

## Metastatic pancreatic ductal adenocarcinoma in a teenage girl: A rare disease

Sutaryo, Scolastika Dita Kristian

**P**ancreatic ductal adenocarcinoma (PDAC) is highly uncommon in patients < 20 years of age, at less than 0.1% of population.<sup>1</sup> Pancreatic tumors in children and adolescents can develop from endocrine or exocrine cells. The tumor types include solid pseudopapillary tumor, ductal adenocarcinoma, pancreatoblastoma, acinar cell carcinoma, and pancreatic endocrine neoplasm (malignant and benign).<sup>2</sup> Other types of tumors may be attached to it or secondarily engage the gland or emerge from other kinds of non-pancreatic cells inside the pancreas. The prevalent type of classic PDAC in adults is highly uncommon in children. We report here on a fifteen-year old girl with metastatic pancreatic ductal adenocarcinoma (mPDAC), who presented with abdominal discomfort and jaundice. [Paediatr Indones. 2020;60:341-3; DOI: 10.14238/pi60.6.2020.341-3 ].

**Keywords:** *pancreatic ductal adenocarcinoma; pediatric*

### The Case

A 15-year-old girl came to Dr. Sardjito General Hospital, Yogyakarta, with jaundice and abdominal discomfort for the past three months. We found hepatosplenomegaly on physical examination. Total bilirubin level was 16.79 mg/dL and conjugated bilirubin was 15.54 mg/dL. Aspartate aminotransferase and alanine aminotransferase levels were 448 IU/L and 453 IU/L, respectively. Abdominal ultrasound revealed hydrops vesica fellea and dilatation of the intrahepatic and extrahepatic bile ducts. Hypodense lesions of the head of the pancreas measuring 49x42x72 mm and dilatation of intra- and extra-hepatic bile ducts were observed by magnetic resonance imaging and computerized tomography

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From the Department of Child Health, Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada/Dr. Sardjito General Hospital, Yogyakarta, Central Java, Indonesia.

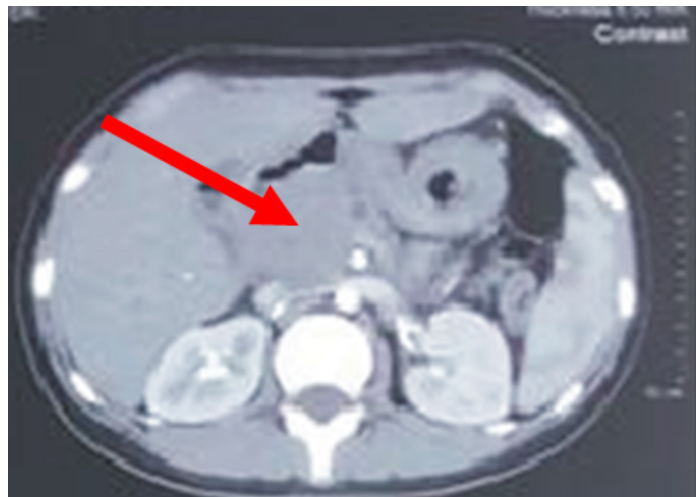
Corresponding author: Sutaryo. Department of Child Health, Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada/Dr. Sardjito General Hospital, Yogyakarta, Indonesia. Email: sutaryo@ugm.ac.id.

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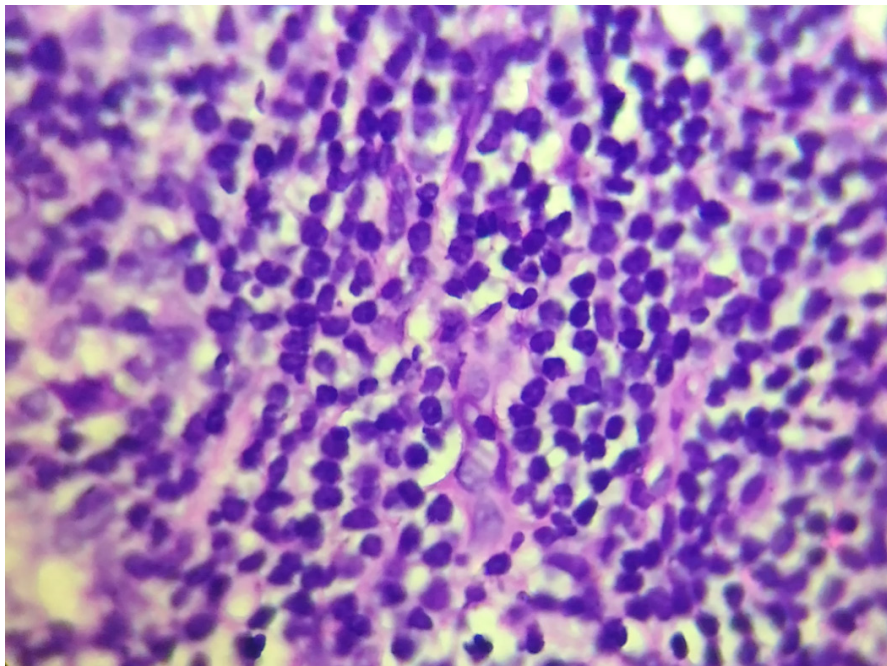
(CT) (Figure 1). The metastatic pancreatic ductal carcinoma (mPDAC) diagnosis was confirmed by histopathological examination from the pancreatic lesion and lymph node biopsy (Figure 2). The patient died during the third cycle of gemcitabine because of pneumocystis.

## Discussion

Our understanding of this tumor is restricted to this case. Ductal adenocarcinoma is rare in the first four decades of life and even rarer during childhood and adolescence.<sup>1</sup> Most PDAC reports are in older publications. In 31 years of registry, the *Surveillance, Epidemiology and End Results Registry* (SEER) had 58 patients with malignant pancreatic neoplasms. The



**Figure 1.** CT scan of ductal pancreatic mass (red arrow)



**Figure 2.** Histopathology of the lymph node (100x)

ratio of women to men was 1.9:1, with age population-adjusted incidences of 0.021 and 0.015 per 100,000, respectively.<sup>3</sup> Seven children were identified with PDAC. In a 90-year (1918-2007) retrospective review of a single institution, 18 patients with seven different histopathological subtypes were recognized. The most common subtypes were solid pseudopapillary, neuroendocrine gastroenteropancreatic, and acinar tumors.<sup>4</sup> Pancreoblastoma is the most common childhood pancreatic tumor. It is associated with several cancer predisposition syndromes, such as Peutz-Jeghers syndrome, familial atypical mole and multiple melanoma (CDKN2 mutations), hereditary pancreatitis (PRSS1 mutations), and other hereditary nonpolyposis colon carcinomas (STK11 and germline mismatch repair genes), and syndromes associated with DNA repair gene mutations (such as ATM and BRCA2).<sup>5</sup>

Ductal adenocarcinoma generally manifests with abdominal pain or palpable abdominal mass. Jaundice happens less frequently in children than in adults.<sup>2</sup> Surgery remains, as in adults, the key to treating non-metastatic pancreatic tumors in the pediatric population.<sup>6</sup> Long term disease-free survival is achieved with full surgical resection in childhood pancreatic malignancies. Chemotherapy or radiation has no documented significance,<sup>4</sup> but their importance is disputed. In unresectable and metastatic disease, these modalities should be reserved. Pancreatic ductal adenocarcinoma has bad prognosis, with only 23% surviving for 15 years.<sup>7</sup> It is one of the most aggressive cancers with high metastatic potential. Clinical observations suggest that there is disease heterogeneity among patients with different sites of distant metastases, yielding distinct clinical outcomes.<sup>8</sup> In pediatric populations, malignant pancreatic tumors, particularly ductal carcinomas, are uncommon. Appropriate diagnosis and therapy remain a challenge for health care providers. Besides that, this rare cancer is extremely challenging to study because of the low incidence of patients with any individual diagnosis, the predominance of rare cancer in the adolescent population, and the lack of clinical trials for adolescents with rare cancer.

## Conflict of Interest

None declared.

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