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Research Article

Rare Cause of Acute Abdomen in Children: Spontaneous Rupture or Bleeding of Solid Malignant Tumor

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Keywords

- Solid pseudopapillary tumor of the pancreas
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- Gastrointestinal stromal tumors
- Childhood
- Peritonitis
- Acute abdomen

Abstract

Introduction: Pediatric patients with abdominal solid malignant tumor may occasionally present with acute abdomen and required emergency surgery because of peritonitis. This report presents four cases of abdominal solid malignant tumor which are associated with rare emergency presentations.

Methods: This study was carried out on four patients with rare pediatric abdominal malignant tumor who presented to the Pediatric Surgery Unit in an emergency situation. All patients' data, clinical presentations, radiological data, surgical procedures, complications, and survival data were collected, reviewed and analyzed.

Results: Between December 2010 till September 2014, 4 patients were admitted with different emergency presentations of clinically and radiologically. The tumor was identified separately was solid pseudopapillary tumor (2 cases), gastrointestinal stromal tumors located in the transverse mesocolon and undifferentiated embryonal sarcoma of the liver. The common presenting symptom was acute abdomen with peritonitis suspected of tumor spontaneous rupture and bleeding.

Conclusions: The emergency surgeon must be acquainted with the malignant tumor, its emergency presentation and principles of surgery in the presence of malignant tumor in children.

INTRODUCTION

Acute abdomen can be defined as a medical emergency, in which there is sudden and severe pain in abdomen of recent onset with accompanying signs and symptoms that focus on an abdominal involvement. It is a common presenting complaint in pediatric emergency. The pain is often due to a wide range of mild self-limiting medical causes, but it sometimes may be due to an acute potentially life threatening, surgical/medical illness. The clinical symptoms and signs in an acute abdomen may be not typical in many cases [1].

Most cases of abdominal solid malignant tumor in children are asymptomatic. However, patients may occasionally present with acute abdomen because of peritonitis caused by spontaneous rupture, bleeding or infection. Acute peritonitis in children is often associated with high morbidity and mortality [2-6]. Severe intra-abdominal sepsis resulting from intestinal perforation, mechanical intestinal obstruction and neglected abdominal trauma are the common causes of generalized surgical acute peritonitis in developing countries [7,8].

Here, we present four patients who underwent surgery for an acute abdomen because of severe peritonitis and were observed to have abdominal solid malignant tumor including of 1 case of gastrointestinal stromal tumors (GISTs) derived from the transverse mesocolon, 2 cases of solid pseudopapillary tumor of the pancreas (SSPT) and 1 case of undifferentiated embryonal sarcoma of the liver (UESL).

PATIENTS AND METHODS

Between December 2010 and September 2014, four patients (females) were admitted with different acute abdomen and emergency presentations of clinically and radiologically suspected abodomimal malignant tumor.

After approval of local ethics committee, all patients included in the study were informed well about the procedure and an informed written consent was obtained from every patient before carrying the procedure.

All patients' data, clinical presentations, laboratory and radiological investigations, tumor characteristics, pathological findings, surgical procedures, as well as follow-up and survival data were collected, reviewed and analyzed.

Four patients were operated upon as soon as possible. All operated patients underwent surgical resection. Histopathological examinations of surgical specimens and immune histochemical staining were performed in all cases.

RESULTS

This study included four patients who presented with emergency complications of three kinds of rare abdominal malignant tumor in children on clinical and radiological aspects. They were all females. Their mean age at diagnosis was 13 years. They all presented with acute abdomen and local peritonitis suspected of tumor spontaneous rupture and bleeding. The tumor was identified separately SSPT located in the tail of the pancreas (case1), in the body and tail of the pancreas (case2), GIST located in the transverse mesocolon (case3), and UESL (case 4). The overall clinical features of all cases were shown in (Table 1).

Enhancement CT scan of the abdomen and pelvis was performed in all the cases. The CT scan of case1 demonstrated a 9.4x7.1cm irregular mixed-density space-occupying lesion in the tail of the pancreas (Figure 1A). The CT scan of case3 demonstrated a 7.3x5.8cm cystic mass in the right quadrant containing air and fluid surrounded by dilated and thickened bowel (Figure 1B). The CT scan of case4 demonstrated a mass with a 20.2x13.6x21.7cm no-defined border in the right lobe and medial segment of left lobe of the liver (Figure 1C).Complete macroscopic resection was achieved in all patients. Pathology analysis revealed clear margins in three specimens (except case4).

The detailed immunohistochemical results are listed in (Table 2).

Cytokeratin profiles (CK) characterize ducts and ductular cells of the pancreas. Chromogranin A (CgA) and synaptophysin (Syn) are considered neuroendocrine markers. Immunohistochemical analysis was performed on samples from both patients. Both were positive for vimentin, β-catenin, and progesterone receptors (PR) whereas CK, CgA and Syn were negative. CD10 in one case was positive and the other was negative (Figure 2a-d).

GIST diagnosis was based on histology and verified by immunohistochemistry (IHC) using KIT (CD117), CD34, smooth muscle actin (SMA), and S100 protein. The immunohistochemistry results of case3 were as follows CD117, Dog-1 and SMA was positive, whereas CD34, Desmin and S-100 were negative. In addition, Ki67 were positive (50%) (Figure 3a-d).

The immunohistochemistry results of UESL were as follows: Broad spectrum creatine kinase (CK) and CD34 were negative, SMA, a-ACT and vimentin (a component of the cytoskeleton of mesenchymal cells) were positive (Figure 3a-c). Hospital stay ranged from 7 to 36 days.

The UESL patient's parents refused adjuvant chemotherapy and demonstrated a right liver mass 3 years post 1st surgery (Figure 1D). Microscopic examination revealed that the tumor was evidence of UESL recurrence (Figure 3d). However, the patient was comfortable and physical examination revealed no abnormal conditions. In addition, the laboratory results were normal. Abdominal computed tomography scan and ultrasound were performed every 3 months to monitor the tumor recurrence. At the time of writing, it has been 6 months after the second surgical procedure and there has been no appearance of abnormalities. The GIST patient's parents received Gleevec therapy and did not develop metastases or recurrence. Two cases of SSPT did not develop metastases or recurrence too.

Table 1: Overall clinical features of all cases.

Patient	1	2	3	4
gender	female	female	female	female
age at presentation	13	13	13	13
chief complaint	vomitting abdominal pain	vomitting abdominal pain	fever abdominal pain	vomiting abdominapain fever
physical examination	local peritonitis	local peritonitis	local peritonitis	local peritonitis
tumor location	tail of pancreas	body and tail of pancreas	transverse mesocolon	liver
tumor size (cm)	9.5*7.5*7.5	8*8*7	7*6*6	20*14 *22
tumor presentation in the operation	bleeding	bleeding and rupture	bleeding rupture and infection	bleeding and rupture
metastasis	No	No	No	No
procedure	tumor resection	tumor resection	tumor resection	tumor resection
length of stay	8	7	36	8
chemotherapy postoperatively	No	No	Yes	No
follow-up	18m	2m	11m	4y
histology	SSPT	SSPT	GIST	UESL
recurrence	No	No	No	YES

Abbreviations: SSPT: Solid Pseudopapillary Tumor; GIST: Gastro Intestinal Stromal Tumors; UESL: Undifferentiated Embryonal Sarcoma of the Liver

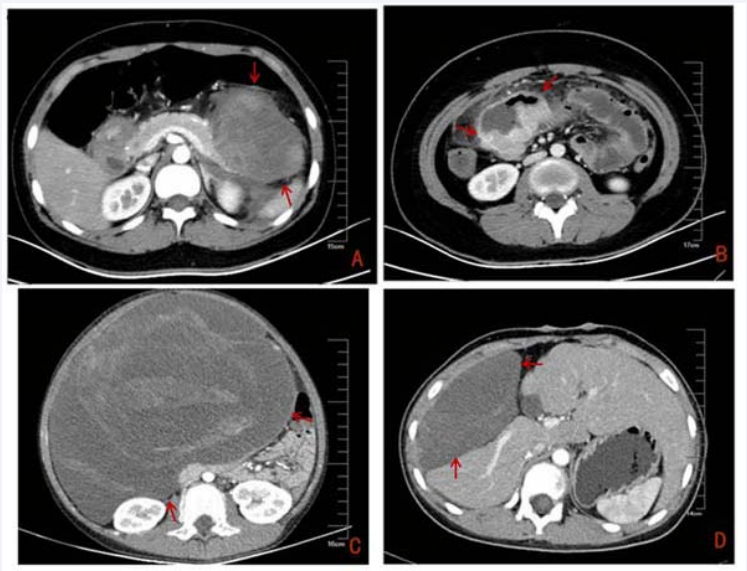

Figure 1 Contrast-enhanced CT imaging of tumor.

(A) Axial computed tomography image showed a 9.4x7.1cm irregular mixed-density space-occupying lesion in the tail of the pancreas (brown arrow), following the administration of a contrast agent, CT scan reveals that the mass is irregularly enhanced.

(B) Axial view of contrast-enhanced CT scan revealed a 7.3x5.8cm cystic mass in the right quadrant containing air and fluid surrounded by dilated and thickened bowel. Following the administration of a contrast agent, CT scan reveals that the mass is obviously enhanced (brown arrow). There was some amount of free air.

(C) Pre-contrast CT scans showing a mass with a 20.2x13.6x21.7cm no-defined border in the right lobe and medial segment of left lobe of the liver. Following the administration of a contrast agent, CT scan reveals that the soft tissue inside the mass is obviously enhanced (brown arrow).

(D) Axial view of contrast-enhanced CT scan revealed a 7.3x5.8cm cystic mass in the right lobe of the liver 3 years post 1st surgery.

Table 2: Positive laboratory examinations and immunohistochemistry results.

Patient	1	2	3	4
Positive laboratory examination preoperatively				
CRP(mg/L)	150	46.4	252	73.3
Blood glucose(mg/L)			9.67	
Immunohistochemistry				
CD10	-	+		
PR	+	+		
β-catenin	+	+		
CD117			+	
chromogranin A	-	-		
vimentin	+	+		+
Syn	-	-		
SMA			+	+
AACT	+	+		+
Desmin			-	
Dog-1			+	
CD34			-	-
CK		-		-
S-100			-	

Abbreviations: CK: Cytokeratin; PR: Progesterone Receptor; Syn: Synaptophysin CgA, Chromogranin A; SMA: Smooth Muscle Actin
"+" indicates positive expression "-" indicates negative expression

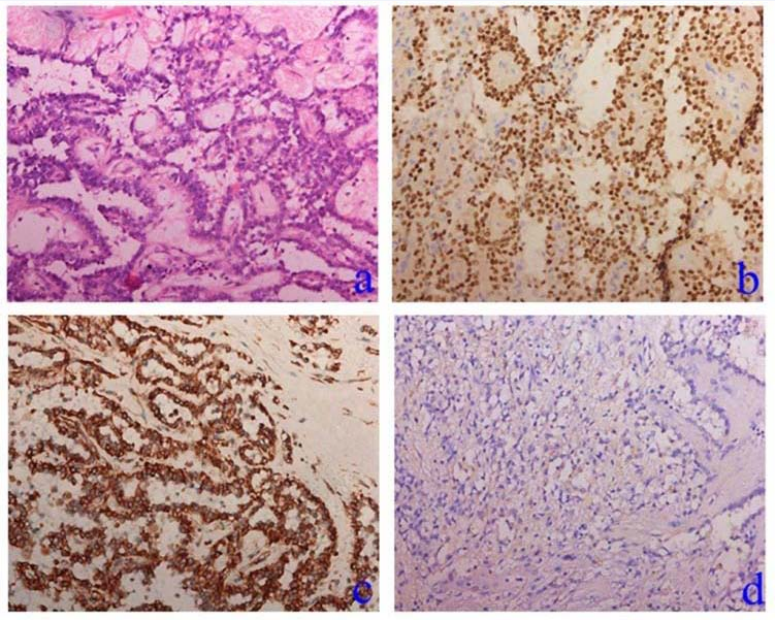

Figure 2 Histopathological and immunohistochemical results of SSPT

a. High power view, showing that the tumor cells have uniform round nuclei with eosinophilic cytoplasm and central microvascular core forming a pseudopapillary pattern (H&E, ×200).

b. PR positive immunohistochemical staining of SSPT (×200).

c. Vimentin positive immunohistochemical staining of SSPT (×200).

d. CgA negative immunohistochemical staining of SSPT (×200).

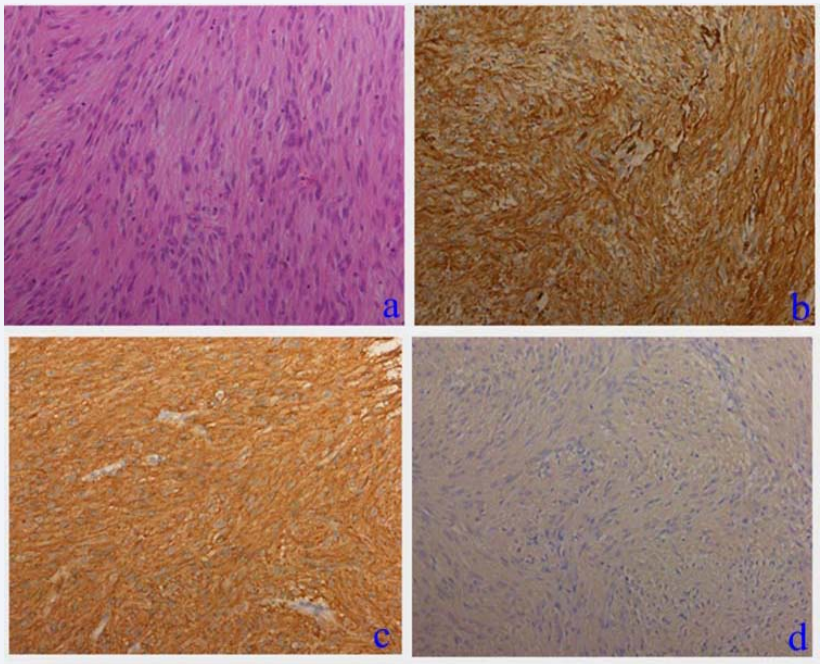

Figure 3 Histopathological and immunohistochemical results of GIST

a. Light microscopy. High power view, showing that continuous tumor nodules separated by interlacing bundles of smooth muscle fibers (H&E, ×200).

b. SMA positive immunohistochemical staining of GIST (×200).

c. CD117 positive immunohistochemical staining of GIST (×200).

d. S-100 negative immunohistochemical staining of GIST (×200).

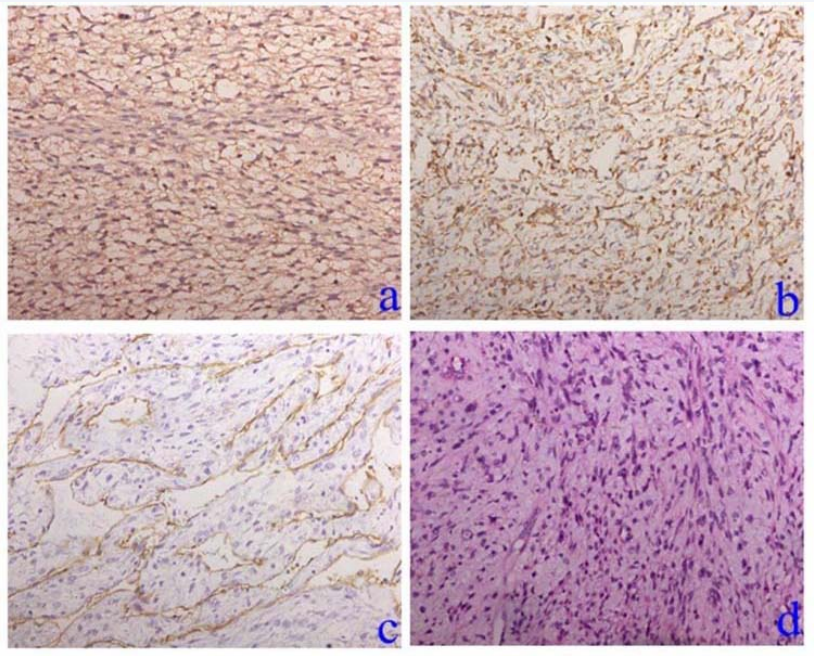

Figure 4 Histopathological and immunohistochemical results of UESL

a. AACT positive immunohistochemical staining of UESL (×200).

b. Vimentin positive immunohistochemical staining of UESL (×200).

c. CD34 negative immunohistochemical staining of UESL (×200).

d. Light microscopy. High power view, showing that the tumor cells exhibited embryonic mesenchymal differentiation and lacked epithelial characteristics (H&E, ×200).

DISCUSSION

This report included four young girls who presented with emergency complications of three kinds of rare abdominal malignant tumor in children on clinical and radiological aspects. The tumor was identified separately was SSPT, GIST located in the transverse mesocolon and UESL. SSPT of the pancreas is a rare neoplasm in children that mainly occurs in young females. SSPT is a rare epithelial solid tumor of the pancreas that invariably develops significant cystic degeneration, acquiring the characteristic solid–cystic appearance. Most SSPT tumors are located in the body/tail of the pancreas, but up to 40% occur in the head [9]. We herein reported two rare cases of SSPT arising from the body and tail of the pancreas.

One 13-year-old girl was admitted to our clinic with abdominal pain and vomitting. No mass was palpated on the physical examination. A 94x71mm, encapsulated, heterogeneous mass with solid and cystic components was defined on computerized tomography (CT). Tumor was rejected during the operation. Histopathological examination revealed that the tumor was a SSPT with positive surgical margins. An eighteen-month follow-up after surgical resection showed no evidence of recurrent disease. SSPT should always be considered in the differential diagnosis in a young female with a palpable mass.

Another 13-year-old girl was admitted to our clinic with continuous abdominal pain. No mass was palpated on the physical examination. An 81x72mm, encapsulated, heterogeneous mass with solid and cystic components was defined on enhanced CT. Tumor was also rejected during the operation. Histopathological examination revealed that the tumor was a SSPT with positive surgical margins. A two-month follow-up after surgical resection showed no evidence of recurrent disease. Certain histochemical markers such as β-catenin, vimentin and progesterone receptors are frequently positive, but non-specific. These distinctive vimentin, PR, CD10, β-catenin and AACT staining pattern were positive whereas CgA and Syn and CK were negative was present in both our cases, throughout the solid and the pseudopapillary regions of all samples.

Reith *et al.,* in 2000 reported another rare GIST that originates outside the gastrointestinal tract; this was termed extraintestinal GIST (EGIST) [10]. In EGIST, just sporadic reports of GIST arising from the omentum, mesentery and retroperitoneum [11]. The most common symptoms associated with mesentery GISTs are vague, non-specific abdominal pain or discomfort. Patients who have mesentery GIST usually suffer from abdominal pain or palpable mass, and also complain of early satiety or abdominal fullness. It is difficult to diagnose preoperatively due to the nonspecific and variable clinical symptoms, and it is also difficult to distinguish the tumor based solely on images. CT scan is a commonly offered imaging modality for patients with suspected abdominal GISTs [12]. Though each disease entity has characteristic features, however, EGIST cannot be completely differentiated from other soft tissue tumors due to overlapping CT findings. In our case, we can't definitely diagnose the tumor but just acute abdomen and peritonitis. The definitive diagnosis of the majority of GISTs

is revealed by histopathological examination of the specimen. Approximately 95% of GISTs express CD117, which is part of the KIT receptor tyrosine kinase. Additionally, DOG1, a recently defined monoclonal antibody against a chloride channel protein expressed by GIST, is positively expressed in 95% of GISTs [13]. DOG1 is a novel marker of GISTs as it has a higher sensitivity and specificity compared with CD34, particularly in the detection of moderate and high risk GIST. Therefore, the present case was diagnosed by immunohistochemical examination of C KIT and DOG1 positivity.

To date, surgery is the only potentially curative therapy for patients with primary, resectable GIST. A lymphadenectomy is not conducted because lymph node metastases are rare [14]. Although the size of tumor in this case was large, there were no harvested positive lymph nodes in the present case. The management of GIST has undergone significant revolution over last decade. Understanding that the molecular pathogenesis of GIST is linked to deregulated KIT tyrosine-kinase activity has resulted in the successful application of a novel systemic tyrosine-kinase inhibitor, imatinib (Gleevec; Novartis Pharmaceuticals Corporation, East Hanover, NJ), in the treatment of GIST patients with malignant metastatic or unresectable disease. Adjuvant imatinib for 3 years should be considered in patients undergoing resection for primary disease [15].

In conclusion, we report the case of a young girl with a perforated EGIST in the transverse mesocolon causing acute local peritonitis. The clinical outcome is worse when this tumor presents with bowel perforation and peritonitis; therefore, if an abdominal mass presents with peritonitis, the possibility of transverse mesocolon GIST perforation should be considered, even though it is extremely rare. A high degree of suspicion is necessary in view of the high morbidity rates resulting from a delayed diagnosis of the disease. High and intermediate-risk patients may need routine follow-up with CT scanning or MRI every 3~6 months during adjuvant imatinib therapy and then after cessation of adjuvant therapy, every 3 months for 2 years, and then every 6 months until 5 years after stopping adjuvant therapy and then annually [16-17].The histology of our case indicated the high-risk tumor and she accepted the adjuvant imatinib therapy and routine follow-up.

UESL is a rare hepatic mesenchymal tumor that was first reported and classified by Stocker *et al.,* in 1978 [18]. In addition, 90% of patients are children aged 6 10 years, and the disease accounts for 5-8% of hepatic tumors in children. The tumor is mainly localized or found in the hepatic right lobe (59%), while it rarely develops in the hepatic left lobe (22%) or the bilateral lobe (20%). Hemorrhaging, necrosis and cystic degeneration are frequently observed, while clinical manifestations include abdominal mass, pain, fever and rarely jaundice [19]. Generally, a definite diagnosis of UESL is not able to be determined preoperatively; a case history combined with imaging analysis has been demonstrated to be beneficial for the discrimination of UESL. The diagnosis relies on postoperative pathological analysis and immunohistochemical results. An enhanced CT scan of the tumor solid part, septation and pseudo-capsule shows different degrees of reinforcement. The immunohistochemistry results reveal the positive expression of SMA, a-ACT, desmin, vimentin and actin in UESL patients, and minorities of cases are positive for PCNA, CK8/18 and p53; while AFP, S-100, CEA, CA 19-9 and cytokeratin have negative expression. The indices of the patient were as follows: vimentin, SMA and AACT were positive whereas CD34 and CK were negative.

Sakellaridis *et al.,* [20] described a 20-year-old patient who presented with spontaneous abdominal haemorrhage due to a liver mass; this was managed with direct suture of identifiable vessels, but the patient died of uncontrolled bleeding and disseminated intravascular coagulation after lobectomy. Fatal tumor rupture after partial tumor excision undertaken for the purpose of tissue diagnosis has been reported in an 8-year old girl with UESL of the liver. The patient died of hypovolaemic shock with haemothorax and haemoperitoneum [21]. In the present study, we describe a case of UESL in a 13-year-old female, whose initial symptoms included the upper severe abdomen pain and a palpable enormous irregular tumor accompanying with peritonitis. Abdominal CT in our patient showed a large heterogeneous liver mass in the right lobe. Hepatic malignant tumor with a high possibility of hepatoblastoma was diagnosed. The tumor was surgically removed and confirmed to be UESL by postoperative pathology and immunohistochemical staining analysis. The key management of the disease is total resection followed by postoperative combined therapeutic measures, including chemotherapy, radiotherapy and interventional therapy, which are able to significantly improve survival rates. Unfortunately, the patient in our report then rejected chemotherapy. To date, the patient has survived for 50 months, and demonstrated a right liver mass 3 years post 1st-surgery. Microscopic examination revealed that the tumor was evidence of undifferentiated embryonal sarcoma recurrence. The patient is currently in a good general condition without evidence of local metastasis or recurrence, when it has been 6 months after the second surgical procedure. Although UESL has a high malignancy and a poor prognosis, cases of long-term survival with improved diagnosis and therapy have recently been reported. Therefore, it has been proposed that UESL should not be considered as a hepatic tumor with a poor prognosis. Total resection with preoperative or postoperative radio-chemotherapy is currently considered to be the key treatment choices for USL, especially with intraperitoneal rupture, are uncertain.

Early diagnosis and treatment would save life of many patients who presented with malignant tumor in children related acute abdomen. Surgery is still the gold standard treatment in localized malignant tumor in children. The prognosis is strictly related to size and completeness of surgical resection. We strongly advocate that all patients with a malignant tumor in children be carefully and regularly followed-up for an indefinite period. The large number of patients in this series is an alarming signal for further studies to elucidate the pathogenesis of these diseases.

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