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PICTORIAL REVIEW

Isolated intraventricular haemorrhage in adults

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ABSTRACT

While intraventricular haemorrhage is frequently found in association with intraparenchymal or subarachnoid haemorrhage, isolated intraventricular haemorrhage (iIVH) is rare in adults and seldom described. Awareness of possible causes is important in order to guide patient management. After elimination of a traumatic cause, numerous aetiologies remain possible. The most frequently found underlying lesions are arteriovenous malformations and aneurysms, but other vascular causes should also be sought, including cavernous malformations and moyamoya disease. Arterial hypertension, anticoagulant use, coagulopathies and certain toxic substances are also associated with iIVH. Finally, iIVH may be caused by intraventricular tumours. In a high number of cases, the cause remains unknown. Vascular and non-vascular causes should be searched through an imaging work-up (with CT angiography, MRI and catheter angiography when necessary) and correlation with clinical information to yield a diagnosis. The aim of this pictorial essay was to review the aetiologies of iIVH in adults.

INTRODUCTION

Intraventricular haemorrhage (IVH) is often found as a complication of intraparenchymal haematomas or in association with subarachnoid haemorrhage (SAH). Historically, only discovered at autopsy, IVH was considered to present with sudden coma and death. With the advent of CT imaging, IVH has become more commonly diagnosed in association with non-specific symptoms (headache, nausea, altered mental status, focal neurological deficits), often similar to those of SAH. It may even be an incidental radiological finding.

In infants, IVH is a frequent complication of prematurity and most often results from a germinal matrix haemorrhage, while truly isolated intraventricular haemorrhage (iIVH) is less common even in newborns. In adults, iIVH is rare, accounting for $\leq 3\%$ of intracerebral haemorrhages.¹

The purpose of the present review was to illustrate the aetiologies of adult iIVH, *i.e.* blood in the ventricular system only. Awareness of these entities (Table 1) is important to ensure the proper diagnostic work-up of this rare type of intracranial haemorrhage.

AETIOLOGIES

Vascular pathologies

The lesions most frequently detected as underlying iIVH are of vascular origin.^{2,3}

Arteriovenous malformations

Arteriovenous malformations (AVMs) can lead to haemorrhage restricted to the ventricular system and have been identified as causes of iIVH in adults in up to 20–25% of cases.^{1,3}

Intraventricular AVMs (of the choroid plexus) are often small and may be angiographically occult.⁴ They can cause iIVH (Figure 1), but may present with large haemorrhages extending into the adjacent cerebral parenchyma. Extraventricular AVMs may lead to iIVH by contact of the nidus with a ventricle or by angioarchitectural weak points such as flow-related aneurysms (Figure 2), venous ectasias or deep venous drainage (Figure 3). Small and deeply located AVMs may be more prone to rupture and rebleeding.⁴

Aneurysms

Although aneurysmal rupture typically causes SAH, it may rarely result in iIVH in the case of aneurysms developed on intraventricular arterial segments or in proximity to the ventricular walls or openings. The rupture of aneurysms of the posteroinferior cerebellar artery (PICA) is often associated with IVH (up to 95%), although only in a minority of cases with iIVH (5%).⁵ The haemorrhage is usually seen in the fourth ventricle (Figure 4) but can extend supratentorially. Occasionally, it may be apparent supratentorially only.⁵ Both proximal and the less frequent distal PICA aneurysms can present with iIVH without visible SAH. Distal PICA aneurysms have been found to rupture at

Table 1. Isolated intraventricular haemorrhage: causes and predisposing factors

Vascular pathologies
AVM
Aneurysm
Cavernous malformation
Moyamoya disease (and other carotid occlusive disease)
Other vascular pathologies (dAVF, vasculitis)
Tumour
Predisposing factors
Arterial hypertension
Coagulopathy
Anticoagulation
Sympathomimetic abuse
Trauma
Surgery
Recirculation of SAH

AVM, arteriovenous malformation; dAVF, dural arteriovenous fistula; SAH, subarachnoid haemorrhage.

smaller sizes than those at other sites. Small size and proximity of the enhancing choroid plexus may render their identification difficult on CT angiography and even catheter angiography.

Aneurysms of the anterior communicating artery and of the basilar tip may directly rupture into the ventricular system through the lamina rostralis and the floor of the third ventricle, respectively. Aneurysms of the supraclinoid internal carotid,

pericallosal, anterior choroidal and lenticulostriate arteries have also been reported as causes of iIVH.

Cavernous malformations

Intraventricular or paraventricular cavernous malformations occur in 2–10% of patients with cerebral cavernous malformations,⁶ but seem to have a higher tendency to rerupture than those in other locations,⁶ causing iIVH (Figure 5) or combined IVH and parenchymal haemorrhage. Their hemosiderotic rim may be thin or absent and the core less heterogeneous, differing from the classical MRI aspect.⁶

Moyamoya

Moyamoya disease, characterized by progressive carotid occlusion and the development of collateral vascular networks, commonly presents with haemorrhage in adults, the most frequent site being the ventricular system.⁷ Arterial fragility within the choroidal collateral system has been implicated as a cause.⁷ The choroidal collateral system is more developed in adults, as opposed to the basal network and collaterals from the external carotid artery (which are more prominent in children); the increased haemodynamic load on the dilated choroidal and medullary arteries in the ventricles and paraventricular region may lead to rupture of these vessels and iIVH (Figure 6). IVH may also result from the rupture of lenticulostriate and choroidal artery aneurysms.⁷

Moyamoya syndrome (moyamoya pattern from a known associated condition) and other carotid occlusive diseases may result in similar changes and IVH.

Other vascular causes

Dural arteriovenous fistulas typically present with parenchymal haematoma or SAH but have been reported to cause iIVH. Isolated cases of vasculitis complicated by iIVH have been

Figure 1. Choroidal arteriovenous malformation (AVM): (a) axial brain CT is showing intraventricular haemorrhage in the lateral ventricles. (b) Asymmetry of the choroid plexus vessels on axial CT angiography maximum intensity projection reformat, with prominence on the right (white arrow). (c) Vertebral digital subtraction angiography lateral projection is demonstrating a small AVM supplied by a dilated lateral posterior choroidal artery (black arrow) with an early draining vein (asterisk) towards the internal cerebral vein.

Figure 2. An arteriovenous malformation (AVM) with nidal aneurysm: (a) axial brain CT is showing intraventricular haemorrhage in the lateral ventricles. (b) The left post-central tangle of flow voids on T_2 weighted imaging is corresponding to an AVM nidus. (c) Left internal carotid artery digital subtraction angiography is showing the wedge-shaped AVM with a small aneurysm extending into the left lateral ventricle (arrow).

reported. Although a haemorrhagic choroid plexus infarction could lead to iIVH, evidence of cases is scarce in the literature.

Tumours

IVH can correspond to intralesional or extralesional haemorrhage of intraventricular or paraventricular tumours (Figures 7 and 8) and may obscure the presence of an underlying mass lesion on initial imaging. Haemorrhage may result from pathologic tumour vessels or the effect the tumour exerts on adjacent structures. It has been reported with primary and secondary intracranial lesions, including choroid plexus

papilloma, neurocytoma, subependymoma and meningioma, the latter of which has been noted to have a higher bleeding risk when in an intraventricular location. Among secondary lesions, choroid plexus metastases from thyroid and renal cell carcinomas have most frequently been reported as causes of iIVH.

Predisposing factors

While arterial hypertension is an established cause of parenchymal haemorrhage and secondary IVH, it is not clear whether it causes iIVH by the same mechanisms. Noted as the most

Figure 3. An arteriovenous malformation (AVM) with venous ectasia: (a) axial brain CT is showing intraventricular haemorrhage in the lateral and third ventricles. (b) CT angiography axial maximum intensity projection reformat and (c) lateral view volume rendering technique reformat are demonstrating an AVM nidus with deep venous drainage and intraventricular ectasias (arrows) of the draining veins.

Figure 4. Aneurysms associated with intraventricular haemorrhage (IVH): (a) sagittal CT angiography (CTA) maximum intensity projection (MIP) reformat is showing a distal posteroinferior cerebellar artery (PICA) aneurysm projecting into the fourth ventricle (white arrow) and fourth ventricular blood (asterisk). (b) Right vertebral digital subtraction angiography lateral projection is confirming the choroidal PICA aneurysm. (c) CTA sagittal reformat is showing a basilar tip aneurysm deforming the floor of the third ventricle (white arrow). (d) Anterior communicating aneurysm is shown to be touching the lamina rostralis on a sagittal CTA reformat (white arrow). (e) Axial brain CT is showing IVH in the lateral ventricles and a giant aneurysm of the left internal carotid artery (ICA) ophthalmic segment with intramural haemorrhage. (f) CTA sagittal MIP reformat is showing the patent lumen to be remote from the haemorrhage, whose origin was supposed to be the rupture of vasa vasorum in direct proximity to the adjacent ventricle.

frequent risk factor or cause of iIVH in some studies,^{2,3,8} it is considered a risk rather than a definite causative factor of iIVH.² It may originate from the choroid plexus or thalamic and

caudate nucleus haematomas. The latter should be searched ("false" iIVH) (Figure 9) but may be too small to be visible² or differentiated from IVH on CT or MRI. In some cases, it might

Figure 5. Cavernous malformation: (a) a mixed density lesion in the right frontal horn on axial CT. (b) T_2 weighted imaging is demonstrating an irregularly shaped hyperintense/isointense lesion with a discontinuous hypointense rim (cavernous malformation) (arrow) and adjacent predominantly hypointense intraventricular haemorrhage.

Figure 6. Moyamoya disease: (a) axial CT is showing intraventricular haemorrhage in the lateral ventricles. (b) CT angiography volume rendering technique reformat, superior view: occlusion of the supraclinoid internal carotid arteries (ICAs) and proximal M1 and A1 segments with hazy basal collaterals and dilatation of the choroidal and thalamoperforating arteries. (c, d) Right internal carotid digital subtraction angiography frontal and lateral projections are confirming the moyamoya pattern with marked dilatation of the choroidal arteries (asterisks).

be difficult to determine on imaging whether the haemorrhage is indeed restricted to the ventricles.

Other reported systemic predisposing or causal factors are coagulopathies⁸ and anticoagulation therapy or antiplatelet agents,³ possibly in association with underlying structural lesions. Sympathomimetic abuse has been reported in association with IVH and iIVH. The pathophysiologic mechanism has not been clarified, and a proportion is possibly related to pre-existing underlying lesions such as AVMs or aneurysms.

Locally, infections of the choroid plexus may be complicated by haemorrhage, as well as the rupture of infectious aneurysms.

Trauma and surgery

Traumatic IVH is uncommon and isolated in a minority of cases (<5% in one study),⁹ although it may be more frequently diagnosed since the advent of higher resolution imaging. It has been postulated to result from the rupture of the choroid plexus or tela choroidea vessels or of vessels in the ventricular walls, resulting from pressure changes induced by a head impact.⁹ In

the majority of cases, it is found in the lateral ventricles, but it may concern all ventricles or the third or fourth ventricles in isolation.⁹ Associated findings such as extracranial soft-tissue swelling, cranial fractures or more distant traumatic lesions can reinforce the diagnosis (Figure 10a). IVH that is isolated on CT may be found to be associated with other lesions such as small contusions or diffuse axonal injury on MRI (Figure 10c). Patient outcome is generally determined by associated injuries.

Apart from surgery directly implicating the ventricles (such as drain or shunt insertion and removal), spinal interventions (including lumbar puncture) can lead to apparent iIVH. This is also true for recirculation of blood from cranial or spinal SAH of other origin (e.g. spinal dural arteriovenous fistulas).

Unknown cause

In a considerable proportion of cases (25–45%), the aetiology of iIVH is unknown.^{3,8} The number of cases in which no aetiology can be determined varies among series and according to the intensity of work-up.³ It has been suggested that some cases of IVH of unknown origin may be due to haemorrhage from small

Figure 7. Intraventricular metastasis: (a) the axial gradient echo (GRE) image is showing a small left occipital intraventricular haemorrhage (arrow). (b) Axial T_2 weighted imaging and (c) T_1 weighted imaging (T1WI) are showing an isointense mass involving the left lateral ventricle (arrows). (d) T1WI with contrast is demonstrating homogeneous enhancement of the lesion (arrow), corresponding to a metastasis of a diffuse large B-cell lymphoma with an extracranial primary on systemic work-up.

vascular malformations which are not visible on catheter angiography but only microscopically,² and which are possibly compressed by the IVH or destroyed upon haemorrhage; cases

of microscopic malformations have been described in autopsy studies, as well as cases of lesions visible on repeat but not initial catheter angiography.

Figure 8. Ependymoma: (a) axial CT is showing a fourth ventricular hyperdensity. (b) Axial T_2 weighted imaging and (c) T_1 weighted imaging (T1WI) are showing a fourth ventricular mass lesion with intrinsic blood products, demonstrating enhancement on (d) axial T1WI with contrast.

Figure 9. Arterial hypertension: (a) intraventricular haemorrhage (IVH) ascribed to hypertension—the right lateral ventricle has isolated intraventricular haemorrhage and there are extensive white matter hypodensities on axial CT. The patient was known for arterial hypertension; work-up was negative for other causes. (b) Axial CT in a different patient is showing secondary IVH from a hypertensive thalamic haematoma (asterisk), the more common scenario.

Small subependymal (parenchymal) haemorrhages indiscernible on imaging have also been postulated as a possible explanation for IVH of unknown origin.¹⁰ It may be difficult to distinguish

between a primary IVH with dissection into the adjacent brain parenchyma and a parenchymal haemorrhage with secondary IVH even microscopically.¹⁰ Alternatively, choroidal arteries may

Figure 10. Traumatic intraventricular haemorrhage (IVH): (a) axial CT is showing a left occipital horn blood level (arrow) and right periorbital swelling. (b) Sagittal CT reformat in a different patient is showing a non-dependent IVH in the anterior portion of the third ventricle (arrow), which resolved on follow-up imaging. (c) Axial CT in a different patient is showing small occipital horn blood levels (arrows). (d) Axial susceptibility-weighted imaging in the same patient is demonstrating IVH not to be isolated: Additional right mesencephalic and left temporal haemorrhagic foci (arrows) are related to Grade III diffuse axonal injury.

be at the origin of (hypertensive) haemorrhages directly into the ventricles.²

Differential diagnoses of IVH include intraventricular pus, colloid cysts and hyperdense tumoral lesions (e.g. lymphoma).

With the ascent of high-resolution CT and the more widespread availability of MRI, iIVH may now be more frequently diagnosed in settings differing from its “classical” presentation, with only small quantities of blood in some cases. Awareness of

possible aetiologies is essential for a proper assessment of this finding in the individual context.

CONCLUSION

A variety of pathologies can be responsible for isolated IVH. Upon identification of IVH, CT/CT angiography, MRI/MR angiography and catheter angiography can provide complementary information for a confident evaluation and a potential treatment access.

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