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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, K W Tsang

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 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.

CASE REPORT

A 34 year old southern Chinese man presented to our accident and emergency department with his first grand mal seizure in April 2002. There was no history of recent viral illness or systematic symptoms. A history of recurrent right foot cellulitis and benign colonic tumour, which was resected in his teenage years, was noted. His older brother suffered from an aural septal defect, which was repaired but this was later complicated by infective endocarditis. There was no recent history of dental or other surgical procedures. Physical examination revealed a fever of 38.2°C and finger clubbing. His oxygen saturation was 98% on 4 l/min oxygen therapy via a nasal cannula. There were no focal neurological signs. The patient became conscious and orientated on arrival to our accident and emergency department. Cardiorespiratory examination was normal. Urgent computed tomography of the thorax was, therefore, performed which revealed no evidence of pulmonary embolism. However, there were multiple enhancing serpinginous structures in both lung bases suggestive of small vascular masses (fig 3A and B). Magnetic resonance angiography (MRA) confirmed that these were multiple PAVMs with arterial supply from dilated descending pulmonary arteries with drainage into the pulmonary veins in both lung bases (fig 4). There were more PAVMs in the right lower lobe than in the contralateral lobe. The presence of this shunt was also confirmed by right heart catheterisation, which showed PAVMs in the right lower lobe and anterior segment of left lower lobe.

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: 0.41–1.29 g/l), and IgG3 of 0.09 g/l (normal: 0.2–1.0 g/l). C3 of 88% and 92%, arterial oxygen pressure of 8.3 kPa, and arterial carbon dioxide pressure of 5.0 kPa while breathing room air. Chest radiography showed only prominent vascular shadows but was otherwise unremarkable (fig 2). Contrast enhanced spiral computed tomography of the thorax was, therefore, performed which revealed no evidence of pulmonary embolism. However, there were multiple enhancing serpinginous structures in both lung bases suggestive of small vascular masses (fig 3A and B). Magnetic resonance angiography (MRA) confirmed that these were multiple PAVMs with arterial supply from dilated descending pulmonary arteries with drainage into the pulmonary veins in both lung bases (fig 4). There were more PAVMs in the right lower lobe than in the contralateral lobe. The presence of this shunt was also confirmed by right heart catheterisation, which showed PAVMs in the right lower lobe and anterior segment of left lower lobe.

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, Fremont, 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

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 IgG2. In a recent study, 13 of 56 bronchiectasis patients had deficiencies of one or more IgG subclass,7 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.8 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.9 10 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.11 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.12

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.

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