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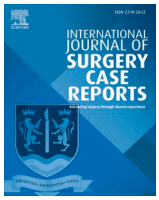

Case report

Endodermal sinus tumor with pancreatic origin: A case report

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A R T I C L E I N F O

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A B S T R A C T

Endodermal Sinus tumor is a rare malignant type of germ cell tumor (GCT). Primary endodermal sinus tumor, especially in the head of pancreas, is extremely rare. The case under consideration is of a 22 year old male who presented with pancreatic mass causing postprandial nausea and vomiting, and raised AFP levels. The biopsy of the mass revealed the endodermal sinus tumor, and the pancreatic origin was confirmed by a CT scan and a negative scrotal ultrasound. As the tumor was unresectable initially, a gastrojejunostomy was done to relieve the obstructive symptoms. Whipple procedure was performed two months after 4 cycles of BEP chemotherapy to remove the residual tumor in pancreas. It is crucial to highlight this uncommon case in order to increase surgeons' and oncologists' knowledge of the malignant extra-gonadal GCTs diagnosis and treatment.

1. Introduction

(The work has been reported in line with the SCARE criteria*).

Neoplasms originating from germ cells, germ cell tumors (GCT) can comprise both immature and mature components developing into various tissue types [1]. These tumors often begin in the gonads, but they can also develop in extragonadal locations (EGCTs), which account for 1–5 % of all germ cell tumors [2]. The two basic explanations for the occurrence of primary extragonadal GCTs are either aberrant somatic cell differentiation or improper implantation of germ cells during embryogenesis [3,4]. The brain, neck, mediastinum, retroperitoneum, vagina, and sacrococcygeal region are among the areas where extragonadal germ cell cancers frequently exhibit an axial distribution pattern [5,6]. There are several different types of tumors that make up GCTs, with teratomas being the most prevalent and endodermal sinus tumor (EST)/yolk sac tumor being the most frequently occurring malignant form [7]. The likelihood of GCTs in the pancreatic head region is very low, with only 3 reported cases of primary endodermal sinus tumor in the literature till date [7,18]]. In this article, we discuss a EGCT case that proved to be an endodermal sinus tumor (EST). As far as we are aware, there are incredibly few examples of primary extragonadal GCT of the pancreas recorded in the literature, especially in adolescent patients.

2. Case presentation

A 22-year-old male patient presented initially with primary complaints of post prandial vomiting, nausea, loss of appetite and epigastric pain. His past medical history was unremarkable. On admission, the physical examination revealed abdominal distension, mild discomfort and a palpable mass on deep palpation in the epigastrium. All the baseline investigations were in the normal range except mild electrolyte imbalance due to vomiting. Ultrasonography revealed a solid mass in the pancreatic head area with the suspected involvement of adjacent duodenum. An esophagogastroduodenoscopy was ordered which showed that the stomach was full of bilious fluid with dilated pylorus and an obstructive mass in the D2. An Endoscopic ultrasound guided fine needle aspiration (EUS-FNA) was done to take a biopsy sample from the mass and sent for histopathology.

The primary component of the tumor, as seen under a microscope, was endodermal sinus tumor tissue, which had a typical reticular or glandular pattern. The tumor also contained normal pancreatic tissue and mature squamous epithelial cells. Immunohistochemical stains came out positive for Cytokeratin Cam 5.2(suggestive of adenocarcinomas), OCT 3/4, SALL-4, CD 30 and Glypican-30 (suggestive of EST/YST), and Ki-67 marker was also noted to be raised. Based on the immunohistochemical and histological findings, a diagnosis of malignant endodermal sinus tumor was made. Additional clinical and ultrasound exams of this patient's genital system were carried out in an effort

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to rule out the presence of a primary gonadal germ cell tumor, but nothing abnormal was discovered.

The laboratory tests showed a high level of α-fetoprotein (AFP) > 1264 ng/ml (reference: <20 ng/ml). The carbohydrate antigen (CA) 19-9, LDH, β-HCG, blood amylase, aspartate transferase (AST), alanine transferase (ALT) and serum bilirubin levels were within the normal ranges.

Pre-contrast and contrast-enhanced spiral CT of the abdomen was performed, including the arterial phase, portal venous phase and equilibrium phase following bolus injection of intravenous contrast material. A heterogenous, solitary mass was identified in the pancreatic head which was locally compressing on the adjacent part of the duodenum (D2) along with mesenteric lymphadenopathy at station 1. Pancreatic mass was found to be only externally compressing the duodenum and causing the obstructive symptoms without any mucosal invasion. Pancreatic tumor was deemed unresectable as it was completely encasing superior and inferior mesenteric arteries (Fig. 1).

Clinical and radiological findings were also supportive of pancreatic malignancy. Exploratory laparotomy was performed for relieving the obstructive symptoms and uncut Roux-en-Y gastrojejunostomy was done. There was no evidence of intraperitoneal or retroperitoneal spread. Upon recovery, the obstructive symptoms improved drastically.

Once the patient recovered; he was referred to the oncology department for neoadjuvant chemotherapy. He was started on cisplatin (Platinol), etoposide phosphate, and bleomycin sulfate, also called PEB. After receiving 4 cycles of PEB, a CT scan was repeated which showed significant reduction in the size of lesion in the pancreatic head (Fig. 2).

Two months after the fourth chemotherapy cycle, exploratory laparotomy was done to remove the residual tumor and a standard pancreaticoduodenectomy was performed. The entire common bile duct distal to the biliary confluence, gallbladder, head of the pancreas, and duodenum were resected. Bilio-enteric and pancreatico-enteric continuity was restored using a Roux-en Y loop. Entire tumor was resected with free tumor margins on all sides, confirmed with biopsy (Fig. 3).

On post-operative day 2, the patient developed tachypnea and was unable to maintain oxygen saturation on room air above 84–86 %. He had developed complicated nosocomial pneumonia and was shifted to the ICU for intensive care and respiratory support. He was initially started on NIV but later developed pneumothorax in the left lung due to barotrauma, for which chest tube was passed. Patient was also intubated and put on ventilatory support until the lungs expanded completely. Complications resolved over a week and he was gradually weaned off from the ventilator, and extubated eventually. No other complications were encountered and the patient had full recovery. On the 14th post-operative day, the patient was discharged from the hospital. Follow up CT scan at two months showed no recurrence of tumor.

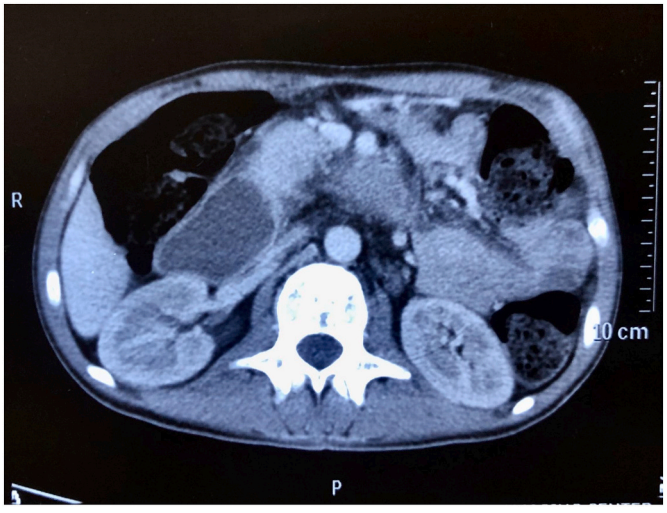

Fig. 1. Pre-chemotherapy CT scan of the patient shows distended stomach and dilated pylorus due to distal obstruction in the duodenum.

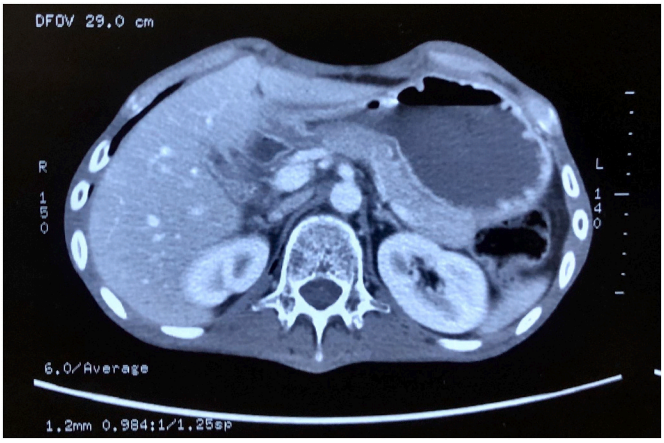

Fig. 2. CT scan film of the patient after 4 cycles of PEB Chemotherapy shows shrunken tumor.

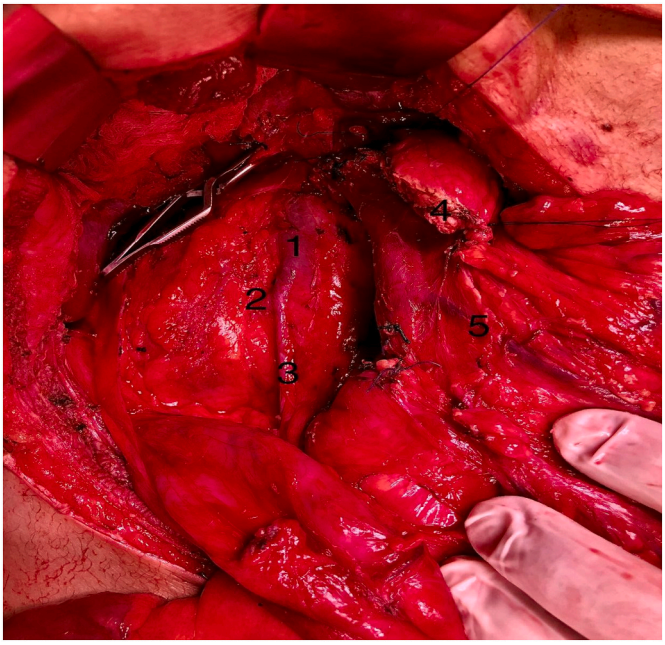

Fig. 3. Exhibiting the abdominal anatomy post pancreatic head tumor resection. (1) Inferior vena cava (2) right renal vein (3) gonadal vein (4) residual pancreas (5) upper colic branch of superior mesenteric vein.

3. Discussion

GCTs arising in the region of the pancreatic head are extremely rare hence making preoperative diagnosis of extra-gonadal mixed GCT extremely challenging [8]. According to reports, endoscopic ultrasonography guided fine needle aspiration (EUS-FNA) is a useful diagnostic technique for examining pancreatic mass lesions [9].

The most prevalent GCTs are teratomas, which can be classified as mature or immature. Neuroectoderm and less differentiated tissue are present in immature teratomas, which range in grade from 1 to 3 [7]. Endodermal sinus tumors are rare and highly malignant germ cell tumors (GCTs) [10]. Large, smoothly marginated, well-enhancing solid masses with a cystic, hemorrhagic, or necrotic part are frequent CT

findings of endodermal sinus tumors. However, no comprehensive study of CT features exists due to their rarity [3]. For the evaluation of ESTs, endoscopic ultrasonography-guided fine needle aspiration may be a useful diagnostic technique. The specimen's histopathology offers a conclusive diagnosis. Schiller-Duvall bodies, which are composed of a monolayer of cubic or columnar neoplastic cells encircling the capillaries, thin-walled blood sinus, or small venous blood arteries, are characteristic architectural patterns that distinguish endodermal sinus tumors from other types of tumors [11]. TP53 and KRAS are the two genes that YSTs most typically have altered, according to genome analysis [12].

Modern neoadjuvant chemotherapy with cisplatin, etoposide, and bleomycin (PEB) has considerably improved the survival of GCT patients [13–16]. Patients with abdominal and retroperitoneal GCTs had excellent prognosis, with an overall survival rate of 87.9 % ± 9.3 % and a 6-year event-free survival rate of 82 % ± 10.9 %, respectively [17]. Surgical excision with combined adjuvant chemotherapy is considered the treatment of choice [3]. However, many patients present with advanced local disease and distant metastasis.

In conclusion, we present a very rare case of malignant extra gonadal germ cell tumor in the pancreas in an adolescent. In order to choose the best course of treatment, a thorough differential diagnosis must be made. It is to be noted that despite not very high levels of AFP compared to other studies, histological and immunohistochemical analysis confirmed EST hence the possibility should be kept in differentials. This unusual instance is very important for raising surgeons' and oncologists' awareness of the malignant extra-gonadal GCTs diagnosis.

Human subjects

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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None.

Ethical approval

Hayatabad Medical Complex, Ethical Review Board approved the synopsis on "Endodermal Sinus tumor with Pancreatic Origin: A Case Report".

Author contribution

All the authors played a substantial role from conceptualisation of the paper to completion of the manuscript and have been given the authorship accordingly.

Registration of research studies

None.

Guarantor

Dr. Musarrat Hussain.
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Declaration of competing interest

The authors have no conflicts of interest to declare. All co-authors have seen and agree with the contents of the manuscript and there is no financial interest to report. We certify that the submission is original work and is not under review at any other publication.

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