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Acute Pancreatitis as an Initial Manifestation of Pediatric AML: A Case Report with Review of Literature

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Abstract

Acute myeloid leukemia (AML) accounts for around 15 to 20% of childhood leukemia. Acute leukemia has variable presenting features and extramedullary manifestations typically occur in 10 to 20% of patients with AML, even before bone marrow involvement. Therefore, leukemia may get unnoticed if symptoms from other organs predominate. Acute pancreatitis, as an initial manifestation of acute leukemia, is seen commonly during and after the course of chemotherapy.

Here, we report the case of a 16-year-old female patient who presented with pain abdomen, multiple swellings in the neck, axilla, and breast for 2 months and had jaundice at presentation was diagnosed to be having acute pancreatitis with AML. Pancreatitis was resolved after starting cytoreductive chemotherapy. Hence, the causation of pancreatitis in the index case was likely due to pancreatic infiltration by the leukemic cells, unlike the common etiologies such as hypercalcemia and chemotherapy drugs seen in the setting of leukemia.

Keywords

- acute myeloid leukemia
- acute pancreatitis
- case report
- extramedullary

Introduction

The most frequently occurring hematological malignancies in children include acute leukemia, of which 80% are classified as acute lymphoblastic leukemia and 15 to 20% as acute myeloid leukemia (AML).[1] Pediatric AML has an incidence of 7 cases per million children younger than 15 years.[2] The incidence of AML in infants is 1.5 per 100,000 individuals per year, it decreases to 0.9 per 100,000 individuals aged 1 to 4 and 0.4 per 100,000 individuals aged 5 to 9 years, after which it gradually increases into adulthood, up to an incidence of 16.2 per 100,000 individuals aged over 65 years.[3]

The clinical features of AML occur due to bone marrow and organ infiltration by leukemic cells.[4] Acute pancreatitis at presentation due to AML is very rare. So far, there is no pediatric case reported in the literature. We diagnosed a case of AML in a 16-year-old female patient with pancreatitis as an initial manifestation.

Case Report and Literature Review

A 16-year-old female patient was brought to the emergency department with complaints of pain in abdomen in the left hypochondrium and epigastrium radiating to back. It developed gradually over the past 2 months, was mild to moderate grade, aggravated on food intake, and decreased on bending forward. She had also developed multiple swellings in the neck, axilla, and breast in the past 2 months. The swellings were progressively increasing in size, noticed as a hard and painless lump in both the breasts. A few days before admission, the patient complained of high-colored urine and parents noticed yellowish discoloration of eyes and body.

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The patient had attained menarche at 12 years of age and had regular menstrual cycles.

On examination, she was anxious and dyspneic (respiratory rate of 32 per minute and oxygen saturation of 88% in room air) with accessory muscle use. Chest examination demonstrated decreased air entry in bilateral infra-axillary and infra-scapular regions. Her oxygen saturation improved with oxygen support.

On general physical examination, pallor and icterus were present. She had lymphadenopathy involving bilateral cervical lymph nodes which were nontender, firm to hard in consistency, and matted. Also, bilateral multiple axillary lymph nodes were enlarged up to 3×3 cm in size, nontender, matted, and hard. These axillary swellings were in proximity to bilateral breast tissue lumps which were nontender, hard, and fixed to the anterior chest wall with no discrete borders or discharge from nipple or skin changes.

On abdominal examination, the patient had distension with hepatosplenomegaly with a liver span of approximately 15 cm in size, nontender with smooth surface, firm consistency, and spleen 4 cm below costal margin, firm, and nontender.

Blood investigations were sent that revealed anemia (hemoglobin 62 g/L), hyperleukocytosis (total count: 128×10^9/L), and thrombocytopenia (platelet count: 45×10^9/L). Serum lactate dehydrogenase levels were elevated (929 IU/L) with hyperbilirubinemia (total bilirubin: 176 micromol/L and direct bilirubin: 103 micromol/L) and mild transaminitis (serum glutamic-oxaloacetic transaminase: 121 U/L and serum glutamic-pyruvic transaminase: 107 U/L). The serum lipase and amylase levels were also elevated to 251 and 104 U/ L, respectively. Serum calcium (9.2 mg/dL) and phosphorus (3.7 mg/dL) were in normal range. C-reactive protein was negative (0.9 mg/dL) and cultures of blood and urine were sterile. Lipid profile was within normal limits (total cholesterol: 143 mg/dL, low-density lipoprotein: 76 mg/dL, triglycerides: 126 mg/dL). Peripheral smear showed presence of 46% blasts. Bone marrow aspirate smear was suggestive of myeloperoxidase-positive acute leukemia. Flow cytometry confirmed the diagnosis of AML M2 with 41.66% blasts with bright expression of CD 13 and moderate to bright expression of HLA DR and CD 117, moderate expression of MPO and CD 34, and dim to moderate expression of CD7, CD 19, and CD 33. Polymerase chain reaction panel for common AML mutations was done and it was negative for CEBPA, NPM1, PML RARA, FLT3–ITD/D835, CBFB/ MYH11, and AML1/ETO.

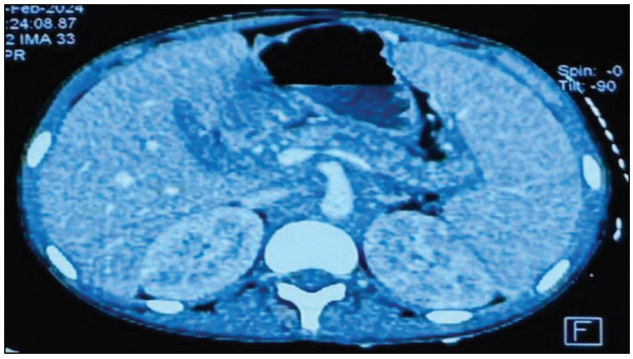

Fig. 1 Contrast-enhanced CT abdomen showing pancreas heterogenous in morphology with prominent main pancreatic duct suggestive of acute pancreatitis. CT, computed tomography.

USG (ultrasound) breast tissue (bilateral) showed heterogeneous echotexture with multiple hypoechoic nodules scattered throughout the breast tissue s/o (suspected of) granulomatous lobular mastitis with lymphadenopathy.

USG abdomen showed a bulky heterogeneous pancreas with parts of the tail and body appearing bulky approximately 23 mm in size suggestive of acute pancreatitis. The contrast-enhanced computed tomography scan of abdomen showed heterogeneous pancreas with prominent main pancreatic duct suggestive of pancreatitis as in ►**Fig. 1**.

The patient was started on cytoreductive chemotherapy with low-dose cytarabine at 10 mg/m^2/dose 12 hourly for 7 days after which she started improving. It was followed by clinical and laboratory resolution of pancreatitis over the next 4 to 5 days as pain abdomen was improved and initially serum lipase and amylase levels were elevated to 251 and 104 U/L and decreased to 150 and 61 U/L, respectively, after 7 days.

Discussion

Acute pancreatitis is generally a reversible process without any lasting effects on the pancreatic parenchyma or function. The main causes of pancreatitis are infections (viral, bacterial, mycotic, and parasitic), trauma, drugs, systemic diseases, and metabolic conditions like hyperlipidemia (hypertriglyceridemia) and hypercalcemia.[5] Existing literature suggests the occurrence of pancreatitis in a patient of leukemia is due to chemotherapeutic drugs, especially L-asparaginase.[6] Pancreatitis has also been reported in AML patients on treatment with cytarabine and all-*trans* retinoic acid.[7] The pancreas can be directly infiltrated by acute leukemia, especially myeloblasts, which is a rare cause of acute pancreatitis.

AML with acute pancreatitis as an initial presentation has been reported in few adult patients in the literature, but none in pediatrics. The summary of cases reported so far is presented in ►**Table 1**.

Here we have reported pancreatitis due to AML in a 16-year-old girl. The strength of this case report is that metabolic conditions like hyperlipidemia and hypercalcemia were ruled out. There was no history of chemotherapy or drugs causing pancreatitis prior to presentation. We were unable to perform a biopsy because of the high possibility of pancreatic hemorrhage after the procedure. This is the only limitation to this case. Moreover, after cytoreductive chemotherapy, pancreatitis improved.

Leukemia can present with a variety of symptoms due to extramedullary infiltration, which can impact any organ. Research has demonstrated that AML cells can hasten the leukemia progression by creating a favorable malignant microenvironment that results in extramedullary infiltration.[8] AML patients with extramedullary infiltration have been found to have a significantly higher bone marrow

Table 1 Summary of previous similar case reports

Serial No.	Reference No.	Age/sex	Chief complaint	Bone marrow findings	Flow cytometry	Sub-type of AML	CT scan abdomen
1.	[9]	61 years Male	Pain abdomen	BM morphology: increased myeloblasts; IHC staining revealed an increase in myeloblasts staining strongly for myeloperoxidase	Myeloblasts (68.93%) were positive for CD34 and CD117 and negative for CD15, CD36, and CD14	FAB AML M2	Diffuse edema of the pancreas and peripancreatic effusion splenomegaly with gallbladder stones
2.	[10]	33 years Male	Fever, back pain	Increased monoblasts (38.6%) and eosinophils (8.1%), with 270,000 copies/μg RNA of the CBFβ/MYH11 fusion gene	Monoblastic cells were positive for CD 13, 33, 34 and HLA- DR and negative for CD 2, 3, 4		Dilatation of the main pancreatic duct, swelling of the pancreatic head, multiple cuneiform defects of the bilateral kidney and splenomegaly
3.	[11]	35 years Male	Pain abdomen, vomiting	Markedly hypercellular bone marrow and expansion of the peroxidase-positive myeloblasts (90.8% of all nucleated cells)	Myeloblasts express CD13, CD33, CD34, CD117, MPO, CD11c, and CD56	AML (FAB M1)	Edematous pancreas due to the inhomogeneous tumor in its head
4.	[12]	36 years Female	Epigastric pain	Presence of 59% of undifferentiated blasts of medium size, a rounded or oval nucleus, with the presence of nucleoli fine, chromatin, and more or less basophilic cytoplasm strewn with azurophilic granulations and myeloblasts positive for myeloperoxidase activity	NA	NA	Suggestive of acute pancreatitis

Abbreviations: AML, acute myeloid leukemia; CT, computed tomography; S/O, suspected of.

plasma matrix metalloproteinase 9 levels.[9] The most plausible explanation of pancreatitis seems to be leukemic infiltration as both pancreatitis and leukemia responded to cytoreductive chemotherapy.

Conclusion

This is the first case of pediatric AML presenting with pancreatitis at initial presentation. Based on the above details and existing literature in adults, it is proposed that leukemic infiltration caused acute pancreatitis in this patient. Given that acute pancreatitis is an uncommon but possible manifestation in pediatric AML, it necessitates urgent attention as delayed diagnosis and treatment can result in systemic inflammatory response syndrome and organ dysfunction if not timely diagnosed and treated. Hence a high index of suspicion is essential as leukemia may be a potential cause of pancreatitis in pediatric patients.

Authors' Contributions

The manuscript has been read and approved by all the authors and requirement for authorship of this document has been met. Each author believes that the manuscript represents honest work. T.S. and S.K.A. conceptualized the study. T.S., S.K.A., and A.H. helped in collection of data, searched literature, drafted the manuscript, and gave final approval for the same.

Patient Consent

Written informed consent has been taken from the parent/guardian.

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Conflict of interest

None declared.

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