



MANAGEMENT OF A 5-YEAR-OLD PATIENT WITH PANCREATIC SEROUS CYSTADENOMA: A RARE CASE REPORT

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Abstract

Introduction: Pancreatic serous cystadenoma is a benign primary pancreatic cystic neoplasm that typically presents asymptotically. It is rare in the pediatric population and often poses diagnostic challenges.

Case Report: A 5-year-old girl presented to the emergency department at Arifin Achmad Hospital with severe pain in the left upper quadrant of the abdomen, which had persisted for 5 days. Clinical examination revealed swelling and erythema in the epigastric region. Imaging studies identified cystic lesions in the pancreas. The patient underwent a pancreatectomy, and the pathological examination confirmed the diagnosis of pancreatic serous cystadenoma.

Conclusion: Diagnosing pancreatic masses requires a comprehensive evaluation, with radiological imaging being crucial for accurate diagnosis and treatment planning. For lesions located in the caudal region of the pancreas, distal pancreatectomy is the recommended surgical approach.

Keywords: Pancreatic Serous Cystadenoma, Cyst, Distal Pancreatectomy

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INTRODUCTION

The literature estimates the prevalence of pancreatic cystic tumors to be between 2.6% and 19.6%, and their incidence is rising due to breakthroughs in diagnostic imaging. Clarification of the clinical, pathological, and imaging findings of serous cystic neoplasm of the pancreas is required, as the justification for surgery in individuals with this tumor is occasionally disputed.¹ Even in their diverse morphological appearances (microcystic, oligocystic/macrocystic) and cystadenocarcinomas, which are seldom documented, serous cystic neoplasms are virtually always benign tumors.²

A third of benign primary pancreatic cystic neoplasms consist of pancreatic serous cystadenoma. Patients might typically present without symptoms. In addition to mucinous cystic neoplasms, intraductal papillary mucinous neoplasms, neuroendocrine cystic neoplasms, solid pseudopapillary neoplasms, acinar cell cystic neoplasms, and ductal adenocarcinoma with cystic degeneration are all prevalent kinds of primary pancreatic cystic neoplasms.¹

The exact etiology of serous cystadenoma is not well understood. It is possible that genetic mutations may contribute to the formation of serous cystadenomas. Approximately 40-75% of patients with cystic tumors of the pancreas are asymptomatic, whereas other cases are incidentally detected by imaging analysis.³ Evaluation of serous cystadenomas involves laboratory and imaging modalities to differentiate these types based on morphological and radiological patterns.⁴⁻⁷

The subject of this case study is a female patient who was diagnosed with a pancreatic serous cyst.

CASE ILLUSTRATIONS

A young child of age five was admitted to the emergency room at Arifin Achmad Hospital because she complained of severe pain in the upper left abdomen since 5 days before she was still in the hospital. The patient complained of fever and cough and cold three days before complaining of abdominal pain. The patient's mother gave paracetamol purchased at the pharmacy to treat fever, then abdominal pain appeared suddenly, which made the patient unable to walk and when lying down the patient was unable to tilt left and right. The lower abdominal area feels hard and swollen, reddish in color

compared to other abdominal areas and there is no palpable bulge in the abdominal area. Defecation is rare, sometimes the bowels are dark black and brown.

Patients complain that they cannot eat too much because it makes patients experience nausea and vomiting. The patient was referred with a benign tumor on CT scan examination. Physical examination when the patient arrived at the emergency room found the patient was fully conscious with moderate general condition. The patient's blood pressure was 105/73 mmHg, heart rate = 102 beats per minute, respiratory rate = 22 beats per minute, and the patient had no fever. General examination was found within normal limits. Local examination found tenderness in the epigastrium, that part felt hard and swollen reddish in color.



Figure 1. The mass indicated by ultrasound examination

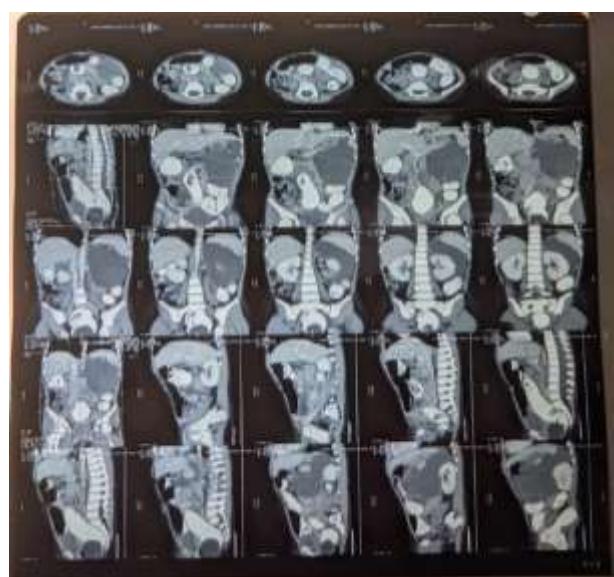


Figure 2. CT scan results

The patient was found to have hypochromic and microcytic anemia, according to the findings of the laboratory examination; nevertheless, the patient's pancreatic lipase and amylase levels were found to be within normal ranges, with values of 26 U/L and 25 U/L respectively. An ultrasound scan revealed that there was a tumor in the patient's pancreas. An ultrasound examination suggested that there was a mass, therefore a CT scan was performed to locate it. The CT scan revealed that there was a cystic mass in the cauda of the pancreas, which results in a pancreatic pseudocyst. It had been decided that the patient would go through with a distal pancreaticectomy.

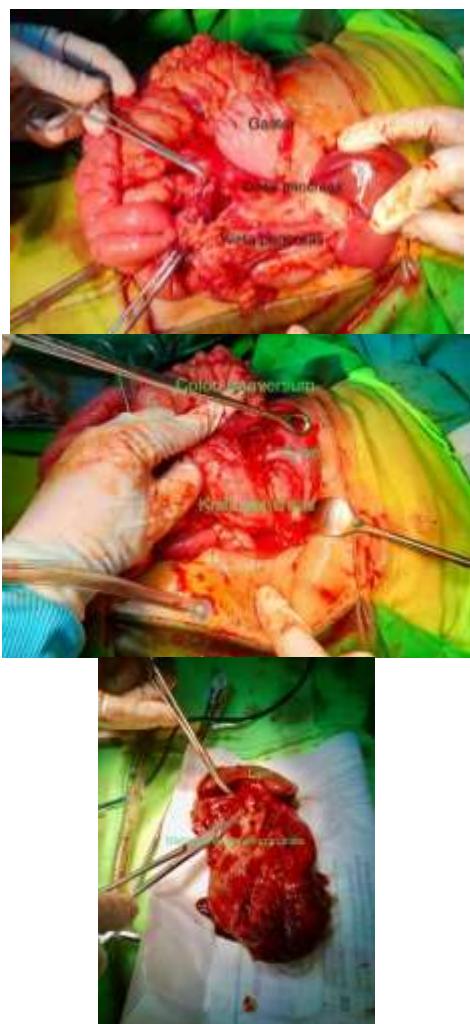


Figure 3. Intraoperative photo

The organ specimens are sent to the anatomic pathology laboratory for examination. The patient was returned to the inpatient room after undergoing surgery. Postoperative

monitoring is carried out by assessing vital signs, fluid balance, and pain experienced by the patient. The patient is temporarily fasted until bowel sounds return. The results of anatomic pathology examination showed a pancreatic serous cystadenoma.

DISCUSSION

Pancreatic serous cystadenoma constitutes the majority of benign primary pancreatic cystic neoplasms, comprising one-third. Patients can usually present asymptomatic. Other common types of primary pancreatic cystic neoplasms include mucinous cystic neoplasms, intraductal papillary mucinous neoplasms, neuroendocrine cystic neoplasms, solid pseudopapillary neoplasms, acinar cell cystic neoplasms, and ductal adenocarcinoma with cystic degeneration.¹ Increasing use and advances in cross-sectional imaging of the abdomen have resulted in increased detection of pancreatic cystic neoplastic neoplasms, especially in asymptomatic patients.^{8,9}

The exact etiology of serous cystadenoma is not well understood. It is possible that genetic mutations may contribute to the formation of serous cystadenomas. Pancreatic serous cystadenoma is almost always benign and occurs more frequently in women. Patients usually presented between the 5th and 7th decades of life (mean age 62 years), and the lesions were more common in the corpus or caudal pancreas. Approximately 77% of patients with von Hippel–Lindau syndrome (VHL) have cystic pancreatic lesions, of which 9% are serous cystadenomas.^{4–7}

The pathophysiology of pancreatic serous cystadenoma is not well understood. Morphological and immunohistochemical features of the serous neoplasm suggest pancreatic centroacinar, intralobular, and ductular cells. The World Health Organization's (WHO) classifies it into serous microcystic adenomas and serous macrocystic (oligocystic) adenomas.⁷ There is a macroscopic variant of the serous cystadenoma; microcystic serous cystadenoma (most common), macrocystic serous cystadenoma (also called oligocystic), solid serous adenoma, VHL-associated serous cystadenoma, and mixed serous–neuroendocrine neoplasm.¹⁰

Macrocystic serous cystadenomas include oligocystic serous and poorly demarcated serous adenomas. Solid serous adenomas are well demarcated and have a rough, dense appearance. They had the cytology and immunohistology of a

classic serous cystadenoma. The serous cystadenoma that occurs in VHL syndrome involves the entire pancreas or unequally. Mixed serous neuroendocrine neoplasm is a rare entity of serous cystadenoma associated with pancreatic neuroendocrine tumors. It is also suggestive of VHL syndrome.^{11,12}

Pancreatic serous neoplasms are unifocal, round, well-defined, and often honeycomb in shape. The cyst contains serous fluid which is free of mucin and is lined by squamous or cuboidal epithelium. Its size can reach more than 20 cm. The cyst is not associated with the pancreatic duct. Typically, a dense fibronodular scar is found in the center of the lesion (also called stellate dibrosa scar) consisting of acellular hyalinized tissue and clusters of small cysts, lined by a single layer of cuboidal epithelial cells. Many patients present without specific signs or symptoms. Pancreatic serous cystadenoma is often detected incidentally by abdominal ultrasonography or cross-sectional imaging studies performed for other conditions.¹²⁻¹⁴

The most common symptom is abdominal pain and/or a palpable mass in the upper abdomen. Patients with advanced cystic lesions rarely present with symptoms similar to pancreatic ductal carcinoma, such as abdominal pain, weight loss, early satiety secondary to compression of the gastric wall by an adjacent pancreatic mass (gastric outlet obstruction), obstructive jaundice due to biliary tree obstruction and pancreatic duct obstruction which can lead to exocrine pancreatic insufficiency and recurrent pancreatitis. Large pseudocysts can compress the stomach, duodenum, or bile ducts, which can cause the patient to experience vomiting, early satiety, or obstructive jaundice.¹²⁻¹⁴

Evaluation of serous cystadenomas involves laboratory and imaging modalities to differentiate these types based on morphological and radiological patterns. In the last 15 years, there has been a 20-fold increase in the detection of pancreatic serous cystadenoma, especially with cross-sectional imaging such as computed tomography (CT) and magnetic resonance imaging (MRI). Emerging imaging modalities led to the detection of four morphological patterns of serous cystadenoma: microcystic, macrocystic, mixed microcystic, and macrocystic and solid. The microcystic pattern was defined as multiple cysts <2 cm in size and separated by thin fibrous septa that give a honeycomb appearance.^{13,14}

Ultrasound usually shows a well-defined loculated lesion. The fibrous portion of the lesion is hyperechoic, and the cystic portion appears hypoechoic. In lesions where the cyst is only a few millimeters in size (microcyst), the tumor appears solid because of the innumerable interfaces. Areas of calcification also appear hyperechoic with posterior acoustic shadowing. Computed tomography (CT) scan shows a lobular shape. They appear hypodense on unenhanced CT because the tumor is densely packed with water. The calcifications that occur in 30% of serous cystadenomas are pathognomonic, and a "sunburst" pattern is seen less frequently.¹³⁻¹⁶

Fluid analysis includes chemical analysis (amylase), tumor markers (carcinoembryonic antigen [CEA], carbohydrate antigen (CA19-9), and cytology. Serous cystadenoma shows low viscosity, low amylase level, and low CEA level (<5 to 20 ng/ mL) with variable levels of pancreatic enzymes. Cancer antigen levels (CA 72-4) are also low. Cytology reveals cuboidal cells with glycogen-rich cytoplasm. This combination of parameters helps in classifying these cysts and distinguishing serous cystadenomas from malignant or mucous cysts. DNA analysis Cystic fluid may reveal mutations in VHL among patients with VHL syndrome.^{13,14}

The patient in this case underwent surgery. Non-surgical management should be recommended in asymptomatic cases, although patients should undergo regular follow-up using radiological examinations. This is due to the small risk of malignancy (<3%) in these cases.¹⁷ Surgical resection should be the next step in the management of symptomatic patients because of rapidly growing lesions (cystadenoma itself vs. hemorrhage), giant tumors >10 cm (causing mass effect, obstructive jaundice, exocrine pancreatic insufficiency or gastric outlet obstruction) or when malignancy (eg. serous cystadenocarcinoma) cannot be excluded.¹³

The surgical approach depends on the location and may include a distal pancreatectomy with or without splenectomy, mid pancreatectomy, or the Whipple procedure. Enucleation was previously reported as a safe option for well-selected patients with serous cystadenoma, with preservation of endocrine and exocrine pancreatic function.¹⁸⁻²⁰ Lesions on the caudal as in this case are usually treated with distal pancreatectomy with or without splenectomy, whereas lesions on the neck or

corpus are treated with middle segment pancreatectomy.²¹

Resection of a serous cystadenoma is possible in most cases, although complaints and sheathing of the mesenteric portal vein can increase the difficulty and danger of surgery. Recurrence of fully resected serous cystadenomas is virtually never seen, and even incompletely excised serous cystadenomas (such as when a small margin of the tumor remains in or behind the mesenteric vein) also do not cause recurrent cysts.²¹

CONCLUSION

The diagnosis of masses that occur in the pancreas must involve multimodal examination, where radiological examination is the most decisive in making decisions. If the lesion is located caudal, a distal pancreatectomy is performed.

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