

Intravascular papillary endothelial hyperplasia in an intracranial thrombosed aneurysm: 3T magnetic resonance imaging and angiographical features

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ABSTRACT

Enhancement of intracranial thrombosed aneurysms is an uncommon finding on magnetic resonance (MR) imaging, and can present diagnostic difficulties and complicate management decisions. We report a 46-year-old man who had an enhancing thrombosed intracranial aneurysm observed on 3T MR imaging. There was angiographical correlation, with follow-up serial MR imaging documenting regression and improvement. Findings are typical for benign intravascular papillary endothelial hyperplasia. Differential diagnoses of giant serpentine intracranial aneurysm and malignant angiosarcoma are discussed.

Keywords: 3T magnetic resonance imaging, angiosarcoma, enhancing intracranial thrombosed aneurysm, giant serpentine intracranial aneurysm, intravascular papillary endothelial hyperplasia

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INTRODUCTION

Enhancement of intracranial thrombosed aneurysms is uncommon on magnetic resonance (MR) imaging, and can present diagnostic difficulties as well as complicate management decisions. We present the serial MR imaging findings of a patient with an enhancing thrombosed intracranial aneurysm observed on a very high field (3 Tesla [T]) MR scanner, with angiographical correlation.

CASE REPORT

A 46-year-old Chinese man presented with a two-month history of giddiness and numbness of his right upper limb. He had previously been well, with no significant prior medical history. MR imaging was performed on a very high field 3T clinical scanner (Gyrosan Intera, Philips Medical Systems, Best, The Netherlands) with the following sequences: axial T1-weighted before and after gadolinium-DTPA (TR/TE 500/8ms, 5mm thick sections/1mm interslice gap, matrix 512X 205, and 240mm FOV), axial T2-weighted (TR/TE 4000/80ms, 5mm

thick sections/1mm interslice gap, matrix 512X 300, and 240 mm FOV), axial gradient (GRE) (TR/TE 500/21ms, flip angle 18 degrees, 5mm thick sections/1mm interslice gap, matrix 512X 256, and 240 mm FOV), axial single shot spin-echo planar diffusion-weighted imaging (DWI) (TR/TE 5000/89ms, b=1000, 4mm thick sections/1mm interslice gap, matrix 215X 112, and 240mm FOV) and 3D time-of-flight (TOF) MR angiography (MRA) (TR/TE 18/3.2ms, flip angle 20 degrees, 5mm thick sections, matrix 512X 400, and 200mm FOV).

A 1.8 x 1.6 x 1.9cm mass was noted at the distal right vertebral artery, just proximal to and near the origin of the posterior inferior cerebellar artery (PICA). There was no significant flow signal within the lesion on 3D TOF MRA. The lesion was isointense with heterogeneous areas of high signal intensity on T1-weighted images (Fig. 1a), and was hypointense on T2-weighted images (Fig. 1c). There was local mass effect against the adjacent brain stem but no evidence of T2 prolongation in the brainstem to suggest oedema, ischaemia or infarct. Following intravenous administration of gadolinium-DTPA, there was heterogeneous contrast enhancement within the mass (Fig. 1b). There was no evidence of prior subarachnoid haemorrhage. MR imaging findings suggested a thrombosed aneurysm at the distal right vertebral artery, which showed enhancement.

Digital subtraction angiography (Fig. 1d) showed a small-calibre right vertebral artery terminating into smaller branches at the region of the foramen magnum, around a rounded mass. This eventually reformed into the right PICA. No early venous drainage was noted. The left vertebral and basilar arteries were essentially normal. The angiographical diagnosis was that of a thrombosed aneurysm, with differential consideration of a vascular wall tumour. Close clinical and MR imaging follow-up at one month and six months intervals was performed (Figs. 2a-b). These showed significant decrease in size of the thrombosed aneurysm, with some enhancement within the thrombus and in its peripheral wall. In view of the decrease in size of this aneurysm and

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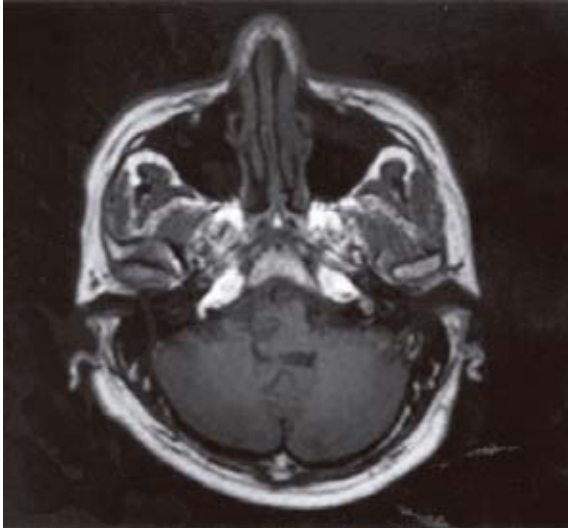


Fig. 1a Unenhanced axial T1-W MR image shows a thrombosed aneurysm arising from the proximal right vertebral artery.

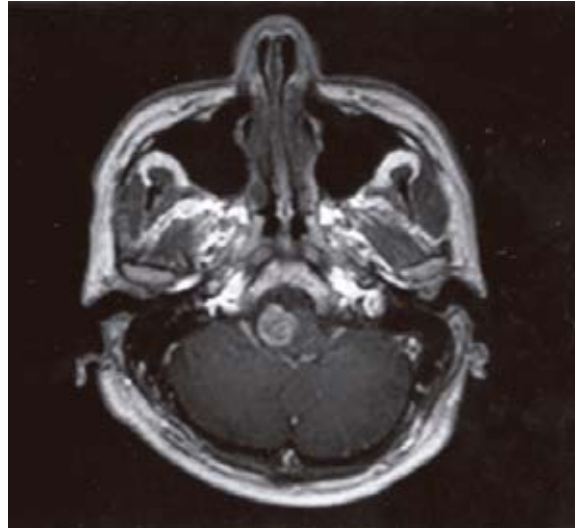


Fig. 1b Enhanced axial T1-W MR image shows heterogeneous enhancement of the aneurysm.

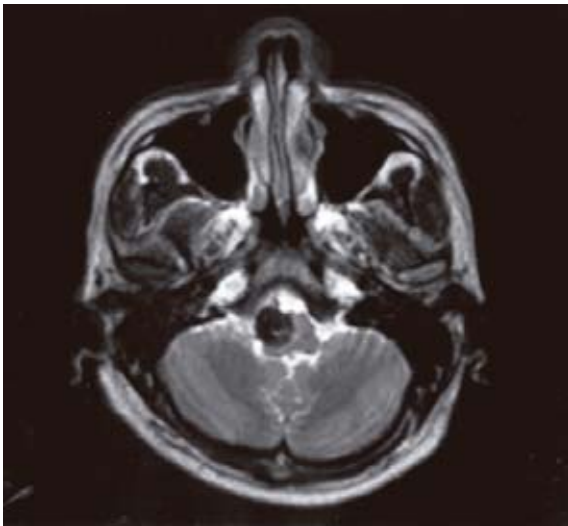


Fig. 1c Axial T2-W MR image shows hypointensity of the thrombosed aneurysm. There is a compressive effect upon the adjacent brainstem without associated parenchymal oedema.



Fig. 1d Digital subtraction angiogram (anteroposterior projection) shows a small-calibre right vertebral artery terminating into smaller branches at the region of the foramen magnum, around a rounded mass. This eventually reformed into the right PICA .

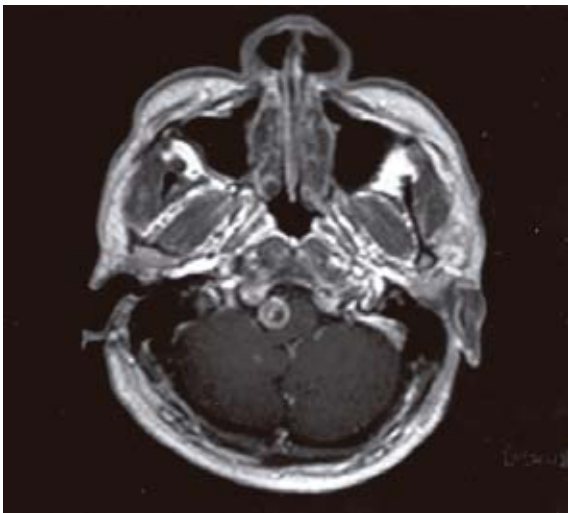


Fig. 2a Enhanced axial T1-W MR image taken at one month shows decrease in size of the aneurysm but with persistent enhancement.

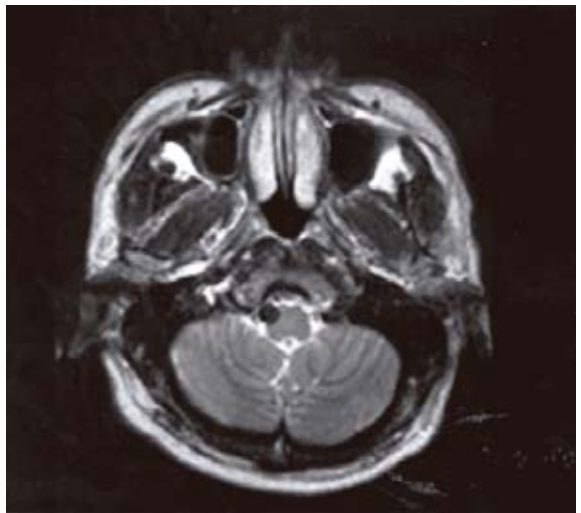


Fig. 2b Axial T2-W MR image taken at six months shows significant decrease in size of the aneurysm.

improvement in clinical symptoms, the patient declined surgery and was managed conservatively. The patient remained clinically well and asymptomatic at two years follow-up.

DISCUSSION

Contrast enhancement of a thrombosed intracranial aneurysm on MR imaging is uncommon, and can result in both diagnostic and management difficulties. The major differential concern, as in our case, is the exclusion of a highly malignant vascular wall tumour such as angiosarcoma, and the differentiation from benign lesions such as intravascular papillary endothelial hyperplasia (IPEH) and giant serpentine intracranial aneurysm. Appreciation of the imaging characteristics of each of these lesions aids the diagnosis and patient management.

In our case, the short duration of symptoms in a previously-well and healthy patient, with no other imaging abnormality other than an enhancing thrombosed aneurysm, suggested the rapid development of an endothelial proliferative process. Both intravascular papillary endothelial hyperplasia (IPEH) and malignant angiosarcoma were considerations. Follow-up imaging at one and six months showed persistence of enhancement with reduction in size of the aneurysm indicating regression, thus favouring IPEH. Clinically, as there was complete resolution of symptoms supported by improvement on imaging, the patient declined surgery and was managed conservatively.

Intravascular papillary endothelial hyperplasia (IPEH), or otherwise known as Masson tumour, is a benign condition resulting from atypical proliferation of endothelium. It can occur in the setting of an intracranial aneurysm with an organising thrombus⁽¹⁻⁴⁾. It is clinically important as it can present as an enhancing mass on MR imaging, simulating a tumour. First described pathologically by Masson in 1923 in a haemorrhoidal vein⁽⁴⁾, similar lesions have since been described elsewhere in the body, most commonly in thrombosed subcutaneous blood vessels. Common locations include the fingers, scalp, neck, and trunk, where these lesions appear as reddish blue, firm, small superficial masses. Central nervous system occurrence is rare⁽⁶⁻¹¹⁾. It can develop within a pre-existing vascular malformation or thrombus, with reported locations in the meninges, cavernous sinus/sellar, torcula, and superior orbital fissure⁽⁷⁻¹¹⁾. Lesley et al⁽³⁾ reported a case of IPEH occurring at a thrombosed posteroinferior cerebellar artery aneurysm.

Endothelial proliferation from chronic inflammation and irritation is believed to be causative, although the exact aetiology is unclear. The original description by Masson considered it a true neoplasm, with intravascular

cells proliferation from the vein walls⁽⁴⁾. Many other hypotheses have since been proposed. Salyer and Salyer considered it to be an atypical manifestation of an organising thrombus, a pseudotumoral lesion, and called it intravascular angiomatosis⁽⁵⁾. Clearkin and Enzinger supported the theory but suggested the term, intravascular papillary endothelial hyperplasia (IPEH)⁽²⁾. A thrombus is believed to serve as a matrix for the development of the endothelial proliferative fronds, hence the association^(4,12). In some cases where no thrombus is found, primary endothelial proliferation with secondary thrombus formation, or simply persistence of endothelial hyperplasia after disappearance of thrombus, has been postulated⁽⁴⁾.

When IPEH develops, endothelial proliferation results in continuing growth and enlargement of the aneurysm. Such enlargement can occur within a relatively-short time frame of 29 months for the development of symptomatic IPEH in a previously-normal patient⁽³⁾. Follow-up imaging showed a gradual decrease in size of the aneurysm in our case, which we believe, documents spontaneous regression of IPEH in a patient that was treated conservatively. IPEH is generally hypervascular and will show enhancement on imaging. On computed tomography (CT), the lesion is generally slightly hyperdense pre-contrast, with variable enhancement. On MR imaging, T2-weighted images generally show signal hyperintensity within the lesion, and show hypointensity to isointensity on non-enhanced T1-weighted images. Avid enhancement is noted post-intravenous administration of gadolinium-DTPA. Angiographically, significant variability has been reported⁽⁶⁻¹⁰⁾, but a vascular blush is expected⁽³⁾.

Angiosarcomas are exceedingly rare malignancies, comprising less than 1% of all sarcomas. They arise from endothelial cells of arteries, veins or lymphatic channels, and are located mainly in the head and face, liver, skin, and other soft tissues. Primary brain angiosarcomas invariably show rapid onset of symptoms, and are extremely vascular and aggressive. They often extend beyond the confines of the vessel lumen, with invasion and a tendency towards haemorrhage. Such features were not seen in our case. Microscopically, they form irregular vascular spaces and can be confused with IPEH. Many authors have reported histological analogies between IPEH and low-grade angiosarcoma^(4,12). It is the lack of cellular atypia, mitoses and necrosis that allow an angiosarcoma to be excluded^(10,13). Distinguishing features of IPEH are that it is frequently well-circumscribed or encapsulated, being limited entirely by the vascular wall and is characterised by papillary fronds, which either appear as bundles or as two endothelial cells layers^(4,12). In angiosarcoma, proliferative fronds are also present, protruding into the vascular lumina similar to

those found in IPEH, but display aggressive features, invariably extending beyond the vascular lumen with haemorrhage.

Giant serpentine intracranial aneurysm is another entity that may present with enhancement in a partially-thromosed intracranial aneurysm. The term was first introduced by Segal and McLaurin in 1977 to describe a partially-thromosed aneurysm containing tortuous vascular channels internally with a separate entrance and outflow pathway⁽¹⁴⁾. They belong to a subgroup of giant intracranial aneurysms, and have distinct clinical presentation and radiological features on CT, MR imaging and angiography⁽¹⁵⁾. It can be mistaken for neoplasm since its presentation is often as an intracranial mass, with adjacent oedema and mass effect, in a patient with progressive neurological deficits. Depending on the location of the aneurysm, signs and symptoms include headache, hemiplegia and hemiparesis, visual disturbance, cranial nerve palsy, dysphasia and aphasia, nausea, vomiting, seizure and vertigo. On pre-contrast CT, an oval-shaped mass of mixed density, with areas of increased attenuation representing the thrombus, and tubular regions of decreased attenuation representing the patent vascular channel, is characteristic. After contrast administration, there is enhancement of the serpentine vascular channel^(16,17). The MR imaging findings consist of a mass lesion with a heterogeneous signal representing areas of haemoglobin degradation and flow voids, and the vascular channel may be demonstrable on MR angiography. Conventional angiography is a diagnostic and useful test in evaluating the location and the state of flow in a giant serpentine aneurysm⁽¹⁵⁾.

The management approach differs for each of the conditions discussed. In general, the management of IPEH occurring anywhere in the body is local surgical excision, with histological examination for a definitive diagnosis. They generally have good prognosis but there are cases of IPEH recurring locally, should the excision be incomplete^(2,6,18). Adjuvant radiotherapy or chemotherapy has also been offered for incompletely resected or multiple intracranial IPEH^(6,8,11). For angiosarcoma, aggressive surgical removal has been

recommended as the treatment of choice, however, with risk of possible massive blood loss during surgery. The treatment of giant serpentine aneurysms is largely by endovascular means aimed at arresting growth and mass effect through obliteration of the abnormal vascular channel⁽¹⁵⁾.

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