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A rare case of Ectopic Pancreatic Papillary Intraductal Mucinous Neoplasm in a 10-year-old child: A Case Report

Edrick Elias

mtalemwae@gmail.com

Catholic University of Health and Allied Sciences

Mary Ndibalema

Catholic University of Health and Allied Sciences

Gandom Dashtaban

Catholic University of Health and Allied Sciences

Subira Matiku

Catholic University of Health and Allied Sciences

Oscar Ottoman

Catholic University of Health and Allied Sciences

Raphael Mwita

Bugando Medical Centre

Peter Rambau

Catholic University of Health and Allied Sciences

Case Report

Keywords: Ectopic pancreas, pediatrics, IPMN, case report

Posted Date: September 23rd, 2026

DOI: https://doi.org/10.21203/rs.3.rs-10718356/v1

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Additional Declarations: No competing interests reported.

Abstract

Background

Ectopic pancreas is an uncommon condition, with an incidence of 0.2% identified during laparotomy and 0.5–13.7% during autopsy, with the most frequent location in the gastrointestinal tract. Intraductal papillary mucinous neoplasm (IPMN) is common in the native pancreas, accounting for 12% of pancreatic tumors in patients older than 80 years, but is rare at ectopic sites. To the best of our knowledge, this is the first pediatric case report to be reported in the literature.

Case presentation:

A 10-year-old female Sukuma presented at Bugando Medical Centre with chronic abdominal pain followed by constipation. Intraoperatively, the duodenal mass was identified, and the resection and anastomosis were done. A cystic and solid submucosal mass measuring 4.5 x 3 x 1.5cm was identified, and histology revealed pancreatic tissue with variable-sized cystic spaces and a few papillary projections lined by a single- to double-layer of intestinal-type epithelium with low-grade dysplasia. No recurrence or any symptom reported after one year of follow-up.

Conclusions

This is the first case reported on pediatric ectopic pancreatic intraductal papillary mucinous neoplasm, low grade, highlighting the consideration of this condition as a possible differential diagnosis in the pediatric population

Background

An ectopic pancreas, also referred to as a heterotopic pancreas, is a condition in which pancreatic tissue is found away from its usual anatomical location and lacks vascular, neural, or anatomical connections to the primary pancreas[1–3]. The pancreas is a retroperitoneal organ normally located posterior to the stomach, between the duodenum on the right and the spleen on the left. It overlies and transversely crosses the bodies of the first and second lumbar vertebrae in the posterior abdominal wall[4]. Ectopic pancreas is an uncommon condition, with an incidence of 0.2% identified during laparotomy and 0.5–13.7% during autopsy[1]. The most frequent location is the gastrointestinal tract, although occurrences have also been reported in the biliary system, lungs, liver, mediastinum, and brain. Within the gastrointestinal tract, the stomach is the most common site, followed by the duodenum and then the jejunum [3,5]. The development of ectopic pancreas is associated with abnormal embryogenesis, although the precise mechanisms underlying this association remain unclear. Three main hypotheses have been proposed: the dislocation or misplacement theory, the metaplasia theory, and the totipotent stem cell theory. The dislocation or misplacement theory suggests that during embryonic rotation of the dorsal

and ventral pancreatic buds, some pancreatic tissue becomes separated and is deposited in other regions of the developing gastrointestinal tract[6]. According to the metaplasia theory, endodermal tissue migrates into the submucosa and subsequently transforms into pancreatic tissue, but the totipotent stem cell theory posits that endodermal cells lining the gastrointestinal tract and other regions may differentiate into pancreatic tissue [6,7].

In most cases, an ectopic pancreas is an incidental finding; however, it may be symptomatic due to neoplastic transformation, which may include both benign neoplasms, including intraductal papillary neoplasms, and malignant neoplasms [8–11]. The occurrence of intraductal papillary mucinous neoplasm (IPMN) is common in the native pancreas, accounting for 12% of pancreatic tumors at the age above 80 year [12], but extremely rare in the pediatric population as only 3 cases have been reported, aged 12 and 18 years, respectively[13,14], and one case was reported in a neonate who had a de novo germline mutation in the SKIL gene[15]. All 24 cases of ectopic pancreatic intraductal papillary mucinous neoplasm (IPMN) reported in the literature were from adults ranging from 28 to 80 years [16,17], and five cases originated from the duodenum[16,17]. No ectopic pancreatic IPMN pediatric case has been documented in the literature. The literature review is summarized in Table 1.

Table 1
Literature review cases summary

S/N	Author, year, and reference	Age and gender	Presentation	Site	Diagnosis
1	Justin M. M. Cates, 2005[17]	73 years, Male	Acute lower abdominal pain, vomiting, and bloody diarrhea	Meckle diverticulum	Intraductal Papillary Mucinous Adenoma
2	Jason Phillips, 2006[18]	80 years, Male	Recurrent dyspepsia and intermittent diarrhea	Gastric	Intraductal papillary mucinous neoplasm
3	Ho Sung Park, 2008[19]	66 years, Male	Asymptomatic	Gastric and pancreas	Gastric intraductal papillary mucinous neoplasm and synchronous pancreatic papillary mucinous adenocarcinoma
4	Bård I Røsok, 2008[10]	59 years, Female	Upper abdominal discomfort	Duodenum	Intraductal papillary mucinous carcinoma
5	Niravkumar Patel, 2010[11]	44 years, Female	Abdominal bloating and epigastric pain in a person living with HIV	Gastric	Intraductal papillary mucinous neoplasm
6	Dimitrios Tsapralis, 2010[20]	60 years, Male	Epigastric pain and obstructive jaundice	Gastric and pancreas	Synchronous gastric and pancreatic Intraductal papillary mucinous neoplasm with high-grade dysplasia.
7	Ju Young Song, 2012[9]	74 years, Male	Jejunal perforation	Jejunum	Adenocarcinoma associated with intraductal papillary mucinous neoplasm
8	Hiroshi Okamoto, 2013[21]	75 years, Female	Incidental findings	Jejunum	Intraductal papillary mucinous neoplasm
9	Alistair J Lawrence, 2015[22]	47 years, Female	Right upper quadrant pain	Proximal hepatic duct	Intraductal papillary mucinous neoplasm
10	Sang Hwa Lee, 2015[23]	49 years, Female	Follow-up CT scan post-colon and endometrial cancer treatment	Ileum	Intraductal papillary mucinous neoplasm with high-grade dysplasia

S/N	Author, year, and reference	Age and gender	Presentation	Site	Diagnosis
11	Changqing Ma et al, 2015[24]	70 years, Male	Crohn's disease	Ileum	Intraductal papillary mucinous neoplasm with high-grade dysplasia
12	Changqing Ma et al, 2015[24]	74 years, Male	Ileocecal fistula	Ileum	Intraductal papillary mucinous neoplasm with high-grade dysplasia
13	Sarah M Choi et al, 2015[25]	54 years, Female	Metastatic neuroendocrine tumor	Duodenum	Intraductal papillary mucinous neoplasm
14	Sarah M Choi et al, 2015[25]	72 years, Male	Gastrointestinal reflux	Duodenum	Intraductal papillary mucinous neoplasm
15	Wade O Christopher et al, 2017[8]	Unknown	Unknown	Gastric	Invasive Pancreatic Adenocarcinoma Arising in Intraductal Papillary Mucinous Neoplasm
16	Michaela Cellina et al, 2017[26]	56 years, Female	Recurrent epigastric pain	Double sites: Duodenum and gastric	Double Intraductal Papillary Mucinous Neoplasm with multiple foci of adenocarcinoma
17	Eiji Noda et al, 2018[27]	54 years, Female	Abdominal pain	Jejunum	Intraductal Papillary Mucinous Neoplasm
18	Pietro Addeo, 2019[28]	69 years, Male	Renal disease	Gastric	Intraductal Papillary Mucinous Neoplasm
19	Etsuko Hisanaga et al, 2020[29]	70 years, Female	Asymptomatic	Duodenum	Adenocarcinoma with intraductal papillary mucinous neoplasm
20	G H Kim and D Y Park, 2021[30]	46 years, Female	Asymptomatic	Gastric	Intraductal Papillary Mucinous Neoplasm with low-grade dysplasia
21	Naoko Nambu et al, 2021[31]	71 years, Male	Epigastric pain	Gastric	Intraductal Papillary Mucinous Neoplasm

S/N	Author, year, and reference	Age and gender	Presentation	Site	Diagnosis
22	Jun-Hao Huang et al, 2023[32]	56 years, Female	Incidental findings, traumatic abdominal injury	Jejunum	Intraductal Papillary Mucinous Neoplasm
23	Yu Pang et al, 2023[33]	28 years, Male	Epigastric pain	Gastric	Intraductal Papillary Mucinous Neoplasm
24	Rinrada Worapongpaiboon et al, 2025[17]	73 years, Female	Chronic reflux	Gastric	Intraductal Papillary Mucinous Neoplasm
25	Edrick Elias	10 years, Female	Constipation	Duodenum	Intraductal Papillary Mucinous Neoplasm with low-grade dysplasia

Case Report

The present case report is of a 10-year-old female Sukuma who presented at Bugando Medical Centre with chronic abdominal pain for 2 years followed by constipation for two weeks. The family and past medical history were uneventful, with no previous history of surgery. The general examination was normal, and there was moderate abdominal tenderness. The abdominal ultrasound showed a duodenal mass with cystic and solid components. The abdominal CT scan and endoscopic studies were not done due to financial constraints. Intraoperatively, the duodenal mass was identified, and the resection and anastomosis were done. In the histopathology department, the bowel segment of 5 cm in length and a circumference of 2–5 cm was received. There was a cystic and solid mass in the submucosa measuring 4.5 x 3 x 1.5 cm, and the bowel mucosa was macroscopically normal. In histology examination, the duodenal mucosal epithelium and glands were normal, but in the submucosa and muscular layer, there was pancreatic tissue with variable-sized cystic spaces with few papillary projections. The cystic spaces were lined by a single to double layer of intestinal-type epithelium with low-grade dysplasia (Fig. 1). The immunohistochemical stains and molecular tests were not available in our setting.

After a year of follow-up, the patient reported no complaints.

Discussion

To the best of our knowledge is the first pediatric ectopic pancreas intraductal papillary mucinous neoplasm confirmed by histopathological examination to be described in the literature, but the lack of immunohistochemical and molecular tests in our setting is a significant limitation to this case report. In the reported 24 cases, even though the data are not enough to conclude, the female gender was predominantly affected by ectopic intraductal papillary mucinous neoplasm (IPMN) reported in adults,

12(52%%), excluding one case whose gender is unknown[17], and the abdominal pain without features of intestinal obstruction was the common clinical presentation[16,17]. This is different from our case, which is presented with abdominal pain followed by constipation. The intraductal papillary mucinous neoplasm (IPMN) is a precancerous lesion; as in the native pancreas, above 15% of pancreatic cancers arise from IPMN, with increased frequencies in the high-grade subtype[34,35]. In documented ectopic cases, 3 patients had intraductal papillary carcinoma[8,9,26]. In a systematic review, 4 patients with malignancy from other sites, including thyroid, gastric, colon, and pancreas, were reported to have ectopic intraductal papillary mucinous neoplasm[16] supporting a well-established genetic linkage between IPMN and cancer syndrome[34]However, in pediatric populations, pancreatic malignancies are rare; in our case, the risk of malignancy is likely to be low.

Despite having established radiological diagnostic criteria for IPMN[36], there is limited accessibility and affordability of diagnostic CT scans, MRI services, and endoscopic studies in Sub-Saharan countries, including Tanzania, which poses preoperative diagnostic challenges[37]. This case was histologically confirmed, which is a standard diagnostic criterion of ectopic IPMN, which requires tissue excision and histopathology evaluation of the resected specimen[17]. In all the literature on ectopic IPMN cases, the treatment modality was surgical resection[14,16,17,19], similar to the treatment approach used in this case. Being a pediatric patient and given the significant relationship between IPMN and cancer, genetic counselling and testing are crucial to our case. Although genetic testing services are not available in our setting, based on the epidemiology, this diagnosis is likely genetically predisposed, making genetic counselling necessary. Communication was made with the attending doctor to enquire about the family history of cancer and related risks for possible cancer counselling and screening. The patient did not report any complaints after one year of follow-up.

Conclusions

This is the first case to be reported on pediatric ectopic pancreatic intraductal papillary mucinous neoplasm, low grade, highlighting the consideration of this condition as a possible differential diagnosis in the pediatric population.

Abbreviations

BMC
Bugando Medical Centre
CT
Computed tomography
CUHAS
Catholic University of Health and Allied Sciences
IPMN
Intraductal Papillary Mucinous Neoplasm

IRB
Institutional review board
MRI
Magnetic resonance imaging

Declarations

Ethics and consent to participate

Witnessed informed consent and assent were obtained from the guardian and patient to participate in the clinical report. The case report does not require approval from the Joint CUHAS/BMC IRB

Consent to publish

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

Availability of data and materials

Clinical and pathological data for this case report can be made available upon formal request

Competing interests

All authors have no conflict of interest

Funding

No funds were received

Authors contributions

EE, MN , GD wrote the main text; RK case management; retrieval of patient information; PR, EE, SM, and OT examined the pathology specimen and reviewed the manuscript. All authors approved the final version of the manuscript.

Acknowledgements

N/A

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Figures

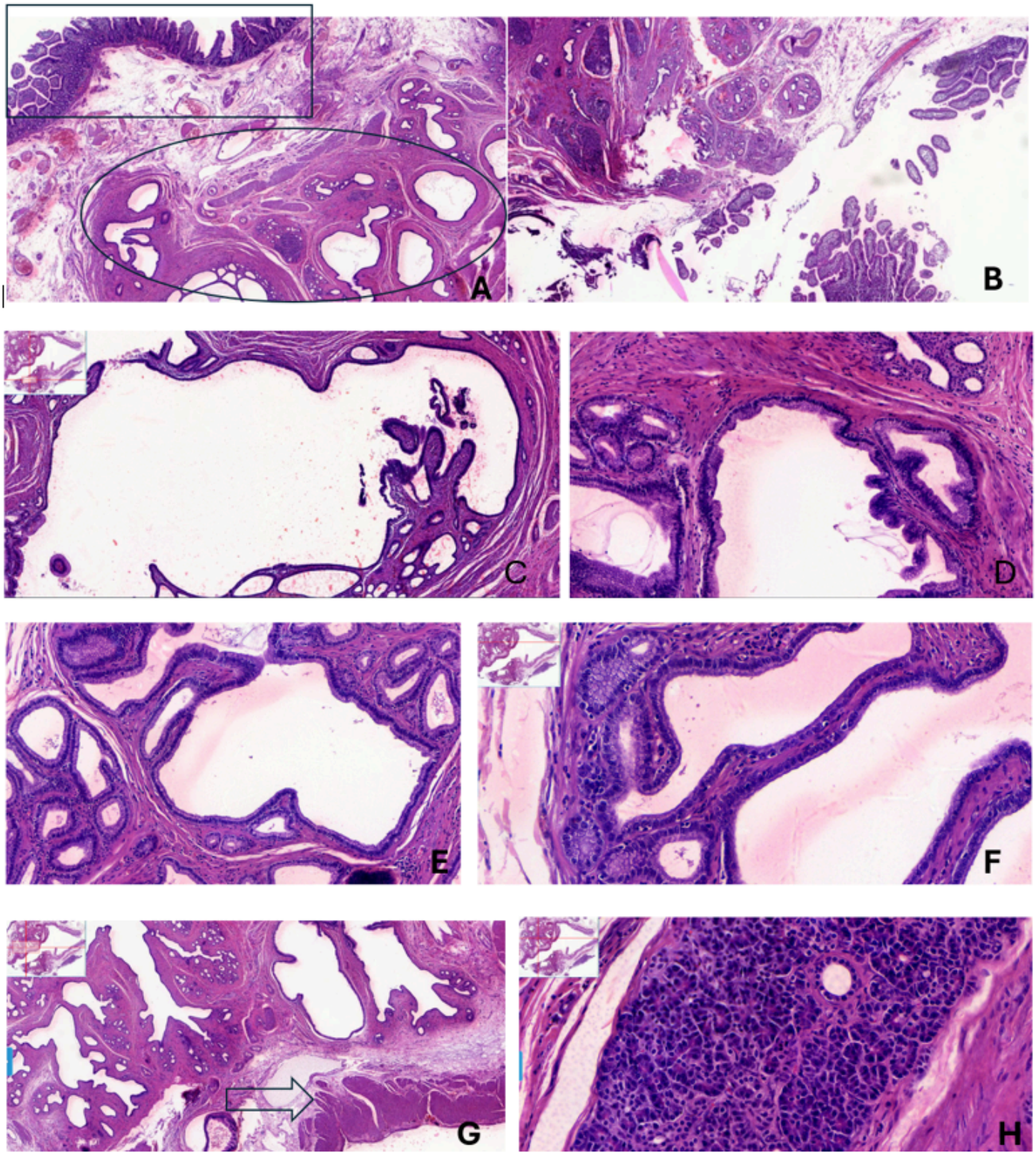


Figure 1

Histomorphology features of ectopic pancreatic intraductal papillary mucinous neoplasm in the duodenum: A:Square indicating normal duodenal mucosa, round shape indicating the duodenal submucosa with ectopic pancreatic tissue, and cystic pancreatic dilated ducts at x2, B: Other view of normal duodenal mucosa, ectopic pancreas and cystic dilated ducts at x2, C-F: Cystic dilated pancreatic ducts, and papillary projections all lined by intestinal type epithelium at x2, x20, x10 and x20 respectively,

G: Square arrow indicating normal duodenal muscular layer above which there is ectopic pancreatic tissue with cystic dilated ducts x2, and H is a normal pancreatic tissue at x10.