## Table of Contents

### Original Articles

**Outcomes and Impact of a Universal COVID–19 Screening Protocol for Asymptomatic Oncology Patients** .................................................. 07  

**Onset of Arterial and Venous Thrombosis and Safety of Antithrombic Therapy in Patients with Gastrointestinal Cancer** .................. 13  
Sayuri Hashimoto, Yoichiro Yoshida, Daisuke Kato, Fumihiro Yoshimura, Suguru Hasegawa

**Cytoreductive Surgery plus Hyperthermic Intrapertitoneal Chemotherapy in Primary Advanced Ovarian Cancer: The First Reported Pilot Experience from Saudi Arabia** ........................................................................................................ 19  
Ahmed Abu–Zaid, Osama Alomar, Mohammd Abuzaied, Mohammad Alsabban, Hary Salem, Ismail A. Al–Badawi

**Frequency and Characteristics of Incidental Pulmonary Embolism in Cancer Patients: A Retrospective Study at a Large Oncology Center in Saudi Arabia** .................................................................................................................. 26  
Mohammed W. Althobaiti, Abdullah M. Al Jehani, Mohammd S. Alqarni, Ziad M. Bukhari, Azzam A. Khankan

**Trends in Thyroid Cancer Incidence in the Gulf Cooperation Council States: a 15–Year Analysis** .......................................................... 31  
Eiman Alawadhi, Amal Al–Madouj, Ali Al–Zahrani

**An Overview of Medullary Thyroid Cancer Cases Treated at Kuwait Cancer Control Center** .............................................................. 39  
Jitendra Shete, Khaled Al Saleh, Reham Safwat, Ahmed Bedair, Mustafa El Sherify, Amany Hussein, Marwa Nazeeh, Asit Mohanty

**Frequency and Characteristics of Incidental Pulmonary Embolism in Cancer Patients: A Retrospective Study at a Large Oncology Center in Saudi Arabia** .................................................................................................................. 26  
Mohammed W. Althobaiti, Abdullah M. Al Jehani, Mohammd S. Alqarni, Ziad M. Bukhari, Azzam A. Khankan

**Lung Cancer in Bahrain: Histological and Molecular Features** ................................................................................................................. 48  
Aalaa Mubarak, Eman Aljufairi, Sayed Ali Almahari

**Current Status of Cancer–Related Pain and Opioid use in South Lebanon: A Pilot Study** ................................................................. 52  
Fadi S. Farhat, Mohamed Tarabey, Feras Chehade, Tarek Assi, Joseph Kattan

**Adjuvant Irradiation in Carcinoma Breast Patients: Comparison of 3DCRT and Semi–automated Complex VMAT Hypofractionated Plans** ......................................................................................................................... 58  

**Relationships between AML1–ETO and MLL–AF9 fusion gene expressions and hematological parameters in acute myeloid leukemia** ................................................................................................................. 65  
Abdel Rahim Mahmoud Muddathir, Tariq A. M. Hamid, Elwaleed Mohamed Elamin, Omar F Khabour

### Review Articles

**Cancer Risk Factors Among Omanis: A Review** ................................................................................................................................. 70  
Abdul Hakeem Alrawahi, Asia AIIaamani, Najia A. Al–Lawati

**Surgical Management of Bone Sarcomas with an Inappropriate Biopsy Site: A Case Series and Review of Literature** .......................... 78  
Subbiah Shanmugam, Syed Afroze Hussain, Kishore Kumar Reddy

### Case Reports

**Basaloid Nasopharyngeal Carcinoma: An Entity That Remains Oblivious** ............................................................................................... 83  
Satesh Kumar Ganeson, Jeyasakthy Sanasiya, Irfan Moghadam, Norhaslinda Abdul Gani

**Pancreatoblastoma: A Case Report** ......................................................................................................................................................... 87  
Turki Alhazmi, Ahmed Abduljabbar, Mohammed Alem, Amr Mansouri, Khaled Bahubalshi

### Conference Highlights/Scientific Contributions

- **News Notes** ..................................................................................................................................................................................... 91
- **Scientific events in the GCC and the Arab World for 2020** .................................................................................................................. 95
Case Report

Pancreatoblastoma: A Case Report

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Abstract

Pancreatoblastoma (PB), also known as infantile pancreatic carcinoma, is an exceedingly rare pancreatic tumor in childhood, which is considered a malignant exocrine pancreatic tumor. Some cases have been reported in Saudi Arabia. Although PB primarily presents during childhood, it may occur in adults too. PB tends to be less hostile in infants and children when compared to adults. Histologically, PB is characterized by distinguished acinar and squamoid cell differentiation. Most of these tumors develop in the head of the pancreas and increase alpha-fetoprotein in up to 68% of patients. Ultrasound and CT scan play a significant role in preoperative diagnosis, which is often quite difficult. The best treatment is surgical removal of the pancreas. The role of adjuvant chemotherapy or radiotherapy is still under consideration due to a minor number of patients treated. Chemotherapy regimens consisting of cyclophosphamide, etoposide, doxorubicin, and cisplatin have been used in the neoadjuvant setting with anecdotal benefit. It was found that a poorer prognosis was associated with patients who had metastasis, and patients who could not be operated on surgically.

Keywords: Pancreatic neoplasm—Pancreatoblastoma

Introduction

Pancreatoblastoma is an extremely rare pancreatic tumor in childhood. It was reported for the first time in 1957. Pancreatoblastoma usually affects children ranging from one to eight years old and also may be seen in elderly individuals. It is usually misdiagnosed as neuroblastoma or hepatoblastoma in children under the age of 7 years. Partially differentiated acini of fetal origin will be noticed in pathology analysis of pancreatoblastoma. The presenting symptoms are palpable mass with anorexia, vomiting, constipation and abdominal pain. The tumor is clinically silent and large in size at the time of presentation. The symptoms are mostly due to the mass effect and include abdominal pain, vomiting and constipation.

Case Presentation

A 9-year-old girl presented with abdominal pain and abdominal mass. The abdominal pain started three years ago, it was mild and nonspecific. The patient was admitted many times as a case of abdominal pain for investigation that was treated as urinary tract infections. Later on, the pain increased in severity with associated weight loss. Upon examination, she looked sick, in pain and cachectic. Cardiothoracic and CNS examinations were unremarkable. Abdominal examination demonstrated a large mass occupying most of the left side of the abdomen. CT scan of the abdomen was performed, which showed an exophytic mass probably arising from the left liver lobe. The CT report offered the following differential diagnoses: primary liver lesions such as hepatoblastoma, HCC, and less likely mesenchymal tumor; Secondary liver lesions such as metastasis and lymphoma. Furthermore, pancreatic origin of the mass could not be entirely excluded due to the absence of fat planes between the mass and the pancreas. MRI of the liver was advised and pediatric surgery consultation was sent. MRI of the abdomen was done and it showed a large upper abdominal mass exerting mass effect on adjacent structures namely the liver, pancreas, and stomach with no clear invasion to the adjacent vessels. This mass, according to the MRI, was likely originating from the pancreas, however hepatic origin could not be totally excluded. Further clinical evaluation, surgical consultation, and histopathological correlation was advised. Her laboratory work—up including chemistry was

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unremarkable except for an elevated alpha-fetoprotein. A pediatric surgeon assessed the patient and took a tissue biopsy. The biopsy histopathology report confirmed the diagnosis of pancreatoblastoma – stage III.

The patient received chemotherapy as per PLADO protocol as tolerated. After seven months, CT scan was repeated for the patient (Figures 5, 6, and 7) and showed significant reduction in mass size that was followed by surgical resection. The surgery showed that the tumor was arising from the proximal part of the body of the pancreas, emerging through the lesser omentum, involving the lesser curvature of the stomach, and infiltrating the lower part of the hepatic segment 4 without lymphadenopathy (Figure 8).

**Discussion**

Pancreatoblastoma is an extremely rare primary neoplasm of childhood that has been seen in both young and elderly populations, but usually affects children ranging from 2–8 years old. Pancreatic tumors in children are exceptionally rare, causing less than 0.2% of pediatric deaths due to malignancy. The congenital form is associated with Beckwith–Wiedemann syndrome and has been characterized as a cystic form [9,11]. Patients with PB have been reported with an elevated Alpha-fetoprotein that could reach up to 68%. About 4% of the patients were reported to have the risk of malignancy (nephroblastoma, hepatoblastoma, rhabdomyosarcoma and pancreatoblastoma) that is associated with Beckwith–Wiedemann syndrome. Usually in ultrasound it demonstrates a large, well-defined, heterogeneous mass with solid and multilocular cystic components that contain hyperechoic septae. Dilatation of the biliary duct is not common as the tumors themselves are soft.

Although they are frequently large at presentation, CT findings usually show well defined, heterogeneous masses of solid and multilocular cystic components with enhancing septa, which may demonstrate fine calcifications similar to those seen in neuroblastoma. Generally, it occurs in the head of the pancreas, which...
Pancreatoblastoma is an extremely rare pancreatic tumor in childhood. It usually presents late with upper abdominal pain, and many have a palpable mass in the epigastrium. Alpha-fetoprotein may be elevated in up to 68% of patients.

Radiological findings are not specific. However, the location, the age and the imaging appearance are all helpful to reach the diagnosis. Histologically, PB is characterized by the presence of squamous corpuscles and aggregates or large epithelioid cells with formation of acini. Surgical resection is the optimal treatment method. Chemotherapy and radiotherapy are commonly used in the setting of metastasis or recurrence.

**Summary**

Pancreatoblastoma is an extremely rare pancreatic tumor in childhood. It usually presents late with upper abdominal pain, and many have a palpable mass in the epigastrium. Alpha-fetoprotein may be elevated in up to 68% of patients.

Radiological findings are not specific. However, the location, the age and the imaging appearance are all helpful to reach the diagnosis. Histologically, PB is characterized by the presence of squamous corpuscles and aggregates or large epithelioid cells with formation of acini. Surgical resection is the optimal treatment method. Chemotherapy and radiotherapy are commonly used in the setting of metastasis or recurrence.

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