CASE REPORT

An infant with a pancreatic yolk sac tumour presenting with obstructive jaundice – A case report

K W S M Wijayawardhana[1], U I Wickramarachchi[2], S Jayathunga[1], V J Meegoda[3], D A A Gamage[3], S A Gunarathne[4], L H D M Somathilaka[5], A K Lamahewage[4]

[1] *Faculty of Medicine, University of Kelaniya, Sri Lanka*

[2] *Faculty of Graduate Studies, University of Kelaniya Sri Lanka*

[3] *Postgraduate Institute of Medicine, University of Colombo, Sri Lanka*

[4] *Lady Ridgeway Hospital for Children, Colombo, Sri Lanka*

[5] *National Cancer Institute, Maharagama, Sri Lanka*

Abstract

Yolk sac tumours (YST) are malignant germ cell tumours that might have gonadal or extragonadal location. Extra-gonadal germ cell tumours often show an axial distribution, such as brain, neck, mediastinum, vagina, and sacrococcygeal region. YST in the pancreas are extremely rare. Herein, we aim to describe a rare case of pancreatic YST in a child with anatomical significance.

Case report: An eleven-month-old girl presented with features of obstructive jaundice for a one-week duration. Examination revealed deep icterus, without palpable abdominal mass. Serum alpha-fetoprotein (AFP) was significantly increased with normal serum beta-hCG. Other investigations showed an obstructive pattern liver enzyme, and imaging showed a heterogeneously enhancing, well-defined lesion in the head and neck of the pancreas. Child underwent Whipple's surgery and received adjuvant chemotherapy. Histology and immunohistochemistry confirmed the diagnosis of YST. The patient's condition improved following surgery.

This case demonstrates the importance of considering pancreatic tumours as a differential diagnosis for obstructive jaundice in children. Histology and immunohistochemistry play a significant role in arriving at a definitive diagnosis. Early diagnosis and multidisciplinary management help improve the outcome.

Keywords: Yolk sac tumour, Pancreas, Whipple's surgery, Schiller–Duval bodies.

DOI: https://doi.org/10.4038/slaj.v9i2.285

Sri Lanka Anatomy Journal 2025; 9(II): 101–107

Corresponding author

KWSM Wijayawardhana
E-mail: sameeraw@kln.ac.lk

https://orcid.org/0000-0002-5725-8235

Introduction

Yolk sac tumours (YST), also known as endodermal sinus tumours, are malignant germ cell tumours. They usually arise from the gonads, ovaries or testes. Those can be of extra-gonadal locations, often with an axial distribution, such as the brain, neck, mediastinum, vagina, and sacrococcygeal region. However, YSTs in the pancreas are extremely rare.

These tumours originate from totipotent germ cells that aberrantly migrate during embryogenesis, leading to neoplasms in unusual anatomical sites. Pancreatoblastoma is the most typical pancreatic tumour in infants. The pancreas, a retroperitoneal organ with intricate anatomical relationships to the bile duct, duodenum, and central vasculature, plays a critical role in clinical presentations when affected by tumours. Symptoms of obstructive jaundice suggest compression or invasion of the biliary system, highlighting the anatomical impact of such lesions.

This case report presents a rare instance of a pancreatic YST in an infant presenting with obstructive jaundice, focusing on the anatomical, histological, and embryological findings.

Case presentation

This eleven-month-old girl of a primigravida mother had an uncomplicated antenatal period and was born by an elective caesarean section at term with a birthweight of 3.65 kg. She presented with features of obstructive jaundice for a one-week duration. Examination revealed deep icterus with no palpable abdominal mass. Serum alpha-fetoprotein (AFP) was significantly increased (3678.43ng/ml) with normal serum beta-hCG. Liver enzymes revealed an obstructive pattern.

USS of the abdomen revealed a 4x3x3 cm mass medial to the R kidney displacing the IVC posteriorly. CECT abdomen revealed a soft tissue lesion in the head of the pancreas, suggestive of a neurogenic or pancreatic tumour. MRI abdomen revealed a heterogeneously enhancing, well-defined lesion in the head and neck of the pancreas, medial to the 2nd part of the duodenum, measuring 41mm in diameter (Figure 1). The pancreatic duct was dilated.

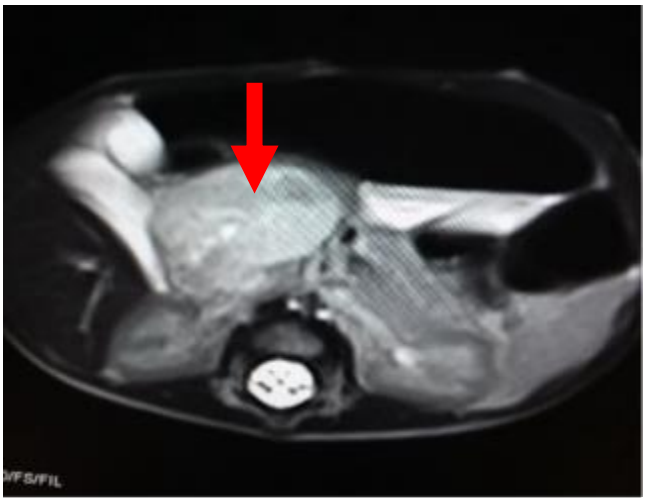

Figure 1: Magnetic Resonance Imaging (MRI) of the abdomen showing tumour in the head of the pancreas (Red arrow - Tumour in the head of the pancreas causing obstruction to CBD)

The child underwent Whipple's surgery and received adjuvant chemotherapy. A histology specimen of Whipple's received, containing the distal stomach, duodenum and head of the pancreas, was examined. Sections reveal a

well-encapsulated tumour with a loose reticular lace-like pattern. Papillary and cord-like patterns of cuboidal and elongated cells were presented with vascular nuclei and moderate cytoplasm. Schiller–Duval bodies around a central capillary were present (Figure 2).

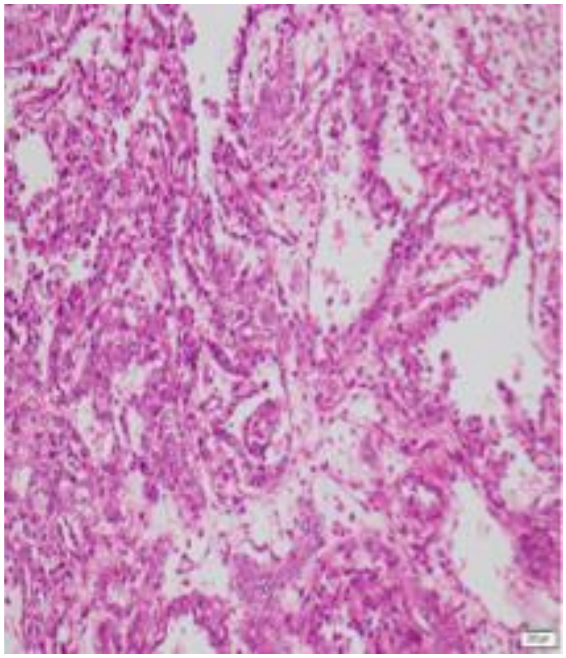

Figure 2: H&E staining of the encapsulated tumour with a loose reticular lace-like pattern. Papillary and cord-like patterns of cuboidal and elongated cells were present with vascular nuclei and moderate cytoplasm. Schiller–Duval bodies around a central capillary were present showing cellular proliferation and spindle cells.

The tumour did not involve the duodenum with clear resection margins. There had been no evidence of vascular and neural invasion. Immunohistochemistry revealed strong positivity for AFP and CK, and PLAP was negative, which confirms the diagnosis of YST (Figure 3). The patient's condition improved following surgery. The child is currently doing well and is being followed up on with AFP levels and USS.

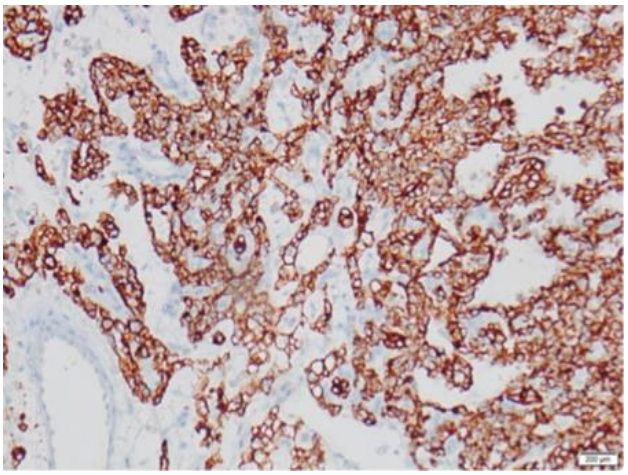

Figure 3: Immunohistochemistry of the tumour showing strong positivity for AFP

Discussion

The pancreatic yolk sac tumour (YST) in this child was located in the head of the pancreas. The pancreatic head is connected to the C-shaped concavity of the duodenum, closely related to the common bile duct, pancreatic duct, and vasculature, Inferior vena cava (IVC) and renal veins. (1) Imaging revealed a 4 cm mass compressing the common bile duct, which explained the clinical presentation of obstructive jaundice. The tumour's proximity to the duodenum and its partial encasement of the pancreatic duct were evident during surgical exploration, complicating resection due to the risk of damaging these structures. While no anatomical variations, such as an aberrant pancreatic duct or accessory bile duct, were observed, the tumour's size and location have displaced the superior mesenteric vein laterally. It is an unusual finding that posed challenges during the surgical exploration. The complex retroperitoneal anatomy of the pancreas underscores the importance of precise imaging to identify the tumour size, location, and impact on the biliary and pancreatic systems. Multimodal imaging,

including ultrasonography (USS), contrast-enhanced computed tomography (CECT), and magnetic resonance imaging (MRI), plays a crucial role in localising pancreatic masses and assessing local invasion. (2)

Only a few cases of pancreatic yolk sac occurring in infants were reported in the literature. The first extragonadal YSTs in the pancreaticobiliary region were reported in an 11-month-old girl presenting with obstructive jaundice, who was found to have a primary YST of the common bile duct. (3) In contrast, Wang et al. described a 12-month-old girl with a primary mixed germ cell tumour of endodermal sinus and mature teratoma in the pancreatic head. (4) This limited literature highlights the rarity of germ cell tumours in the pancreaticobiliary system in infants.

Given the tumour's location in the pancreatic head and its presentation with obstructive jaundice, a detailed understanding of biliary system anatomy becomes crucial for both clinical comprehension and surgical management. The common bile duct passes through or adjacent to the pancreatic head before opening into the duodenum at the ampulla of Vater. This close anatomical relationship makes it liable for patients to develop obstructive jaundice when a space-occupying lesion develops in this region. (5, 6)

The extrahepatic biliary system consists of right and left hepatic ducts, which unite with each other to form the common hepatic duct, then receives the cystic duct to form the common bile duct, which ultimately drains to the second part of the duodenum. The common bile duct measures approximately 7-11 cm in length and 4-8 mm in diameter. It is described in four anatomical segments, which are supraduodenal (2-3cm), retroduodenal (2-3cm), pancreatic(2-4cm), and intraduodenal portions (1-2 cm) (7). The Pancreatic portion, which passes through or in a groove in the posterior surface of the pancreatic head, is surgically significant in pancreaticoduodenectomy procedures. (Diagram 1)

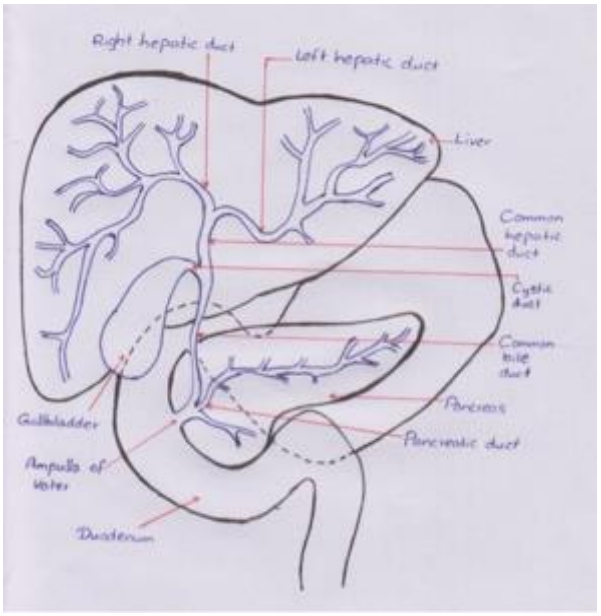

Diagram 1: Biliary tree

The arterial blood supply of the extrahepatic biliary system is by an artery network called 'epicholedochal plexus' that lies along the lateral border of the common bile duct. (Diagram 2) In extensive dissection, it can lead to biliary ischemia and anastomotic complications. (8). The biliary system exhibits anatomical variations in approximately 20-25% of the population, which can significantly impact the planning of surgery. (9) Low confluence of hepatic ducts, aberrant right hepatic duct, and short cystic artery are some common anatomical variations.

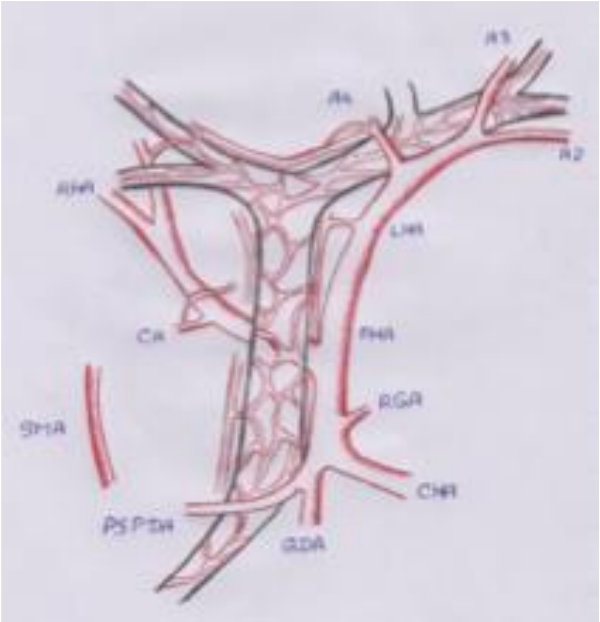

Diagram 2: Arterial blood supply of extrahepatic biliary system; SMA: Superior Mesenteric Artery; GDA: gastroduodenal artery; CHA: Common hepatic artery; PSPDA: Posterior superior pancreaticoduodenal artery; RGA: Right gastric artery; PHA: Proper hepatic artery; LHA: Left hepatic artery; CA: Cystic artery; RHA: Right hepatic artery; a2: artery to laterosuperior segment; a3: artery to lateroinferior segment; a4: artery to medial segment.

In our patient, Preoperative imaging did not reveal a biliary anatomical variation that would facilitate surgical planning. However, a tumour compressing on the common bile duct created a similar functional challenge to anatomical stenosis, requiring attention in preserving the common bile duct continuity in surgery.

Pancreatic yolk sac tumours represent an extraordinary rarity in pediatric oncology. Yolk sac tumour is the predominant variant of germ cell tumours in infants and young children and can manifest in unusual locations, including the abdominal wall and other extragonadal sites. Billmire et al. reported 25 children with abdominal and retroperitoneal malignant germ cell tumours; some had YST histology but did not specify pancreatic involvement. The most common malignant pancreatic tumour in young children is pancreatoblastoma. (10) It has a similar clinical profile and presentation. Case descriptions for pancreatoblastoma describe the occurrence in young children with a mean age of five years. (11)

Histopathological features of the tumour in our patient showed similar microscopic findings in a 70 years old woman with pancreatic YST, presence of schiller-Duval bodies which is a pathognomonic feature of a YST and with strong positivity of AFP. (12) Histopathological examination of the tumour confirmed characteristic features of a YST. Schiller-Duval body is pathognomonic for yolk sac tumours. (13) Strong immunohistochemical positivity for alpha-fetoprotein (AFP). These findings are diagnostic of YST and distinguish it from other pancreatic neoplasms, such as pancreatoblastoma. Schindewolf et al., describe a diagnosis of testicular yolk sac tumour with similar histological and immunohistochemical findings (14). The embryological basis of pancreatic yolk sac tumours is related to the aberrant migration of primordial germ cells during early development. Usually, these cells migrate from the yolk sac to the gonadal ridges between the fourth and sixth weeks of embryogenesis. (15) In our patient, ectopic germ cells are likely to persist in the pancreatic primordium, giving rise to the tumour. This developmental anomaly contrasts with gonadal YSTs, which arise from germ cells in their

typical location. While gonadal YSTs share similar histological features, the pancreatic location is unique due to its rarity and the absence of germ cell tissue in the pancreas under normal conditions, making this case exceptional.

Paediatric germ cell tumours account for approximately 3.5 % of all childhood cancers, with one-third being extragonadal. Paediatric pancreatic yolk sac tumours are sporadic. Rogerro et al reported a 12-year-old girl with ovarian YTS and co-occurrence of a pancreatic solid pseudopapillary neoplasm. (16) Akker et al report YST in the abdominal wall of an 18-month-old girl, with similar histological features. (17) However, there are case reports of pancreatic YST in adults. A 33-year-old male presented with abdominal pain, vomiting, and weight loss without prominent clinical features of obstructive jaundice, and blood tests revealed an obstructive jaundice pattern. (18) The diagnosis was found to be a pancreatic yolk sac tumour. The patient has undergone resection and chemotherapy and was found to be stable in a five-year follow-up.

This case describes an infant with pancreatic YST presented with obstructive jaundice, with emphasis on anatomical findings, including the tumour's location in the pancreatic head, its compression of the common bile duct, and its displacement of the superior mesenteric vein. Histologically, the presence of Schiller-Duval bodies and AFP positivity confirmed the diagnosis, while embryologically, the tumour's origin from ectopic germ cells represents a rare developmental anomaly. This case holds an educational value for anatomical sciences, particularly related to the impact of rare tumours on pancreatic and biliary anatomy. The detailed description of the anatomical relationship of the tumour enhances understanding of how mass lesions disrupt normal pancreatic function and biliary flow. Sri Lanka, despite the limited resources, has pediatric surgical and pathological expertise that is developing. This case serves as a valuable teaching material for recognising rare tumours in local healthcare settings, where access to advanced diagnostics is limited.

In fact, this case study has several limitations. The discussion primarily focused on anatomical details related to diagnosis, rather than detailed management. Additionally, the long-term follow-up was constrained due to the patient's clinical course, which limited insights into the outcomes. Molecular studies and advanced imaging techniques, such as 3D reconstruction of pancreatic anatomy, may further clarify tumour-structure interactions, aiding surgical planning.

Authors' contribution: SAG, LHDM, and LAM participated in the management of the patient and provided clinical care. SAG contributed to the histological and immunohistochemical diagnosis. KWSMW, UIW, SJ, VJM, and DAAG contributed to the article's writing and editing. All authors read and approved the case report prior to submission.

Conflicts of interest: The Authors declare no conflict of interest in preparing this article

Funding: None

References

1. Yuan Q, Pan A, Fu Y, Dai Y. Chapter 1 - Anatomy and physiology of the pancreas. In: Li M, Lu L, Xiao Y, Fu D, Zhang H, editors. Integrative Pancreatic Intervention Therapy: Elsevier; 2021. p. 3-21.

2. Scialpi M, Reginelli A, D'Andrea A, Gravante S, Falcone G, Baccari P, et al. Pancreatic tumors imaging: An update. International Journal of Surgery. 2016;28:S142-S55.

3. Munghate GS, Agarwala S, Bhatnagar V. Primary yolk sac tumor of the common bile duct. J Pediatr Surg. 2011;46(6):1271-3.

4. Wang J, Zheng Z, Qiu Y, Tou J, Liu W, Xiong Q, et al. Primary mixed germ cell tumor arising in the pancreatic head. J Pediatr Surg. 2013;48(1):e21-4.

5. Brahmi SA, Khattab M, Mesbahi OE. Obstructive jaundice secondary to pancreatic head adenocarcinoma in a young teenage boy: a case report. Journal of Medical Case Reports. 2011;5(1):439.

6. Bertens KA, Livingston M, Quan D, Leslie K, Zorzi A, Bütter A. Neuroendocrine tumor of the pancreas causing biliary obstruction in a 12 year-old girl: A case report and literature review. Journal of Pediatric Surgery Case Reports. 2014;2(9):413-6.

7. Dawson PM, Allen-Mersh TG. The anatomical relationship between the retropancreatic part of the bile duct and the main pancreatic duct. Ann R Coll Surg Engl. 1983;65(3):188-90.

8. Nagino M, Kamiya J, Nishio H, Ebata T, Arai T, Nimura Y. Two hundred forty consecutive portal vein embolizations before extended hepatectomy for biliary cancer: surgical outcome and long-term follow-up. Ann Surg. 2006;243(3):364-72.

9. Buddingh KT, Nieuwenhuijs VB, van Buuren L, Hulscher JB, de Jong JS, van Dam GM. Intraoperative assessment of biliary anatomy for prevention of bile duct injury: a review of current and future patient safety interventions. Surg Endosc. 2011;25(8):2449-61.

10. Patterson KN, Trout AT, Shenoy A, Abu-El-Haija M, Nathan JD. Solid pancreatic masses in children: A review of current evidence and clinical challenges. Frontiers in Pediatrics. 2022;Volume 10 - 2022.

11. Glick RD, Pashankar FD, Pappo A, Laquaglia MP. Management of pancreatoblastoma in children and young adults. J Pediatr Hematol Oncol. 2012;34 Suppl 2:S47-50.

12. Yonemaru J, Takahashi M, Nara S, Ichikawa H, Ishigamori R, Imai T, Hiraoka N. A yolk sac tumor of the pancreas and derived xenograft model effectively responded to VIP chemotherapy. Pancreatology. 2020;20(3):551-7.

13. Yolk sac tumor. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 [cited 2025 Jul 19]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK563163/.

14. Schindewolffs L, Dierks C, Heppelmann M, Gähle M, Piechotta M, Beineke A, et al. Testicular yolk sac tumor and impaired spermatogenesis in a Holstein Friesian calf. Systems Biology in Reproductive Medicine. 2015;61(5):314-9.

15. Zhang B, Gao S, Chen Y, Wu Y. Primary yolk sac tumor arising in the pancreas with hepatic metastasis: a case report. Korean J Radiol. 2010;11(4):472-5.

16. Roggero A, Piro L, Paoloni D, Palo F, Avanzini S, Capra V, et al. Co-occurrence of ovarian Yolk Sac Tumor and pancreatic solid pseudopapillary neoplasm in a pediatric patient: A case report. Pediatric Hematology Oncology Journal. 2025;10(2):100457.

17. van den Akker M, Vervloessem D, Huybrechs A, Declercq S, van der Werff Ten Bosch J. Yolk sac tumor in the abdominal wall of an 18-month-old girl: a case report. J Med Case Rep. 2017;11(1):47.

18. Galanis I, Floros G, Simou M, Kyriakopoulos G, Stylianidis G. An Extremely Rare Case of a Primary Pancreatic Yolk Sac Tumor. Cureus. 2022;14(6):e26007.