e-ISSN 2083–4640
ISSN 1507–1367

CLINICAL VIGNETTE

Reports of Practical Oncology and Radiotherapy
2022, Volume 27, Number 5, pages: 929–930
DOI: 10.5603/RPOR.a2022.0095
Submitted: 04.07.2022
Accepted: 04.08.2022

Pancreatic ganglioneuroma in a young female

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Key words: pancreas; pancreatic ganglioneuroma; ganglioneuroma

Rep Pract Oncol Radiother 2022;27(5):929–930

A 24-year-old female was admitted to the hospital complaining of back pain for a few months. She had no history of a disease. She denied complaints of early satiety, weight loss, jaundice, or fever. The clinical examination was unremarkable. Her blood work was all normal. The patient was tested negative for catecholamine excess. Tumor markers were normal as well.

She underwent contrast-enhanced computerized tomography (CT) imaging which showed a well-defined solid mass in the porta hepatis (Fig. 1). A magnetic resonance imaging (MRI) scan was obtained for further characterization of the lesion. MRI axial images showed a well-defined solid mass in the porta hepatis. The mass demonstrated heterogenous enhancement on the delayed postcontrast sequences (Fig. 2).

Fine needle aspiration was ordered for diagnosis. It demonstrated a neoplasm that consists of spindle-shaped cells. Surgical removal of the lesion was performed, with negative margins. Postoperative pathology showed tumor cells that are positive for S-100 protein, neurofilament (NF), and synaptophysin; while negative for smooth muscle actin (SMA), desmin, CD117, and DOG-1; confirming the diagnosis of ganglioneuroma involving the pancreatic tissue.

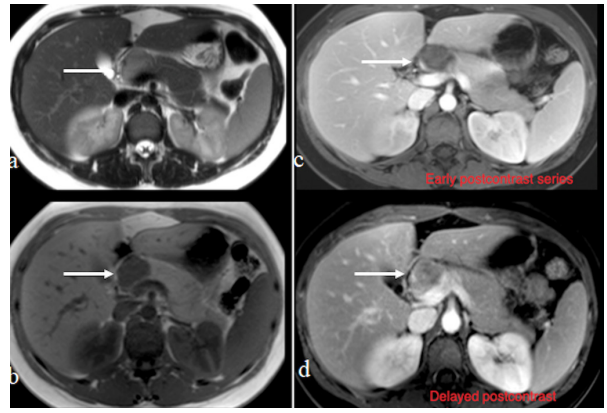

Figure 1. Precontrast and postcontrast computed tomography (CT) scan: axial and coronal images demonstrate a 3.5 × 2.5 cm homogenous low attenuation mildly enhancing well defined solid mass in the porta hepatis (white arrows) with the common hepatic artery, proper hepatic artery, and gastroduodenal artery closely associated with/draped over this mass. The mass abuts the main portal vein and involves the anterior neck of pancreas. No main pancreatic ductal dilatation

Postoperative hospitalization was uneventful. Repeat CT after 6 months revealed no signs of recurrence.

Ganglioneuroma is a benign type of peripheral neuroblastic tumor. It is important to differentiate ganglioneuromas from malignant components of neural crest cell-originated tumors [1].

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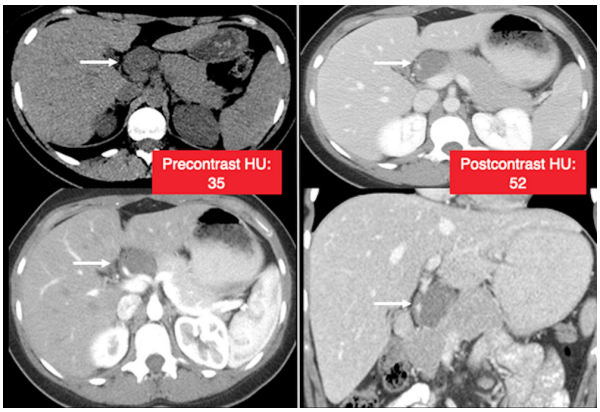

Figure 2. Magnetic resonance imaging (MRI) axial images demonstrate a 3.5 x 2.5 cm T1 hypointense T2 heterogeneously hyperintense well defined solid mass in the porta hepatis abutting the main portal vein which involves the pancreatic neck. The mass demonstrates heterogenous enhancement on the delayed postcontrast sequences (white arrows on image d)

Ganglioneuromas are usually asymptomatic and discovered incidentally. Symptoms, if present, occur due to the growth of the lesion and compression of the surrounding tissues and organs [2].

The precise diagnosis is made by histopathological examination. Ganglioneuroma consists of spindle cells and ganglion cells. The lack of immature neuroblasts, nuclear polymorphism, necrosis, and inflammation is useful for differentiating ganglioneuroma from malignant ganglioneuroblastoma and neuroblastoma. Immunohistochemistry is also helpful and positivity for S-100 protein, neurofilament (NF), and synaptophysin show the tumor's neurogenic origin [3]. Undifferentiated components detected by the endoscopic ultrasound (EUS)-guided biopsy cause preoperative diagnostic limitations.

The pancreatic location of ganglioneuroma is extremely uncommon and less than 10 cases are known from medical literature [1].

The pancreatic location of ganglioneuroma is usually seen in children and young adults [1].

Pancreatic ganglioneuroma has no specific imaging features. CT provides accurate anatomical details, and relationship to the vascular structures [4]. On contrast-enhanced CT, the lesion is usually non-enhanced. Delayed heterogeneous contrast enhancement may be observed. On MRI imaging, the lesion commonly appears as a T1 hypointense, T2 heterogeneously hyperintense mass. Gradually increasing enhancement is observed in dynamic studies [4]. In contrast, higher density on non-enhanced CT and lower hyperintensity on T2 weighted images are seen in ganglioneuroblastoma and neuroblastoma [4]. Discrete and punctate calcifications may also be observed in ganglioneuromas. On the other hand, amorphous and coarse calcifications are more likely to be seen in malignant ganglioneuroblastoma and neuroblastoma [4]. When a solitary, well-demarcated mass occurs in the porta hepatis, differential diagnosis should include the following tumors and conditions: lymphadenopathy, lymphoma, giant lymph node hyperplasia, tuberculosis, leiomyomas, nerve sheath tumors, hemangiomas, and stromal tumors of the gastrointestinal tract.

The case we present here is remarkable as a rare case of pancreatic ganglioneuroma in a young female. The preoperative diagnosis is challenging due to the lack of specific imaging features and the confusing aspects of histopathology. Definitive diagnosis and treatment are only possible with complete surgical resection of the lesion.

Conflict of interest

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Funding

The authors report no involvement in the research by the sponsor that could have influenced the outcome of this work.

Ethical statement

Informed consent was obtained from the patient for the publication of this case report.

References

1. Mazzola M, Bertoglio C, Achilli P, et al. Pancreatic ganglioneuroma: A rare entity with a difficult approach: A case report and systematic review. Dig Med Res. 2019; 2.
2. Poves I, Burdío F, Iglesias M, et al. Resection of the uncinate process of the pancreas due to a ganglioneuroma. World J Gastroenterol. 2009; 15(34): 4334–4338, doi: 10.3748/wjg.15.4334, indexed in Pubmed: 19750580.
3. Christein J, Kim A, Golshan M, et al. Central Pancreatectomy for the Resection of Benign or Low Malignant Potential Neoplasms. World J Surg. 2003; 27(5): 595–598, doi: 10.1007/s00268-003-6848-4.
4. Otal P, Mezghani S, Hassissene S, et al. Imaging of retroperitoneal ganglioneuroma. Eur Radiol. 2001; 11(6): 940–945, doi: 10.1007/s003300000698, indexed in Pubmed: 11419166.