## REFERENCES

1 **Lowder CY**, Foster RE, Gordon SM, Gutman FA. Acute posterior multifocal placoid pigment epitheliopathy after acute group A streptococcal infection. Am J Ophthalmol 1996;122:115–17.
2 **Chiquet C**, Lumbroso L, Denis P, Papo T, Durieu I, Lehoang P. Acute posterior multifocal placoid pigment epitheliopathy associated with Wegener's granulomatosis. Retina 1999;19:309–13.
3 **Brezin AP**, Massin-Korobelnik P, Boudin M, Gaudric A, Lettoang P. Acute posterior multifocal placoid pigment epitheliopathy after hepatitis B vaccine. Arch Ophthalmol 1995;113:297–300.
4 **Gass JDM**. Acute posterior multifocal placoid pigment epitheliopathy. Arch Ophthalmol 1968;80:177–85.
5 **Charteris DG**, Khanna V, Dhillon B. Acute posterior multifocal placoid pigment epitheliopathy complicated by central retinal vein occlusion. Br J Ophthalmol 1989;73:765–8.
6 **Ghazi NG**, Gollance SA, Green WR. Choroidal vascular occlusion in a child with a connective tissue disease and complement C4 deficiency. Ophthalmology 2002;109:1272–7.
7 **Kleiner RC**, Najarian LV, Schatten S, Jabs DA, Patz A, Kaplan HJ. Vaso-occlusive retinopathy associated with antiphospholipid antibodies (lupus anticoagulant retinopathy). Ophthalmology 1989;96:896–904.
8 **Uthman I**, Gharavi AE. Viral infections and antiphospholipid antibodies. Semin Arthritis Rheum 2002;31:256–63.


# Multifocal lymphadenopathy associated with severe Kawasaki disease: a difficult diagnosis

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## CASE REPORT

A previously healthy 3½ month old infant was admitted to the hospital with dyspnoea, malaise, and irritability in August 2001. One week before he had had an upper respiratory infection treated with amoxycillin and inhaled steroids; the day before admission he presented high grade (up to 40°C) temperature.

On admission he was febrile (39°C), pale, restless; his respiratory rate was 50/min and pulse 120 beats/min. Clinical evaluation showed oral cyanosis, redness of the pharynx, and bilateral suppurative conjunctivitis. A chest radiograph showed pulmonary infiltrates in the right lung.

Electrocardiography (ECG) and echo colour Doppler excluded cardiac abnormalities. An electroencephalogram showed diffuse slow waves. A lumbar puncture was performed to exclude a meningeal infection; the cerebrospinal fluid was clear with normal glucose and protein levels and a mild increase in mononuclear cells.

Blood tests showed: erythrocyte sedimentation rate (ESR) (Westergren) 120 mm/1st h, C reactive protein (CRP) 221 mg/l, white blood cells $16.8\times10^9$/l (neutrophils 75.2%), haemoglobin 94 g/l, and platelet count $504\times10^9$/l . Serum urea, creatinine, electrolytes, transaminases, γ-glutamyltransaminase, IgA, IgM, IgG, lactate, pyruvate, lactate dehydrogenase levels, and blood and urine cultures were normal. Serological tests for antibodies to measles, mumps, chickenpox, herpes viruses, adenovirus, HIV, hepatitis C and B, cytomegalovirus, *Mycoplasma pneumoniae*, *Leishmania* were all negative. Vanillylmandelic acid in the urine was in the normal range.

Despite wide spectrum antibiotic treatment (netilmicin, ceftazidime) his general condition deteriorated and he was less alert and extremely pale. He lost weight and developed liver, spleen, and cervical lymph node enlargement.

On day 5 from admission, a diffuse maculopapular rash affecting all the body and the scalp was evident. Over the following days the child had diarrhoea and developed generalised oedema. Fever persisted (up to 39.5°C).

Abdominal ultrasound confirmed hepatosplenomegaly and excluded the presence of other masses. Cardiac evaluation, with ECG and echocardiography performed on days 7 and 10 was normal.

Blood tests were repeated and showed an increased ESR (132 mm/1st h) and CRP (231 mg/ll), and decreased haemoglobin (60 g/l), fibrinogen 4.5 g/l, and platelet count $116\times10^9$/l. After blood transfusion, the child underwent abdominal and chest computed tomographic scan that showed the presence of multiple lymph nodes in the retroperitoneal, cervical, and axillar areas.

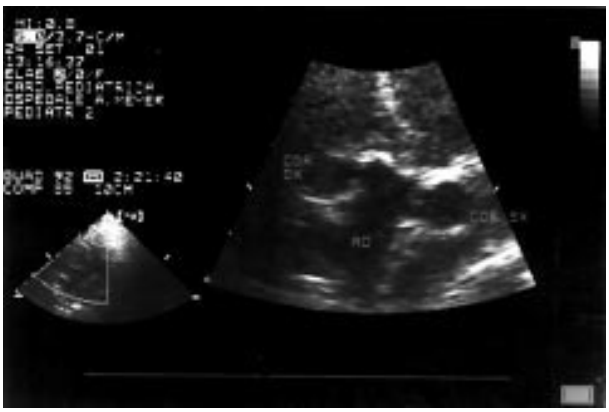

**Figure 1** Giant aneurysms. The left coronary diameter was 13 mm and the right coronary diameter 11 mm. Apical five chambers view modified for coronary arteries.

Lung infiltrates were still present. As a peripheral blood smear showed the presence of myelocytes and promyelocytes a bone aspiration was performed, and several histiocytes were found. Because a lymphoproliferative disorder was suspected a bone marrow biopsy was planned. During anaesthesia the boy underwent cardiac arrest and was transferred to the intensive care unit.

ECG showed inverted T waves and echo colour Doppler two giant aneurysms: the left coronary diameter was 13 mm and the right coronary diameter 11 mm (fig 1); on the posterior left ventricular wall a small area of myocardial necrosis was detected.

The child was then referred to our unit. Kawasaki disease (KD) was then suspected and the appropriate treatment with intravenous immunoglobulin (IVIg) (2 g/kg) was immediately given. Anticoagulation with low weight heparin was also started, owing the appearance of giant aneurisms and myocardial infarction. The fever promptly dropped during IVIg infusion, so high dose aspirin was not started. The patient improved dramatically; he became alert and active.

Blood tests showed a high platelet count $1.2\times10^{12}$/l. Three weeks after admission the patient's two digits of his right hand peeled. Blood tests progressively improved and completely normalised over four weeks. In October the child was discharged in general good condition while continuing to receive treatment with oral anticoagulant. At the last control,

May 2002, a cardiological evaluation showed a slight reduction of the coronary aneurysms (left coronary diameter 9 mm and right 10 mm), and anticoagulation was continued.

## DISCUSSION

KD is one of the most common systemic vasculitides in childhood. Owing to a lack of diagnostic tests, the diagnosis is based on clinical criteria after the exclusion of other febrile diseases in young children.[1]

The presence of histiocytes, probably a reactive event, led us to consider the diagnosis of histiocytosis. The differential diagnosis includes sepsis, scarlet fever, toxic shock syndrome, viral infections, in particular due to enterovirus, adenovirus, measles, parvovirus, cytomegalovirus, Epstein-Barr virus, and *Mycoplasma pneumoniae*[2]. Lymphoproliferative disorders are usually not included in the differential diagnosis of KD.

In fact, recently an increasing number of children with atypical onset of KD have been reported,[3–5] but to our knowledge a patient with clinical signs mimicking a lymphoproliferative disorder has not been described so far.

This case emphasises that KD must be suspected in any child with high, persistent fever, even in absence of all diagnostic criteria; IVIg can be recommended before the typical manifestations are all present.[6] Delayed treatment might be responsible for severe coronary complications, and even sudden death, especially in infants.[7]

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## REFERENCES

1 **Anonymous**. Diagnostic guidelines for Kawasaki disease. Circulation 2001;103:335–6.
2 **Petty RE**, Cassidy JT. Kawasaki disease. In: Cassidy JT, Petty RE, eds. *Textbook of pediatric rheumatology*. 4th ed. Philadelphia: Saunders, 2001:580–94.
3 **Rowley AH**, Gonzales Crussi F, Gidding SS, Duffy CE, Shulman ST. Incomplete Kawasaki disease with coronary artery involvement. J Pediatr 1987;110:409–13.
4 **Rowley AH.** Incomplete (atypical) Kawasaki disease. Paediatr Infect Dis J 2002;21:563–5.
5 **Witt MT**, Minich LA, Bohnsack JF, Young PC. Kawasaki disease: more patients are being diagnosed who do not meet American Heart Association criteria. Pediatrics 1999;104:10.
6 **Newburger JW**. Kawasaki disease. Current treatment options. Cardiovasc Med 2000;2:227–32.
7 **Oki I**, Tanihara S, Ojima T, Nakamura Y, Yanagawa H. A multicenter collaborative study on the risk factors of cardiac sequelae due to Kawasaki disease: a one-year follow-up study. Acta Pediatr 2000;89:1435–8.

# Brain abscess in rheumatoid arthritis

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## CASE REPORT

A 62 year old man had RA for five years that had been diagnosed according to the 1987 American Rheumatic Association criteria. His disease was well controlled with disease modifying antirheumatic agents (DMARDs). However, the DMARDs were discontinued in March 2000, and he entered a leflunomide clinical trial from April 2000 to August 2000. He improved during the trial. After the trial ended and the leflunomide was discontinued, his joint pain and swelling resumed, so DMARDs were started again.

In December 2000, he arrived in our emergency department with the sudden onset of a disturbance of consciousness, fever, chills, throbbing headache, and left limb weakness for a few days. On physical examination, his temperature was 38.3°C, and he had slurred speech, isochoric pupils, a supple neck, and muscle power in all four extremities grade 3/5. However he then had a seizure. Emergency computed tomographic (CT) scanning showed decreased lucency in the right mastoid air cells and a lesion suggestive of a focal infarct or cerebritis affecting the lower part of the right posterior frontal lobe (fig 1). A lumbar puncture was performed with an opening pressure of 240 cm $H_2O$ and a closing pressure of 210 cm $H_2O$. The cerebrospinal fluid (CSF) was clear and he had a white blood cell (WBC) count of 8730/ml with 60% neutrophils and 37% lymphocytes.

The Pandy test was positive, protein 150 (10–45) mg/day. Microbiological investigations (antigens and/or culture) were all negative, as were blood cultures for bacteria and fungi. Penicillin and chloramphenicol were given but did not improve his condition. Phenytoin was also given for seizure control. Ten days after admission, magnetic resonance imaging (MRI) showed focal cerebritis with possible abscess formation in the right frontal and temporal lobes and the right insular cortex and a portion of the right basal ganglion (TR 2500/TE 100) (fig 2). A right frontotemporal craniotomy was performed to drain the abscess. The pathology report was consistent with an abscess,

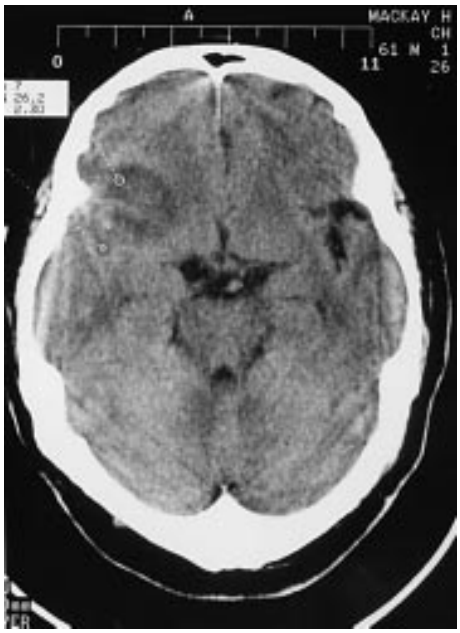


**Figure 1** CT scan of the brain shows an ill defined hypodense area in the right frontotemporal junction.