RECURRENT INTRACRANIAL HAEMORRHAGE DUE TO CRYPTIC ARTERIOVENOUS MALFORMATION (A Case Report)

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Introduction

Arteriovenous malformation (AVM) is an uncommon cause of intracerebral haemorrhage (ICH). It is usually suspected in a patient who is young and normotensive. The diagnosis is usually confirmed by angiographic studies. However, these studies may be normal due to various reasons. ICH recurs unpredictably in patients with AVM and is an important cause of mortality and morbidity. We describe a case of recurrent ICH due to AVM in whom cerebral angiography was normal and diagnosis could be confirmed only on autopsy.

CASE REPORT

A 40-year-old male first presented with a generalized tonic-clonic seizure and prolonged postictal unconsciousness of 4 hours duration in August 1994. On regaining consciousness, he was found to have right-sided hemiparesis. He recovered completely in 2 weeks. In October 94, he had a right focal motor seizure with secondary generalization followed again by right-sided hemiparesis. CT scan of the brain done at that time showed ICH in the left fronto-parietal region with surrounding oedema and mass effect (Fig 1). While recovering from hemiparesis, he had another episode of right focal motor seizure and loss of consciousness in December 94. He denied history of headache, vomiting, or visual disturbances in the past. There was no history of hypertension, diabetes mellitus, bleeding disorders, connective tissue diseases, anticoagulant therapy or drug abuse. No history of similar ailment in other family members was present.

On admission in December 94, he was fully conscious and normotensive. There were no neurocutaneous markers. He had motor aphasia and right hemiparesis. Optic

fundi were normal. Other systemic examination was normal. Blood haemoglobin, total leukocyte count and differential counts were normal. Bleeding time was 2 min 15 sec, clotting time 4 min 30 sec, prothrombin time control 11 sec and test 13 sec, partial thromboplastin time with kaolin control 45 and test 43 sec, platelet count was 1,60,000/cumm. Blood for VDRL and HIV was negative. He was managed with cerebral decongestants and anti-convulsants. Considering a possibility of AVM, a left-sided carotid angiography was performed after 6 weeks. It did not show any vascular malformation or abnormal vasculature. Four days after angiography, the patient had another seizure. Following this he became deeply unconscious, developed brain-stem herniation and died after 10 hours.

Post-mortem examination showed a haematoma measuring 5x4 cm and subarachnoid haemorrhage in the left parietal lobe extending to frontal and occipital lobes with evidence of brain-stem herniation. No AVM or aneurysm was discernible on gross examination. Histopathological examination of brain showed numerous dilated and tortuous intercommunicating vessels in the

Fig. 1 : CT scan of the brain showing an area of high attenuation due to haemorrhage in the right parietal lobe.

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Fig. 2 : Section of the brain showing jumbled mass of arteries and veins with evidence of rupture of internal elastic lamina in some arteries (EVGx40).

left parietal lobe (Figs 2 & 3). There was disruption of internal elastic lamina and hyalinization of the vessel wall of some arterioles. There was evidence of gliosis. A few vessels showed evidence of thrombosis and haemosiderin laden macrophages were seen in the surrounding tissue.

Discussion

AVM of the brain is a vascular lesion of congenital origin representing persistence of embryonic pattern of blood vessels which are dilated and tortuous [1]. Though present at birth, AVMs grow in size with age and become symptomatic during the second or third decade [2]. Aneurysms and vascular malformations account for nearly 5% of ICH [3]. With better control of hypertension in the population and improved diagnosis of AVM, the latter is now emerging as a more important cause of ICH.

Arteriography establishes the diagnosis with certainty and demonstrates AVMs larger than 5 mm in diameter. Crawford and Russel, introduced the term "cryptic" to describe small vascular malformations which are not evident on angiography [4]. In about half of the patients, AVM first presents as intracerebral or subarachnoid haemorrhage. In 30% of the cases, a seizure is the first and only manifestation. Focal neurologic deficit, audible bruit or cardiac failure are uncommon manifestations [1]. The natural history and bleeding characteristics have been studied by many workers. Forster et al found a 25% risk

Fig. 3 : Section of the brain showing two arteries of different sizes and one vein in the sub-arachnoid space (EVG-40).

of rebleed in the 4 years after the first haemorrhage, whereas in patients who had 2 episodes of bleeding, the risk was 25% in 1 year [5]. Graf et al estimated the risk of recurrent ICH to be 6% in first year and about 2% per year thereafter [6]. Prognosis in AVM bleeding is considered better than that of bleeding due to aneurysm [7]. However, our patients had 4 episodes of ICH over a period of 6 months which is rather unusual. The cryptic nature of AVM in this case could be either due to its small size, compression by haematoma, or thrombosis of AVM [8]. Small AVMs are more likely to bleed than large ones due to significantly high arterial and draining venous pressure [9]. Enhanced CT scan of brain may also demonstrate 95% of true AVM particularly when calcified or when a small haemorrhage has taken place. However, findings may not be specific. In evaluation of patients with ICH, angiography may not reveal any cause in 12% cases. In 17% of such cases MRI may aid diagnosis [10].

Histopathologically, AVMs may either be true AVMs, venous angiomas, cavernous angiomas, telangiectasia or varices [1]. Some authors describe venous angioma, cavernous angioma and telangiectasia under one group broadly designated as cavernous angiomas due to their common characteristics like causing small and clinically silent ICH [2]. More than 50% of them are also cryptic [11]. True AVMs are usually large and are rarely

cryptic [2]. Dilated and tortuous arteries as well as veins with normal intervening brain parenchyma were seen in our patient which are the features of a true AVM.

We report an uncommon case of histopathologically proven true cryptic AVM. The presence of gliosis and hyalinization of vascular walls indicated an old lesion. Histopathological evidence of old haemorrhage, in the form of haemosiderin laden macrophages, was also present. The patient had recurrent ICH and died within 6 months of initial manifestation.

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