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RESEARCH ARTICLE

ESOPHAGEAL COMPOSITE TUMOR WITH NEUROENDOCRINE COMPONENT: A RARE ENTITY

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Abstract

Gastro-entero-pancreatic neuroendocrine neoplasms are a rare entity, esophageal localization is even rarer and unfortunately known for its aggressiveness, early dissemination and poor prognosis. We document the case of a 44 years old woman with a metastatic squamous cell carcinoma neuroendocrine features which presented as dysphagia with right side abdominal pain.

Introduction:-

Gastro-entero-pancreatic neuroendocrine neoplasms are a rare entity, representing approximately 1% of digestive tumors[1]. Esophageal localization is even rarer and unfortunately known for its aggressiveness, early dissemination and poor prognosis.

Due to frequent metastasis, neuroendocrine carcinomas (NECs) may present as distant metastasis before a primary site is recognized[2]. In some cases, it may be found in composite malignant esophageal tumors, which are characterized by different contingentsincluding endocrine and more differentiated cellular proliferation associated in different proportion in the same tumor[3].

Case Report

We present the case of a 44 years old woman with history of heavy drinking and smoking habits, hypertension and familial history of breast cancer. She was admitted for dysphagia, black emesis and upper right abdominal pain, no weight loss or other symptoms were reported. Physical examination revealed normal vital signs and anicteric conjunctiva. The patient had upper quadrant tenderness, a firm hepatomegaly with irregular surface, reaching to the umbilicus, extendedto the left hypocondrium. The laboratory tests showed no anemia, the hemoglobin was 15,3g/dl,

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a hyperleukocytosis 13,1Giga/l, normal platelet count, TP 95%, elevated aminotransferases AST 135UI/l (3,3N), ALT 108UI/l (2,7N), and cholestasis (GGT 367UI/l (9,6N),ALP 223UI/l (1,9N)) the c-reactive protein was 49mg/l andbilirubin 11µmol/l. Tumor markers were negative (ACE, Ca19-9).Chromogranin A was 2367 ng/ml.

An esogastroduodenal endoscopy revealed a tumoral mass of the middle third of the esophagus with esophageal varices of grade I and cardio tuberositaryvarices of grade II, biopsies were performed and showed composite squamous cell carcinoma and high gradeneuroendocrine carcinoma.

Image 1:- HES x 40: invasive squamous cell carcinoma.

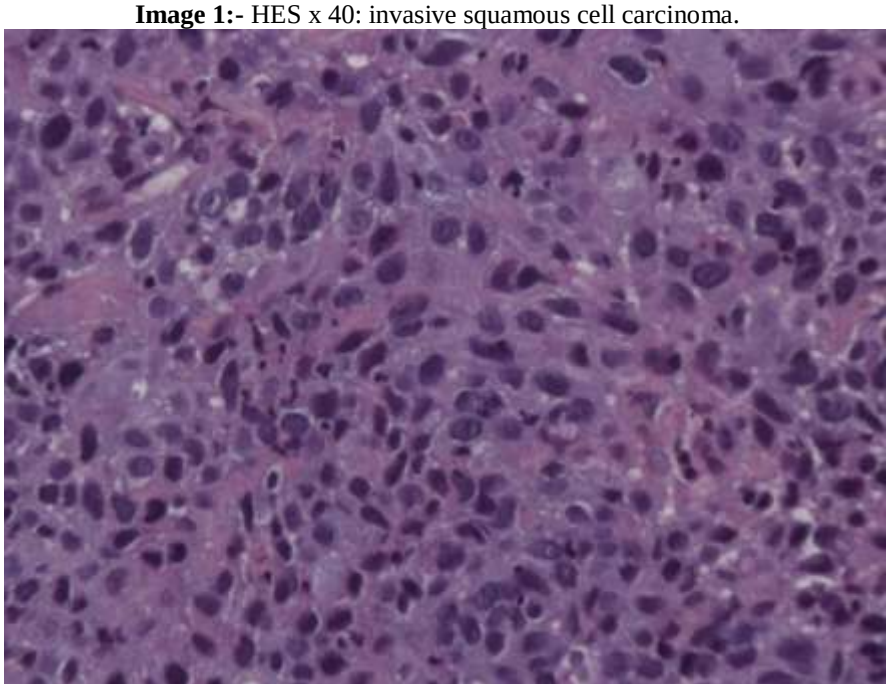


Image 2:- HES x40: small cell neuroendocrine carcinoma.

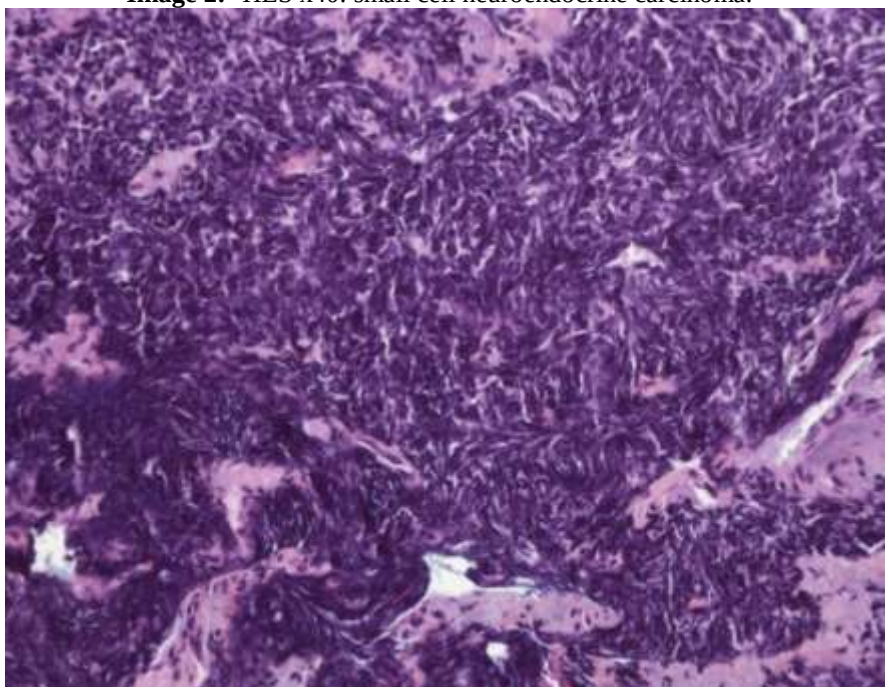
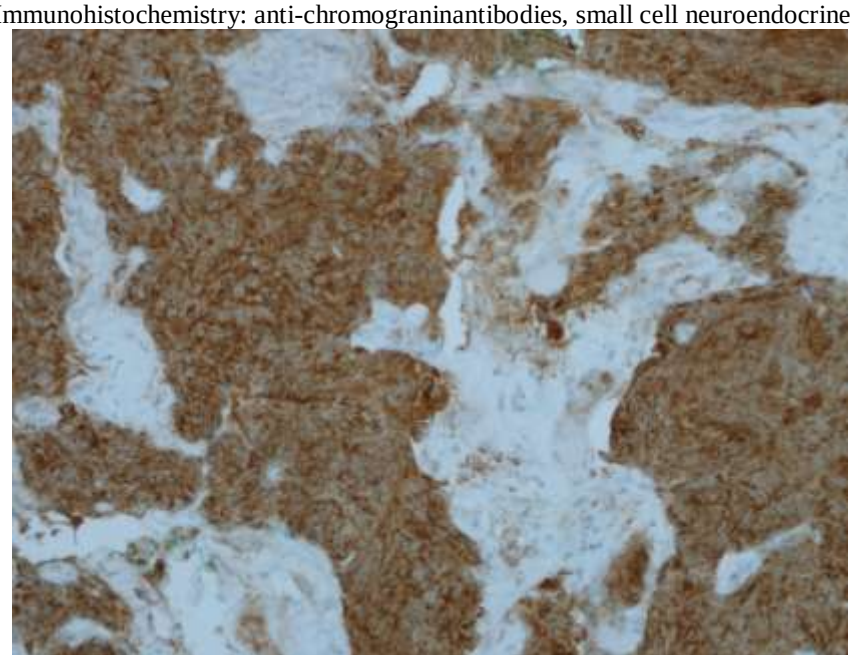
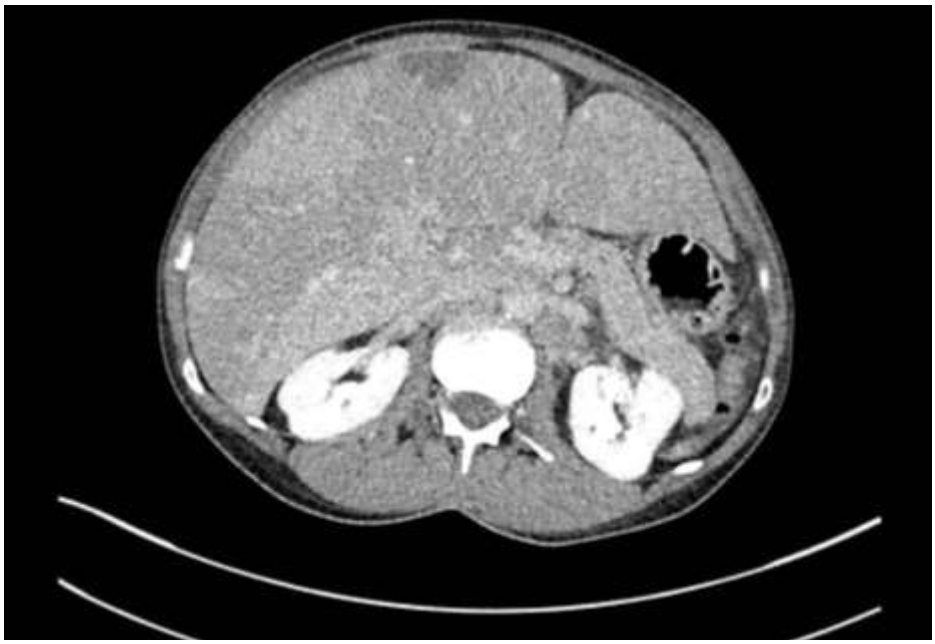


Image 3:- Immunohistochemistry: anti-chromograninantibodies, small cell neuroendocrine carcinoma.



A CT scan for the assessment of tumor local and metastatic extension showed a hepatomegaly measuring 23cm, site of an extensive, hypodense, heterogeneous infiltration, most probably of metastatic origin.Extensive venous thrombosis ofcommon femoral veins, iliac veins and partially thrombosed inferior vena cava. Laterogastricadenopathies of necrotic appearance, heterogeneous, located incontact with the anterosuperior part of the pancreas. An abdominal ultrasound with elastometry -ARFI was performed and showed a dysmorphic hepatomegaly with an elasticity of 50.4kPa and moderate ascites.



During her hospitalization, the patient presented progressively increasing intensity abdominal cramps requiring the administration of morphinics.

Her case was discussed in a multidisciplinary meeting. The decision was to start emergency chemotherapy in the oncology department using carboplatin and etopozide under cover of hyper hydration and allopurinol with a

controlled lysis syndrome.

The patient improved progressively, the liver enlargement regressed and the abdominal pain was under control, allowing to set up a discharge.

Unfortunately, the improvement did not last and she died a few weeks later of lievr failure before her next chemotherapy appointment.

Discussion:-

Small cell carcinoma of the esophagus (SCEC) is a rare form of esophageal cancer with a low incidence (0.1% to 2.4%)[4][5][6]. It is a highly aggressive tumor, with about half of patients presenting with metastatic disease. Treatment usually involves a combination of chemotherapy, radiotherapy, and/or surgery in cases of localized disease. However, even with multimodal treatment, overall survival is poor, with a median survival of 8 to 21 months. Since randomized studies are not available and are unlikely to be conducted due to the rarity of the disease, large retrospective series that use nationwide databases have been used to study survival based on treatment modality. However, these studies lack patient-specific data and cannot provide a conclusive recommendation for an optimal treatment regimen. Lesions can be mixed, such as neuroendocrine differentiation in adenocarcinoma, and are more commonly found in low-grade dysplasia.Composite tumors are rare and are characterized by the presence of multiple different types of cells within the same tumor, including squamous, glandular, and endocrine cells. These cells are associated in varying proportions within the same tumor[7][8][9]. In the case mentioned, the patient had high-grade dysplasia in the primary diseasewhich is not common.

The clinical presentation of endocrine tumors in the esophagus is similar to squamous cell carcinoma, with symptoms such as dysphagia. However, most patients with this condition have synchronous metastases[10]. The identification of the endocrine component in esophageal carcinoma is critical, as it is associated with a poor prognosis and a median survival of only 6 months. The poor prognosis of composite tumors seems to be linked to the presence of undifferentiated cells. The endocrine component of these tumors can spread independently and aggressively, suggesting a high degree of cellular dissemination.

The treatment of esophageal NEC lacks a standard or agreed upon algorithm in current literature. Endoscopic treatment is a suitable option for tumors between 0.2 and 0.8 cm without regional lymph node metastases, while surgical resection is best for those with metastasis or larger primary tumors. Recurrence of the disease after surgical or pharmaceutical treatment worsens the prognosis for patients with esophageal NEC. Somatostatin analogs, such as SOM230 and octreotide, are used to manage the hormonal symptoms associated with esophageal NEC. Patients with pure NECs have a worse prognosis compared to those with mixed NECs.

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