

# An Unusual Case of IgA Vasculitis with Multisystemic Involvement

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## Research Article

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# Abstract

**Background:** Immunoglobulin A vasculitis (IgAV) is one of the most common vasculitis in children. It is generally a self-limiting disease. Due to its systemic nature, a variety of symptoms in different organs can be observed. We report a case of IgAV characterized by several complications to improve clinicians' understanding of the disease.

**Case presentation:** A 4-year-old boy was admitted to a local hospital because of abdominal pain and skin rash. The skin biopsy showed leukocytoclastic vasculitis with IgA deposition, consistent with a diagnosis of IgAV. He developed clinical signs of intussusception and laparotomy was undertaken. He continued to have intermittent abdominal pain and edema in the four limbs with oliguria. Elevated pancreatic enzymes and swelling of the pancreas on abdominal ultrasound suggested a combination of pancreatitis in the child. The child subsequently developed headache, dizziness and convulsions, and head MRI showed a high signal on the left side of the cortex and subcortical white matter, and he was considered to have developed cerebral vasculitis. He underwent bronchoscopy because of respiratory distress, which confirmed the presence of pulmonary hemorrhage. Combined pulmonary infections added to the severity and complexity of his condition. He received two courses of methylprednisolone pulse therapy combined with IVIG and aggressive anti-infective therapy, but his condition eventually deteriorated and he died.

**Conclusions:** IgAV can involve multiple systems and various complications. There is no definitive evidence to support a single drug or multi-immunosuppressive regimen. IgAV usually runs a benign course, however, the severe cases are critical, with a high mortality rate.

## Background

Immunoglobulin A vasculitis (IgAV), formerly called Henoch-Schönlein purpura<sup>[1]</sup>, is the most common form of leukocytoclastic systemic small-vessel vasculitis in children. The annual incidence of IgAV in children is estimated to be 15/100,000 cases<sup>[2]</sup>. More than 90% of patients are under the age of 10 (mean age, 6 years)<sup>[3]</sup>. The main clinical manifestations of IgAV usually characterized by cutaneous palpable purpura, arthritis or arthralgia, abdominal discomfort, and renal involvement. Other possible complications include intussusception, intestinal perforation, cerebral vasculitis, pulmonary hemorrhage, ureteral or bladder disease, and scrotal, penile, or testicular hemorrhages. The prognosis of IgAV is generally excellent, but severe complications can occur. Here we present a complicated case: a pediatric patient with IgAV who demonstrated the simultaneous involvement of multiple organ systems.

## Case Presentation

A 4-year-old male patient was admitted to a local hospital with the complaints of abdominal pain and skin rash for 17 days. Except for an upper respiratory tract infection 5 days earlier, his previous medical history was unremarkable. His birth and development appeared normal. He had purpuric rash on his

lower extremities and feet which were not faded by pressing. The abdominal pain was periumbilical, intermittent and associated with vomiting. Sometimes the pain was aggravated by food intake. Based on these clinical findings, the patient was diagnosed as IgAV with gastro-intestinal involvement, and the skin biopsy showed leukocytoclastic vasculitis with IgA deposition. Methylprednisolone (1mg/kg/day) was administered, and his abdominal pain quickly improved. Three days later, he complained of increased abdominal pain. Abdominal ultrasonography showed ileo-ileal intussusception. At laparotomy necrotic bowel and intramural hematoma were found, and a partial bowel resection (28 cm) with termino-terminal anastomosis was performed. Histological examination of the ileocecum demonstrated leukocytoclastic vasculitis with IgA deposition, supporting the clinical diagnosis of IgAV. In the following days, he showed ecchymosis of the scrotum, still intermittent abdominal pain, edema in the four limbs with oliguria. On postoperative day 14, he was transferred to our hospital for further evaluation and management.

Physical examination upon arrival revealed the following: the body temperature was 37.0°C, the pulse was 80 bpm, the breathing frequency was 20 times/min, the blood pressure was 109/57 mmHg. The patient's consciousness was normal. He had purpuric rash on his lower extremities and scrotum. The patient was also noted to have generalized depressed edema. There was diffuse abdominal pain and tenderness, especially in the epigastric and umbilical regions. Bowel sound was diminished. Other examinations were unremarkable.

Laboratory tests showed the following: white blood cell (WBC),  $10.98 \times 10^9/L$  with normal differentiation, platelet count,  $153 \times 10^9/L$ , C-reactive protein, 13 mg/L; and serum albumin, 18.9 g/L (35–55 g/L); serum urea nitrogen, 6.8 mmol/L (1.7–7.1 mmol/L); serum creatinine, 35.1  $\mu\text{mol/L}$  (30–104  $\mu\text{mol/L}$ ), total cholesterol, 2.39 mmol/L (1.8–5.2 mmol/L), serum amylase, 298 U/L (0–125 U/L), serum lipase, 292.6 U/L (0–39 U/L). D-dimer, 2.408 mg/L (0–0.243 mg/L), prothrombin time, partial thromboplastin time, and international normalized ratio were all within normal limits. Complement C3, 0.32 g/L (0.85–1.93 g/L); complement C4, 0.061 g/L (0.12–0.36 g/L). antineutrophil antibodies, antinuclear antibodies, double-stranded DNA anti-Sm, and anti-SSA/anti-SSB were negative. Immunological laboratory tests (IgG, IgA, IgM levels, and lymphocyte subpopulations) were normal. Urinalysis indicated hematuria and proteinuria, and proteinuria was quantified at protein of 4.9 g/d. Urine protein to creatinine ratio was 11.93 mg/mg. An acute abdomen ultrasound demonstrated extensive swelling of the small intestine, swelling pancreas with homogeneous echogenicity, and a small amount of free ascites. A chest and abdominal radiograph indicated that the upper right lung was slightly dense and showed air-liquid level in the lower right abdomen. A kidney biopsy was recommended, but the patient's parents refused.

According to the European League Against Rheumatism (EULAR)<sup>[4]</sup>, the patient was diagnosed IgAV with gastrointestinal, pancreatic and renal involvement. The patient was started on total parenteral nutrition. He initially received daily intravenous methylprednisolone (2 mg/kg/d), somatostatin, pantoprazole, transfusion of human albumin for hypoalbuminemia, and intravenous furosemide for edema.

On the second hospital day, the patient developed headache, dizziness and convulsions. He had a clonic seizure in his limbs for one minute, developed binocular gazing, and gradually lost consciousness.

Diazepam was injected for control the seizure. His blood pressure was controlled around 120/85 mmHg. Further work included a magnetic resonance imaging(MRI) (Fig. 1A)of the brain, which showed a high signal of left parietal occipital cortex and subcortical white matter in fluid attenuated inversion recovery(FLAIR). Magnetic resonance angiography(MRA) (Fig. 1B) showed filling defect in the lateral part of the left transverse sinu, suggesting ischemic lesions. Electroencephalogram (EEG) revealed slow-wave activity in the left occipital region. A lumbar puncture was performed. Examination of the cerebrospinal fluid revealed a WBC count of  $8 \times 10^6/L$ , no red blood cells, and normal protein, sugar and cerebrospinal fluid pressure. Subsequent bacterial cultures, herpes simplex polymerase chain reaction, and oligoclonal banding were negative. Neurology consultations were requested to assist in treatment and suggested that the patient developed cerebral vasculitis due to IgAV. Therefore, the patient received a three-day course of methylprednisolone (20 mg/kg/d) pulse therapy, followed by intravenous cyclophosphamide (750 mg/m<sup>2</sup>) along with intravenous methylprednisolone (2 mg/kg/d).

On day 8, the patient developed dyspnea and required non-invasive ventilation (Nasal Continuous Positive Airway Pressure). High-resolution computed tomography scans (Fig. 1C) showed interstitial and parenchymal infiltration in both lungs, with bilateral pleural effusion. The patient underwent a thoracentesis. Cultures from the pleural fluid revealed no growths. The dyspnea was relieved after a bronchoalveolar lavage. We suspected that the pulmonary involvement may be caused mainly by vasculitis, but pulmonary infection could not be completely excluded. He was treated with intravenous cefoperazone sulbactam sodium for presumed pulmonary infection.

On day 14,the patient was started on continuous renal replacement therapy (CRRT) for his renal function deteriorated with repeated edema and oliguria.

On day 20, amylase and lipase levels returned to normal. Repeat ultrasound showed no swelling, bleeding or necrosis in the pancreas.

On day 24, the patient suddenly presented with severe respiratory distress, perioral cyanosis, red secretions in the nasal cavity, and a second convulsion attack. During intubation, blood was seen in the trachea. A repeat chest X-ray (Fig. 1D) showed multiple patchy shadows in both lungs, and the lesions were more severe than before. Bronchoscopy was repeated, which identified active bleeding in the right upper lobe. Intravenous gammaglobulin(IVIG) treatment (2g/kg) was initiated.

On day 28, the patient had a fever (38.7°C). Laboratory investigations showed an increases in C-reactive protein (133 mg/L). *Serratia marcescens* (Carbapenemase (+) ) was isolated from bronchoalveolar lavage fluid cultures. The patient's antibiotics were changed to cefepime.

On day 35, the patient's respiratory status continued to deteriorate, and methylprednisolone (15 mg/kg/d) pulse therapy was given for three consecutive days with no significant improvement. The patient's cardiac function decreased with an ejection fraction of 57%. The subsequent course was characterized by worsening renal function, respiratory failure and heart failure. The patient died despite maximal supportive care.

## Discussion

IgAV is the most common systemic vasculitis seen in childhood, characterised by the deposition of IgA with complex and complementing component 3 proteins on arterioles, capillaries, and venules. These mainly invade the skin, joints, kidneys, gastrointestinal mucosa, and serosa. Gastro-intestinal manifestations occur in approximately two-thirds of children during the course of IgAV. Abdominal pain is a common symptom, with the small intestine being the most frequently involved site, usually due to swelling caused by submucosal and subserosal hemorrhages, and edema from vasculitis. Severe gastro-intestinal tract involvement can lead to intussusception, bleeding, bowel infarction, intestinal perforation, and necrosis. Our patient developed severe gastrointestinal complications that eventually led to intussusception and underwent surgery. Unfortunately, details of the procedure are not available as it occurred prior to the patient's admission to our hospital.

Acute pancreatitis (AP) is a rare gastrointestinal complication of IgAV. IgAV patients with severe abdominal pain should be evaluated for pancreatitis. Tests for elevated pancreatic enzymes levels are useful tools for early diagnosis of IgAV-associated pancreatitis<sup>[5]</sup>. The imaging changes in pancreatitis can be atypical. Ultrasonography or radiological signs, such as a swollen pancreas, necrosis, or pseudocysts, can confirm the diagnosis only if the pancreatitis is complicated. Diagnosis of AP in our case was confirmed based on the presence of abdominal pain, elevated serum lipase levels (three times more than the upper limit of the normal range) and swelling of the pancreas observed on ultrasound. There was no prominent cause, such as biliary stones, overeating, drinking, hyperlipidemia, infection, toxins or drugs. The pathogenesis of AP remains unclear. It may be associated with small blood vessel thrombosis and vasculitis, which activate digestive enzymes and lead to inflammation, edema, vascular damage, and even cellular death.

In addition to the symptomatic and supportive treatment provided, intravenous methylprednisolone was also administered as soon as the diagnosis was established in our case. A recent review of IgAV related pancreatitis cases highlighted the importance of timely steroid therapy in order to obtain a good outcome<sup>[6]</sup>. Methylprednisolone pulse therapy can be used and is effective in alleviating symptoms of AP<sup>[7]</sup>. When the clinical manifestation is steroid-resistant, other immunosuppressive therapies are required. A single high dose of cyclophosphamide may be beneficial in IgAV cases with severe pancreatic involvement that are non-responsive to steroids<sup>[8]</sup>. Somatostatin can inhibit the secretion of stomach and pancreas, thus reducing the enzyme activity. It also reduces capillary permeability, opens the oddi sphincter, and promotes pancreatic enzyme excretion. Somatostatin may be applied after the diagnosis of AP<sup>[6]</sup>. Our patient's symptoms of pancreatitis resolved after using intravenous methylprednisolone plus pulse methylprednisolone.

Our patient experienced two convulsions during the course of the disease. The most likely diagnosis in this case was IgAV-associated encephalopathy. This was deduced after excluding systemic lupus erythematosus, other connective tissue diseases, systemic vasculitides, metabolic diseases, head traumas, and neurological diseases such as intracranial infections. The pathogenesis of IgAV-associated

encephalopathy is not thoroughly understood. It may be a result of central nervous system vasculitis, or related to arterial hypertension in cases of IgAV nephritis<sup>[9]</sup>. Murakam, et al.<sup>[10]</sup> reported a brain biopsy of a fatal case of IgAV, and demonstrated leukocytoclastic vasculitides that had IgA deposits in the vessel walls. However, such histological confirmations are very rare, the diagnosis usually depends upon imaging examination.

MRI is more sensitive than computed tomography for diagnosis of cerebral vasculitis. The most commonly reported lesions were ischemic, followed by hemorrhagic<sup>[11]</sup>. Typically, 14% of neuroimaging results are normal. The diagnosis of cerebral vasculitis may be difficult since neurological abnormalities can be caused by multiple factors. Pulsed methylprednisolone is used as first-line treatment for IgAV encephalopathy<sup>[12]</sup>. Other immunosuppressive drugs, such as cyclosporine A, cyclophosphamide, or mycophenolate mofetil, may be selected for patients who exhibit corticosteroid dependence or who do not respond to treatment. IVIG might be a safe method to treat the cerebral manifestations of IgAV<sup>[13]</sup>. Another treatment is plasmapheresis, which is believed to regulate the immune response by eliminating and reducing the plasma immune mediators that are responsible for IgAV<sup>[14]</sup>. Our patient was treated with methylprednisolone and cyclophosphamide pulse therapy for his first convulsive, but the disease progressed rapidly. Less than a month later, he had another convulsive with a combination of pulmonary hemorrhage.

The occurrence of pulmonary hemorrhage is an unusual and fatal manifestation. The pulmonary hemorrhage symptoms vary greatly, ranging from mild cough, epistaxis, hemoptysis, fatigue, and altered activity tolerance to tachypnea, chest pain, dyspnea, and respiratory failure. A chest X-ray is the first step in the diagnosis of pulmonary hemorrhage, and the common imaging findings are fluffy, diffuse, patchy opacities or opacities with a ground-glass appearance. A bronchoalveolar lavage can be performed to confirm diagnosis and start treatment simultaneously. Pulmonary hemorrhage is likely due to diffuse vasculitis. The deposition and fragmentation of IgA immune complexes, as well as the adhesion of masses of white blood cells are the main causes of pulmonary hemorrhage<sup>[15]</sup>. Lung biopsy reported by W K Wright<sup>[16]</sup> revealed intra-alveolar hemorrhage with vasculitis of the small blood vessels.

Guidelines regarding the treatment of pulmonary hemorrhage associated with IgAV have not been established, and current recommendations are based on retrospective studies. High dose intravenous pulse methylprednisolone has been recommended as a first-line treatment. Immunosuppressive therapy such as pulsed cyclophosphamide should be considered during respiratory failure. Our patient had been treated with pulsed methylprednisolone and cyclophosphamide prior to the pulmonary hemorrhage, but this did not prevent the development of pulmonary involvement. Considering the possibility of pulmonary infection, we opted for a safe IVIG therapy. However, his pulmonary symptoms did not improve. The optimal treatment for pulmonary involvement in IgAV is still controversial and needs further study.

Renal function is impaired in approximately 50% of patients with IgAV, and may manifest in many ways, from asymptomatic hematuria and proteinuria, to nephritic syndrome, rapidly progressive glomerulonephritis, and even renal failure<sup>[17]</sup>. A renal biopsy is recommended if severe proteinuria occurs

in children with IgAV. Renal histopathology is the most reliable standard to identify the stage of the disease and predict prognosis. Severe IgAV nephritis is treated usually with high-dose corticosteroid and intravenous cyclophosphamide to induce remission, and lower doses of corticosteroid combined with azathioprine or mycophenolate mofetil as maintenance treatment<sup>[18]</sup>. In the present case, the patient's massive proteinuria and intermittent oliguria suggested a serious condition. The lack of renal biopsy made it impossible to determine the pathological changes in the kidney, which was a drawback of our case. The rapid progression of the patient's disease might be a function of vasculitis or might be due to severe infection. A total of seventeen CRRT treatments, two methylprednisolone pulse and one cyclophosphamide pulse therapy failed to alleviate the patient's symptoms.

IgAV can and often involves vasculitis in many organs at one time, which can lead to numerous complications. Our patient's clinical manifestations were complex and difficult to control. The coexistence of skin lesions, abdominal pain, severe nephritis and IgA deposits on skin and ileocecum biopsy confirmed the diagnosis of IgAV. Pancreatic, pulmonary and CNS involvement were also seen in our case. The SHARE group<sup>7</sup> recommends that corticosteroid application should be considered for complications of IgAV including nephritis, orchitis, cerebral vasculitis, pulmonary hemorrhage and severe gastrointestinal involvement. In severe cases (e.g., severe cerebral, pulmonary or gastrointestinal involvement), pulsed intravenous methylprednisolone over three consecutive days may be considered. Corticosteroids were our main treatment, including methylprednisolone pulse therapy. Cytotoxic immunosuppressants or even plasma exchange may be added when the condition is complicated or when there are life-threatening involvement. However, there is no standard treatment.

In our case, cyclophosphamide was used for the patient's first convulsive because he also had severe nephritis at that time. This treatment was also seen in other cases and was successful<sup>[14]</sup>. However, our patient was not in remission and subsequently developed pulmonary hemorrhage and pulmonary infection. IVIG might be a safe method. The exact mechanism of IVIG is complex and not fully understood. The interaction between IgG-Fc fragments and Fcγ receptors on target cells seems to be the key to anti-inflammatory effects<sup>[19]</sup>. Unfortunately, a second methylprednisolone pulse after IVIG still failed to relieve the condition of our patient.

In summary, we report a case of IgAV with multisystemic involvement. IgAV is a multisystem disorder and usually runs a benign course. However, the severe cases are critical, with a high mortality rate. The available data suggest an aggressive treatment with pulse methylprednisolone, especially for severe cases. Various immunosuppressive agents have been used in the treatment of multiorgan involvement in IgAV, but there is no definitive evidence to support a single drug or multi-drug regimen. Further studies need to analyse the clinical course and the response to different protocols of treatment in patients with multisystemic involvement associated with IgAV.

## Abbreviations

IgAV: Immunoglobulin A vasculitis; WBC: White blood cell; MRI: Magnetic resonance imaging; FLAIR: Fluid attenuated inversion recovery; MRA: Magnetic resonance angiography; EEG: Electroencephalogram; CRRT: Continuous renal replacement therapy; IVIG: Intravenous gammaglobulin; AP: Acute pancreatitis;

## **Declarations**

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### **Author Contributions**

Lina Du, Chang Liu designed the study, collected the data and drafted the manuscript. Panpan Wang, Shaojing Li and Ziyun Guo analyzed the data and played an important role in interpreting the clinical data. Shuang Yue contributed to figure preparation. Yan Yang reviewed and revised the manuscript. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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Availability of data and materials

The raw data supporting the conclusions of this manuscript will be made available by the authors, without undue reservation, to any qualified researcher.

### **Ethics approval and consent to participate**

The case was approved by the ethics committee of Beijing Children's Hospital of Capital Medical University. Written informed consent was obtained from the parents for publication of this case report.

Consent for publication

Publication consent was obtained from the patient and her guardian.

Competing interests

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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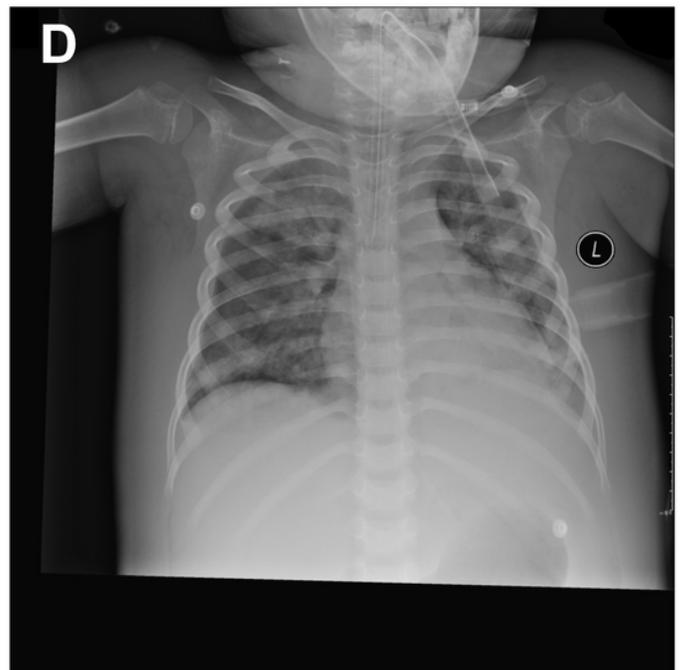
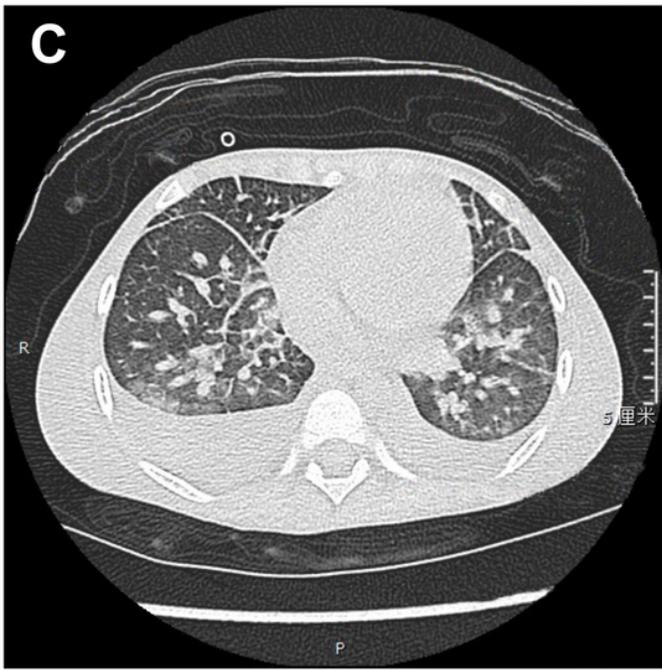
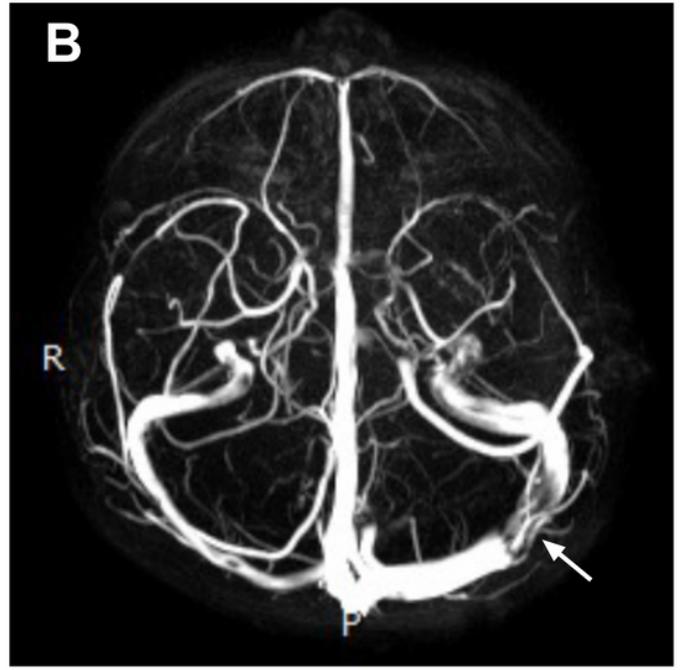
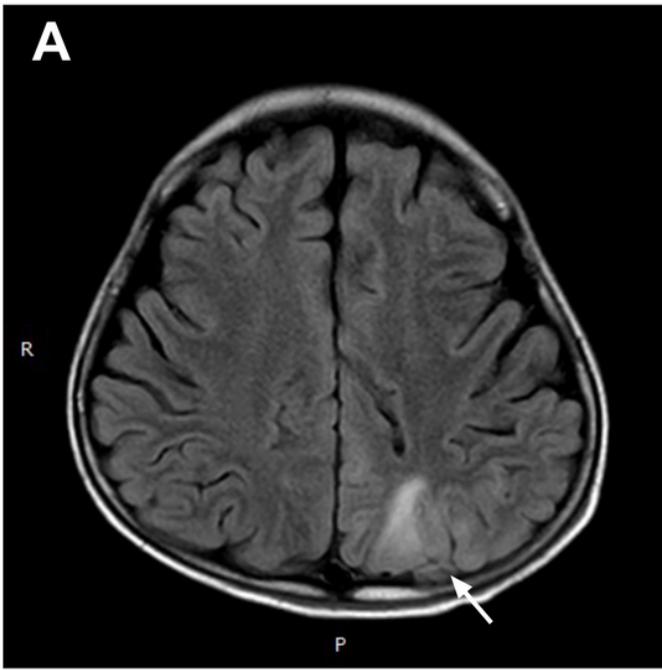
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## Figures



**Figure 1**

a FLAIR image of brain MRI showing a high signal of left parietal occipital cortex and subcortical white matter (arrow). b MRA showing filling defect in the lateral part of the left lateral transverse sinus. c Chest CT scan showing interstitial and parenchymal infiltration between the two lungs, with bilateral pleural effusion. d Chest radiograph showing multiple patchy shadows in both lungs.