

CASE REPORT

A large superior mesenteric artery aneurysm and ileal obstruction: a rare presentation of polyarteritis nodosa in an infant

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Abstract

Polyarteritis nodosa (PAN) is a rare form of vasculitis that occurs in childhood and affects small- and medium-sized arteries. Large aneurysms due to PAN can induce fatal complications like rupturing or occlusion of the affected arteries. Here, we report a case of a 4-month-old girl with PAN complicated by a large superior mesenteric artery aneurysm and ileal obstruction. We controlled her blood pressure to prevent the artery from rupturing. A combination of prednisolone, intravenous cyclophosphamide, and plasma exchange reduced the inflammation. She developed mechanical ileus due to ileum stricture and underwent bowel resection. Histopathological examinations revealed reparative arteritis around the healed ulcer. Her postoperative course was uneventful without further dilatation of the aneurysm. This case highlights the importance of intensive immunosuppressive therapy and appropriate blood pressure control in pediatric patients with PAN complicated by large aneurysms. Mechanical ileus can develop and may require surgical management even after remission of vasculitis.

INTRODUCTION

Polyarteritis nodosa (PAN) is a form of systemic vasculitis that affects small- and medium-sized arteries. It is characterized by necrotizing inflammatory changes and angiographic

abnormalities including aneurysms, stenosis or occlusion of the affected arteries [1]. However, large aneurysms involving the superior mesenteric artery (SMA) are rare [2]. PAN is usually diagnosed in middle-aged or older patients, rarely during infancy [3]. Here, we report a case of PAN complicated by a

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Figure 1: Abdominal contrast-enhanced computed tomography (CT) findings. Axial (A) and sagittal (B) CT images showed segmental and aneurysmal dilatation (13 × 32 mm in diameter) (arrow) at the proximal SMA.

large SMA aneurysm and ileal obstruction diagnosed during infancy.

CASE REPORT

A 4-month-old girl with no significant past medical history was admitted with melena and fever. She exhibited hypertension (127/100 mmHg) and tachycardia (180 bpm). Laboratory testing revealed leukocytosis (18 950/ μ l [normal range: 3500–9800/ μ l]), moderate anemia (hemoglobin 9.8 g/dl [normal range: 11.0–16.0 g/dl]), elevated C-reactive protein (CRP) level (95.3 mg/l [normal range: <3.0 mg/l]) and high erythrocyte sedimentation rate (66 mm/h [normal range: 3–15 mm/h]). Contrast-enhanced computed tomography demonstrated proximal SMA segmental and aneurysmal dilatation (diameter: 13 × 32 mm; Fig. 1). She was then transferred to our hospital for further management. Under sedation, nitroglycerin and furosemide were administered to lower blood pressure and prevent aneurysm rupture. Although she lacked typical findings that are associated with Kawasaki disease, we administered intravenous immunoglobulin (IVIG) (2 g/kg), which resulted in minimal inflammation improvement. Additional testing revealed that anti-nuclear antibodies, proteinase 3-antineutrophil cytoplasmic antibodies, myeloperoxidase-antineutrophil cytoplasmic antibodies, hepatitis B surface antigen and tuberculosis skin test were negative. Interleukin 6 levels were elevated (91.5 pg/ml [normal range: <4.0 pg/ml]). The patient fulfilled the PAN diagnostic criteria proposed by the European League against Rheumatism and the Pediatric Rheumatology European Society (Table 1) [4].

She received plasma exchange (PE) for 5 days. Prednisolone (2 mg/kg/day) was initiated after IVIG. Five doses of intravenous cyclophosphamide were subsequently administered at 3- to 4-week intervals (250 mg/m² for the first and second doses and 333 mg/m² for subsequent doses). Ultrasonography revealed

Table 1: Diagnostic criteria of PAN proposed by the European League against Rheumatism and the Pediatric Rheumatology European Society

Classification criteria for childhood PAN	This case
Systemic illness characterized by:	
Histopathological findings of necrotizing vasculitis of medium- or small-sized arteries, or	+ *
Angiography showing aneurysm, stenosis or occlusion of medium- or small-sized arteries	+
Plus \geq 1 out of 5 of the following	
Skin involvement (livedo reticularis, tender subcutaneous nodules, superficial skin infarctions, ecchymosis or deep skin infarctions)	–
Myalgia or muscle tenderness on examination	–
Hypertension (BP >95th percentile for height)	+
Peripheral neuropathy (motor mononeuritis multiplex, sensory peripheral neuropathy)	–
Renal involvement (proteinuria >0.3 g in 24 hours, hematuria or RBC casts, impaired renal function)	–

*Histopathological findings were obtained by the bowel resection at the age of 10 months.

BP, blood pressure; RBC, red blood cell.

aneurysmal intramural thrombosis; therefore, unfractionated heparin (100 U/kg/day) was administered intravenously. The intravenous unfractionated heparin dose was adjusted up to 400 U/kg/day to achieve an activated partial thromboplastin time (aPTT) within 46–70 seconds. Aspirin was substituted for heparin after 30 days since the thrombosis diminished. Blood pressure and CRP levels normalized and the size of the aneurysm remained unchanged. She was discharged home on a tapering course of prednisolone.

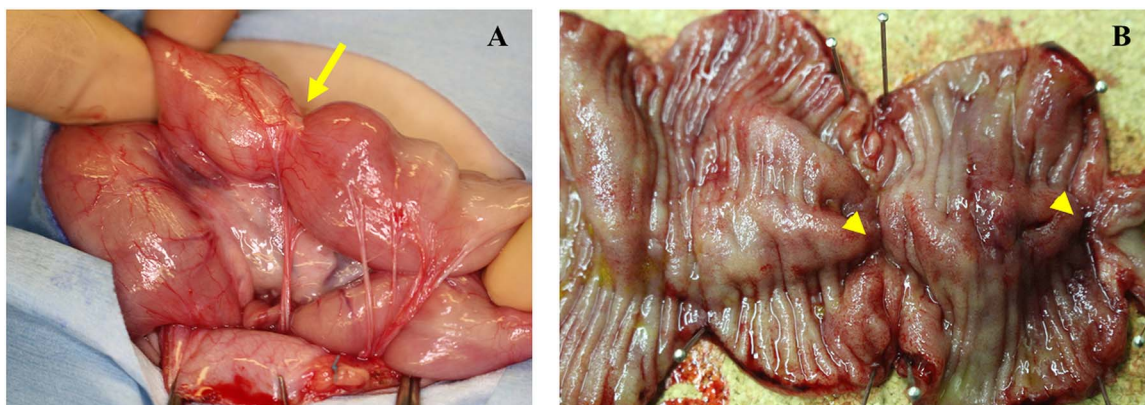


Figure 2: (A) Gross appearance of the ileal stenosis (arrow) with dilated intestinal loops and (B) healed ulcers (arrowheads) in the ileal lumen.

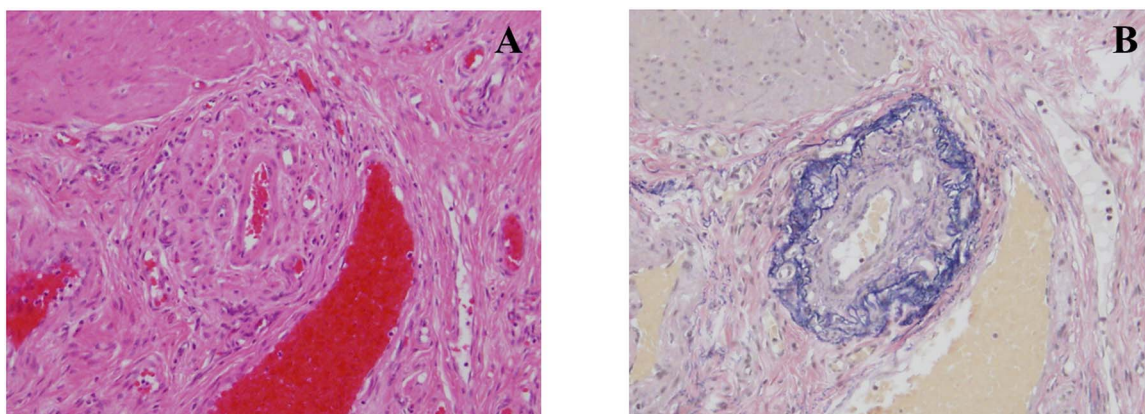


Figure 3: Histopathological analyses of the small arteries of the intestinal wall showed fibrocellular thickening of the intima, stratification of the internal elastic lamina and small vessel proliferation in the media and the adventitia, which were consistent with findings for the reparative stage of arteritis.

At the age of 7 months, the patient developed melena. We discontinued aspirin because peptic ulcer was considered as a differential diagnosis. Her stool became normal and laboratory data showed signs of recovery from anemia. Shortly after feeding her milk and solid food, she vomited and showed abdominal distention. She recovered conservatively but repeatedly exhibited ileal obstruction. An enema examination revealed multiple strictures of the terminal ileum and dilatation of the oral side. She underwent laparotomy at the age of 10 months. Two stenotic sites and inflammatory adhesions were observed at the terminal ileum (Fig. 2). One of the severe strictures of the ileum caused obstruction, and two lesions were resected. Histopathological examination revealed reparative arteritis around the multiple healed ulcers at the ileum stenotic site (Fig. 3), which the healing stage of PAN. The patient followed an uneventful postoperative course without further dilatation of the aneurysm.

DISCUSSION

To our knowledge, this is the first report of infantile-onset PAN complicated by an SMA aneurysm and ileal obstruction that was successfully managed with immunosuppressants and surgical intervention.

The annual incidence of PAN is reported to be 0.7 per million children [5]; however, PAN is rarely reported in infancy [3]. Although aneurysms are common findings associated with PAN, large aneurysms are rarely reported in the SMA [1]. Previous

case studies have reported rupturing of visceral arteries with aneurysm caused by PAN [6]. Litt *et al.* [7] reported an adult case of vasculitis in which the ruptured superior pancreaticoduodenal artery was embolized and aortic ulceration was treated with an endovascular stent. Our case was a 4-month infant; therefore, it was impossible to perform endovascular treatment or repair the aneurysm with a prosthetic graft. Hence, we tried to lower her blood pressure to <100/65 mmHg or within the 90th percentile for 4-month-old girls [8] and sedated her to decrease the risk of aneurysm rupture.

She responded to a combination therapy of prednisolone, intravenous cyclophosphamide and PE. IVIG was not effective in our case. Eleftheriou *et al.* [3] reported that 83% of pediatric patients with PAN received induction therapy with a combination of corticosteroids and cyclophosphamide and only 9% received PE. Although PE may be unnecessary in most cases, it can rapidly suppress inflammation and inhibit the large artery aneurysm rupture. Which PAN patients require PE remains unclear.

We used unfractionated heparin to treat intramural thrombosis in the aneurysm. The intravenous dose was adjusted to achieve an aPTT within 46–70 seconds, which corresponded to an aPTT ratio of 1.5–2.3 times normal values. Our target therapeutic range was consistent with Japanese guidelines published in 2009 [9]. We chose intravenous heparin over subcutaneous low-molecular weight heparin (LMWH) because data about efficacy and safety of LMWH in infants are limited. In addition, LMWH is not approved for clinical use for children in Japan.

Our patient developed mechanical ileus after inflammation resolution. Only 10% of pediatric PAN cases show severe gastrointestinal involvement [3]. Venuta et al. [10] reported a pediatric PAN case and included a literature review. They described that four of seven reported cases died from gastrointestinal tract perforation or vascular occlusion. Gastrointestinal manifestations are infrequent but can be fatal without prompt surgery. In our case, inflammation of the affected arteries induced ileal ischemia and led to ulcer development. It remains possible that intensive induction therapy prevented perforation of the intestinal wall. However, ileal strictures formed during the healing process resulted in mechanical ileus. Further studies are necessary for establishing better strategies for avoiding severe gastrointestinal complications.

In conclusion, our case study suggests that pediatric PAN complicated by large SMA aneurysms is better treated by intensive induction therapy and appropriate blood pressure control. Mechanical ileus can develop even after the remission of vasculitis; close cooperation with pediatric surgeons is essential for managing gastrointestinal complications.

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CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

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ETHICAL APPROVAL

This report was approved for publication by the ethical committee of Gunma Children's Medical Center.

CONSENT

Informed consent was obtained from the patient's parents for publication of this case report.

GUARANTOR

Dr. Akihiko Shimizu

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