Introduction

Intracranial cavernous haemangioma (CH) was an uncommon pathology before the introduction of MRI examination. It was regarded as a mysterious pathology in neurological diagnosis because the lesion could not be easily revealed by cerebral angiogram or CT scan. It was therefore given the description of angiographically occult vascular malformation or cryptic vascular malformation.

When MRI became a common screen examination, CH became a common pathology identified in a routine MRI examination of the brain.

Pathology

CH consists of a group of thin walled vessels as discrete, lobular and well circumscribed lesions inside the parenchyma of the brain. Grossly it appears as a raspberry-like lesion red to purple in colour (Figure 1).

Microscopically it consists of dilated thin walled capillaries with variable thin fibrous adventitia devoid of smooth muscle and elastin. There is no brain parenchyma between the vascular channels. Haemosiderin deposits are always present inside the surrounding normal parenchyma indicating that diapedesis of red blood cells is a common event in all CHs.

Diagnosis

Clinical diagnosis of intracranial CH is impossible. Skull x-rays may occasionally reveal fine calcifications at the lesion but the finding is not diagnostic.

CT scan may be totally normal in most CHs. Hyperintense signals may be seen if old blood is accumulating. Enhancement after contrast CT scan is variable and is usually not diagnostic.

CH is well known to be invisible in cerebral angiograms and is therefore called angiographically occult. Occasionally venous pooling at the lesion may be seen and is again not diagnostic.

MRI is the most important imaging study for confirmation of the diagnosis (figure 2). In the absence of overt haemorrhage, most CHs appear as a hyperintense lesion with a faint hypointense rim in T1 and lobulated heterogeneous signals with a hypointense rim in T2. The most diagnostic image appears in the gradient-echo MRI which produces a blooming artifact from the magnetic susceptibility effect of haemosiderin. CH therefore appears characteristically as a dense hypointense signal in the gradient-echo MRI.

Epidemiology

CH of the brain is found in 0.1% to 4% of all vascular malformations of the brain. In 4,068 cases of prospective autopsy study, McCormick identified CH in 0.4%, arteriovenous malformations in 0.6%, telangiectases in 0.7% and venous malformations in 2.6%.

There is no sex prevalence and most symptomatic cases are found between 20 to 40 years old.

About 75% of the lesions are located in the supratentorial region (1/4 in the frontal and 1/6 in the temporal lobes) and 25% in the infratentorial region (50% in the pons or brainstem).
Multiple lesions are reported in 8 to 18% of cases (figure 3). Multiple CHs are common in the south-western part of the United States among Hispanic patients. Familial incidence is reported in 50% of multiple CHs and an autosomal dominant inheritance with variable penetrance is suspected. There is no convincing hereditary link with single lesions.

Clinical Presentation

The majority of CHs are found incidentally with no clinical symptoms. It is associated with headaches less commonly than arteriovenous malformations. Two major clinical symptoms related to CH are bleeding and epilepsy.

Haemorrhage in CH

The presence of haemosiderin around the lesion is commonly found and represents diapedesis of red blood cells through the thin walled vessels. This results in surrounding gliosis with a fibrotic layer covering the lesion. Minor intralesional bleeding and thrombosis are also commonly found in incidental cases. All these findings should not be considered as overt haemorrhage.

Robinson defined overt haemorrhage as acute or subacute blood accumulation outside the haemosiderin ring of the lesion (Figure 4). With this definition, the incidence of overt haemorrhage in virgin cases is estimated at 0.7% per lesion per year. However the risk of rebleeding after an overt haemorrhage is 25% in one year without treatment. Rebleeding after the first haemorrhage is often related to physical exertion. Incidents of overt haemorrhage are more common in females. A higher chance of haemorrhage during pregnancy has been suggested. The consequence of overt haemorrhage is seldom fatal. The presentation depends on the location of the lesion.

Overt haemorrhage in supratentorial lesions is commonly associated with progressive hemiparesis and is often misdiagnosed as tumours in initial CT scan. Occasionally extensive intraventricular haemorrhage is found in periventricular lesions (figure 5).

Bleeding in the brainstem from CH is more serious and sixth nerve palsy with diplopia is a common initial presentation followed by multiple cranial nerve palsy, ataxia and long tract signs. Coma and life threatening brainstem insult can occur as a consequence of recurrent bleeding.

The chance of haemorrhage is independent of the lesion size. However bleeding from a large CH often results in significant neurological impairment that needs surgical treatment.

Epilepsy in CH

The actual incidence of epilepsy in patients with supratentorial CH is unknown. More incidents of epilepsy should be found in patients with CH at the hippocampus and motor area (figure 5). The abnormal vessels themselves are not epileptogenic. The surrounding tissue is rendered epileptogenic with the effect of pressure and trophic factors such as haemosiderin.

Treatment

Most CHs are diagnosed as incidental findings in MRI during investigation for other problems. As the risk of spontaneous bleeding is low and seldom catastrophic, treatment is not required if there is no clinical...
symptoms. However a detailed history is often required to exclude the presence of complex partial seizure which is often ignored by patients.

**Conservative Treatment**

Conservative treatment for overt bleeding in CH is often successful if neurological impairment is not significant. If rebleeding is minimised by the reduction of physical activities and tranexamic acid, neurological improvement is usually seen in a few weeks to a few months (figure 7). The patient is advised to withhold all activities that will increase venous pressure in the subsequent year.

**Surgical Treatment**

Surgical removal is the only effective method for elimination of a CH. It is not a difficult procedure event in the brainstem. The existing gliosis around the CH serves as a good layer for microscopic dissection of the lesion without major damage to normal tissues. The use of navigator and microscopy allows safe removal of deep seated lesions.

However the indication for surgery must be justified. The common indication for surgical removal is a significant neurological impairment as a result of repeated haemorrhages resulting in a mass effect.

**Epilepsy Treatment**

CH is commonly found in investigations for poorly controlled epilepsy, especially in the hippocampus. Surgical removal of the CH for control of epilepsy is suggested if the epileptic focus is confirmed by electrophysiology studies. Good surgical result in the control of epilepsy has been reported at 70% complete seizure free after operation.\(^1,3\)

Removal is usually performed with clearance of the surrounding haemosiderin-loaded brain parenchyma in order to remove the epileptogenic zone. However there is a controversy in this point because no significant difference is found between surgical removal of the lesion alone and removal of surround haemosiderin.

**Radiosurgery**

The use of radiosurgery such as the Gamma Knife for treatment of CH remains debatable. There is a lack of convincing evidence in the literature to support the effectiveness of radiosurgery for the elimination of CH. The benefit of such therapy must be carefully evaluated against the potential radiation toxicity.\(^1,3\)

**Summary**

Cavernous haemangioma of brain is a common incidental finding in MRI examinations. The associated clinical events are haemorrhage and epilepsy. The risk of overt haemorrhage is 0.7% per lesion per year for virgin cases. Active treatment is usually not required for asymptomatic cases. Surgical removal is the only confirmed effective method for elimination of the CH and is indicated for progressive neurological impairment from repeated bleeding and uncontrolled epilepsy.

**References**