrine tumor at liver

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Introduction

- Neuroendocrine tumors (NETs) are a heterogeneous group of rare tumors with different and complex clinical behaviors, originating from peptidergic neurons and neuroendocrine cells throughout the body
- Most are gastroenteropancreatic neuroendocrine tumors (GEP-NETs).
- Generally, the incidence of GEP-NETs has increased continuously worldwide over the last decades due to the increased availability of diagnostic tools and awareness
- There has been no agreement about the treatment strategy and prognosis prediction

Case History

- A 16-year-old complained of abdominal pain beneath the xiphoid process and diarrhea for a months
- Physical examination revealed tenderness at the right upper abdomen.
- A fecal occult blood test was positive.
- MRI showed a lesion in the liver and near pancreas.
- Endosonographic examination showed a lesion at the left liver lobe and several lymph nodes in liver hilum with an ulcerated surface on duodenum, that was tough in texture.
- No abnormality was found in the tumor markers.

Biopsy

- FNB was performed from the liver mass, hilum, duodenum and lymph nodes from pancreatic tail lesions through trans gastric approach
- Surgery consult

Diagnosis

• The biopsy result was neuroendocrine tumor with 8 mitotic figures within 10 HPF

Clinical data:

Diarrhea, weight loss and abdominal pain since few months ago; liver mass lesion and liver hilar lymph nodes seen on CT scan

EUS impression:

Ampulla of Vater: There was no mass lesion seen at major papilla or ampulla of Vater.

Liver: There were several mass lesions in the left lobe of the liver. Some were hyperechoic and some were hypoechoic. The largest one was measured 17x16 mm in diameter. There was no ascites.

Pancreas: There was no mass lesion seen in the pancreatic head, neck, body, tail, or uncinate process. Lymph node: Several abnormal appearing lymph nodes in the liver hilum. The largest one was measured 36x25 mm in diameter. Two lymph nodes around the pancreatic tail.

Sample:

Immunohistochemical evaluation has been done on paraffin block number P-00-1790/A,D on one slide

Presumptive Histopathologic Diagnosis:

Reported as: In favor of neuroendocrine tumor

IHC Method:

Envision:

The following panels of antibodies are employed and the expressions are as follows:

CK7: Negative in tumor cells, positive internal control (entrapped biliary type epithelium)

CK20: Negative in tumor cells CD45: Negative in tumor cells

Chromogranin: Positive cytoplasmic staining of tumor cells

Ki-67: Less than 1% nuclear staining

CD56: Positive cytoplasmic staining of tumor cells

CDX2: Negative in tumor cells

TTF1: Negative in tumor cells CK19: Negative in tumor cells, positive internal control (entrapped biliary type epithelium)

The above immunostaining pattern is compatible with neuroendocrine tumor, well differentiated

Chemotherapy

• We decided to treat the patient with regimen of neoadjuvant chemotherapy (etoposide 100 mg D1–5 + Cisplatin 40 mg D1–3) after a multidisciplinary discussion in our hospital

LA somatostatin

• In consideration of the neuroendocrine symptom, the patient was also given long-acting octreotide once per month and 30 mg every time

TACE

- After 2 cycles of chemotherapy, a follow-up abdominal and pelvic enhanced CT examination implied the liver mass became larger
- TACE right hepatic artery

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Chemotherapy

- Continue chemotherapy with Etoposide and cisplatin plus 5FU
- After 3 courses
- Progressive disease



خیابان ولیعصر. بالاتر از میرداماد. خیابان رشید یاسمی بیمارستان فـوق تخصـصی خاتـم الاتـبیا (ص) تلفــــــن : ۸۳۵۵۷۰۸ - ۸۳۵۵۷۰۸۰

⁶⁸Ga-DOTA-TATE PET/CT Report

Name: Mr. Ardalani lilya

Date: 1400.09.10 Weight: 56 Kg

Second reviewed by: Dr. HR Amini

Age: 16 Y ID: 01064255

IMPRESSION & COMMENTS:

- * A DOTA-positive lesion in the pancreatic neck, consistent with malignancy.
- * The study is compatible with DOTA-positive metastases in:
 - Multiple liver lesions,
 - · Multiple abdominal lymph nodes.
- Diffuse DOTA uptake in gastric wall is non-specific finding.
 If clinically indicated, endoscopic evaluation is recommended.

Ludotatate

- 3 courses
- Good response

Name: Mr. Ardalani Ilya

Date: 1401.01.23

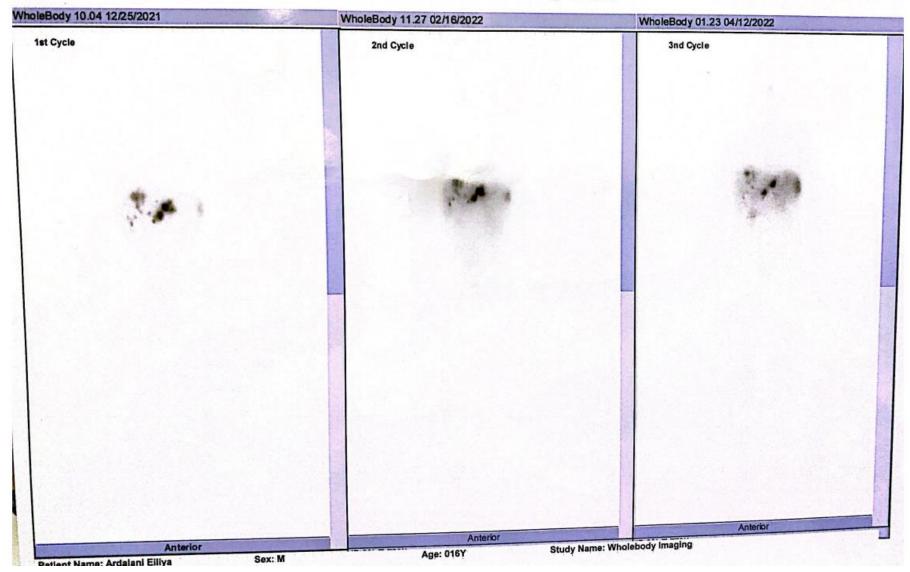
Post-therapeutic whole Body 177Lu-DOTATATE Scan

Procedure: Three days after IV injection of 150 mCi ¹⁷⁷Lu-DOTATATE, whole body scan obtained.

Interpretation:

- > 177Lu-DOTATATE uptake in the pancreatic lesion, multiple liver metastasis and several abdominal metastatic lymph nodes is noticed.
 - O When compared with prior post-therapeutic scans (1400.10.04/1400.11.27); there is interval reduce in radiotracer uptake of the pancreatic lesion along with reduced in size/uptake of some of the metastatic lesions. No new ¹⁷⁷Lu-DOTATATE-avid lesion is detected.

Jam Nuclear Medicine Center 177Lu-DOTATATE WholeBody Scan



Cont'

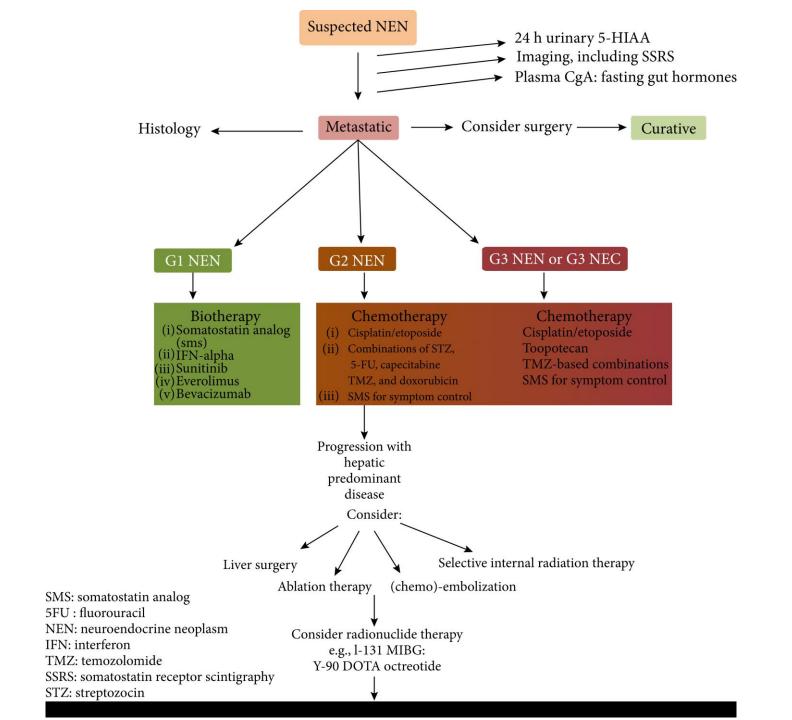
• Pembrolizumab after each therapy started 4 courses

Last scan



Discussion

- Generally, Neuroendocrine tumors (NETs) are rare tumors comprising ~2% of all malignancies with the gastrointestinal tract and the lung as the most common sites
- Neuroendocrine tumors (NETs) are a heterogeneous group of epithelial neoplastic proliferations arising in many body organs
- Irrespective of their primary site and of their grade of differentiation, neoplastic cells share features of neural and endocrine differentiation
- The improvement of the current diagnostic techniques has led to an increased number of patients diagnosed with GIT NET



Summary

- Neuroendocrine tumors (NET) account for about 1% to 2% of all gastrointestinal tumor cases.
- The most common clinical manifestations are abdominal distension and hepatic discomfort
- Abdominal mass, weight loss, nausea, vomiting, and diarrhea (less common)

Summary

- Despite the increasing number of patients diagnosed with GIT NET and also the increasing knowledge within this field, still, most of the cases are diagnosed in advanced stages
- Higher incidence of suspicion is required
- There are still some controversies regarding the sequence of systemic treatments used in the management of GIT NET that needs more clarification

Any Question

