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<td><strong>Author(s)</strong></td>
<td>Tse, KC; Ooi, GC; Wu, A; Ho, PL; Ip, SK; Jim, MH; Lam, YM; Fan, YW; Tso, WK; Tsang, KW</td>
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Multiple brain abscesses in a patient with bilateral pulmonary arteriovenous malformations and immunoglobulin deficiency

K-C Tse, G C Ooi, A Wu, P L Ho, S K Ip, M H Jim, Y M Lam, Y W Fan, W K Tso and K W Tsang

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Multiple brain abscesses in a patient with bilateral pulmonary arteriovenous malformations and immunoglobulin deficiency

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CASE REPORT

A 34 year old Chinese man presented with grand mal seizures complicating multiple brain abscesses caused by mixed oral flora. Because of persistent hypoxaemia contrast spiral thoracic computed tomography was done, which revealed bilateral pulmonary arteriovenous malformations (PAVMs). Concomitant IgA and IgG subclass deficiency was also found. The combination of these two conditions appears to have predisposed this patient to presumably paradoxical septic embolism. The patient’s cerebral condition responded to postoperative antibiotic treatment and he eventually received selective coil embolisation of right lower lobe PAVMs, which relieved his hypoxaemia and dyspnoea.

Pulmonary arteriovenous malformation (PAVM) is a rare congenital vascular anomaly, associated with Osler-Weber-Rendu syndrome, initially described in 1897, and is due to formation of abnormal communications between pulmonary arteries and veins. Clinical manifestations can be subtle and this condition may remain undiagnosed until complications arise in adulthood. We describe a patient who presented with seizures secondary to multiple brain abscesses, which were presumably caused by paradoxical embolism via PAVM and predisposed by IgA and IgG deficiency.

Initial blood tests revealed normal total and differential leucocyte counts, and renal and liver function parameters. Serum IgG was 8.79 g/l (normal: 8.19–17.25 g/l), IgA <0.067 g/l (normal: 0.70–3.86 g/l), and IgM 0.68 g/l (normal: 0.55–3.07 g/l). IgG subclass analysis revealed IgG1 of 2.80 g/l (normal: 4.22–12.92 g/l), IgG2 of 0.70 g/l (normal: 1.17–7.47 g/l), IgG3 of 0.33 g/l (normal: 0.41–1.29 g/l), and IgG4 of 0.09 g/l (normal: 0–2.91 g/l). However, there was no evidence of associated autoimmune diseases with normal antinuclear antibody, C3, and C4. Serum immunoelectrophoresis was also normal, and there was no evidence of underlying haematological malignancy. Serology for HIV was negative.

The patient made an uneventful neurological recovery, and completed his course of intravenous antibiotics uneventfully. In view of the larger and more numerous PAVMS in the right lower lobe, selective coil embolisation of these right lower lobe PAVMs was performed via right common femoral vein puncture under local anaesthesia using 3–6 mm fibred platinum coil vortex 0.35 (Target Therapeutics, Boston Scientific Corporation, Freemont, USA) and 3 mm fibred platinum coil 0.035 type (Target Vascular, Boston Scientific Cork Ltd, Cork, Ireland). The left lower lobe PAVMs were not embolised. At the time of writing, six months after his discharge, he was doing well.

Abbreviations: MRA, magnetic resonance angiography; PAVM, pulmonary arteriovenous malformation
embolisation procedures, this patient has remained asymptomatic neurologically, and has a stable oxygen saturation of 96%–98% (when breathing room air). The patient is undergoing regular intravenous immunoglobulin replacement under the care of an immunologist.

DISCUSSION

Our patient presented with an interesting array of events, which could ultimately be interpreted as the complications of PAVMs and immunoglobulin deficiencies. The occurrence of multiple brain abscesses, caused by oral flora, suggests that paradoxical embolisms could have occurred as the underlying key mechanism for the development of septic embolism, presumably aggravated by underlying immunoglobulin deficiencies. Paradoxical embolism is a well-known complication of PAVMs. Other features of PAVMs include hypoxaemia, dyspnoea, clubbing, cyanosis, polycythaemia, haemoptysis, cerebral infarction, high output cardiac failure, and abnormal chest radiology. The cause of hypoxaemia in patients with PAVMs is due to persistent shunting of deoxygenated blood to the systemic circulation with a similar mechanism to cyanotic congenital heart diseases, except that this shunt is extracardiac in location. Chest radiographs may be abnormal, but changes are frequently non-specific, and the diagnosis may be made incidentally from computed tomography examination particularly in small PAVMs such as found in our case. While pulmonary angiography remains the traditional gold standard, MRA is increasingly used for the non-invasive diagnosis of PAVMs, with contrast echocardiography useful in detecting right to left shunting. Although PAVMs are rare and therefore likely to be under-diagnosed, they are present in 24% of patients with Osler-Weber-Rendu syndrome. Features of associated Osler-Weber-Rendu syndrome, namely the presence of telangiectasia, should therefore be sought in patients with PAVMs, and if present the family should be screened.

Our case is of particular clinical and academic interest as IgA and IgG subclass deficiency exists concomitantly. Selective IgA deficiency is the most common of the immunodeficiency disorders and affects one in 500 indivi-
duals. The majority of these individuals remain asymptomatic, but some patients might develop sinopulmonary infections, chronic diarrhoeal illness, and autoimmune phenomena. Selective IgA deficiency rarely leads to destructive lung disease, unless there is an associated IgG subclass deficiency, especially of IgG 2. In a recent study, 13 of 56 bronchiectasis patients had deficiencies of one or more IgG subclass, although our patient did not have bronchiectasis on computed tomography. Only one other similar case has been reported previously in the literature, which described a middle aged man with telangiectasia of the lips, tongue and PAVMs, and hypogammaglobulinaemia complicated by brain abscesses. Our case is, however, unique as there was a complete absence of cutaneous telangiectasis. A possible association between underlying immunological defects and the development of vascular malformations has recently been proposed, in view of the concomitance of these conditions among patients with ataxia telangiectasia and AIDS. However, the exact mechanism(s) underlying this potential causal relationship is obscure and clearly needs further evaluation. Patients with immunoglobulin deficiency need to be followed up regularly as some of them ultimately develop a common variable immunodeficiency. Maintenance prophylactic antibiotic or intravenous IgG replacement is beneficial, but there is a risk of anaphylactic reaction to the latter due to the inherent presence of anti-IgA in some of these patients.

**SUMMARY POINTS**

- Presentations of PAVMs are frequently subtle and should be suspected in a patient with apparently unexplained hypoxaemia.
- The gold standard for diagnosis is by pulmonary angiography and treatment with coil embolisation is effective especially for multiple PAVMs.
- There is a potential association with primary immunoglobulin deficiency.

**Authors’ affiliations**

K-C Tse, A Wu, S K Ip, M H Jim, Y M Lam, K W Tsang, Department of Medicine, University of Hong Kong, Queen Mary Hospital, Hong Kong, China

G C Ooi, W K Tso, Department of Radiology

P L Ho, Department of Clinical Microbiology

Y W Fan, Department of Neurosurgery

Correspondence to: Dr Kenneth W Tsang, Department of Medicine, Queen Mary Hospital, 102 Pokfulam Road, Hong Kong SAR, China; kwtsang@hku.hk

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