

Intramedullary cervical cavernous angioma

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Abstract

A case of intramedullary cervical cavernous angioma is presented.

The patient was a 68-year-old man with sensory disturbance as initial symptom, followed by a progressive neurological deficit. Magnetic resonance imaging demonstrated the characteristic pattern of a cavernous angioma. C3-C6 laminectomy, mielotomy, and total removal of the lesion were performed. Transient postoperative deterioration was followed by a good functional recovery. Total excision of these lesions should be considered early, before lesions enlargement and recurrent hemorrhage may occur.

KEY WORDS: Cavernous angioma, Carvernoma, Spinal cord, Intramedullary, Complete excision.

Introduction

Cavernous angioma, or cavernoma, represents a type of vascular malformation of the central nervous system, together with arteriovenous malformations (AVM's), venous malformations and capillary telangiectasias^{22,31}.

High definition CT scan^{1,40}, and now high field MRI^{2,3,29}, has led to increasing numbers of these lesions being reported.

They remain rare lesions, particularly in the spinal cord^{2-4,6,9,13,19,20,23,25,26,32,38-41,44}.

The aim of this report is to present a case of an intramedullary cervical cord cavernous angioma which has been radically removed, with good surgical results, and to contribute to a better understanding of the natural history of these unusual lesions.

Case Report

A 68-year-old man presented with a one and a half year history of progressive left elbow pain and left hand and finger numbness, accompanied by forearms and hands atrophy, gait difficulty, urinary hesitancy and frequency, sexual impotence and, subjectively, cold knees.

On admission, general examination was normal. Neurologically he had an ataxic gait, a spastic tetraparesis, more marked on the right, forearms, hands and right quadriceps wasting, with brisk deep tendon reflexes, a right plantar extensor response, a left hemisensory loss to pain and temperature below D3, and a bilateral decrease in vibration sense below D6.

The clinical impression was that of an intramedullary cervical lesion.

Non contrasted cervical CT scan did not show the lesion.

MRI demonstrated a mixed signal lesion on both T1 and T2 weighted series, with minimal gadolinium enhancement, at C5 (Fig. 1).



Fig. 1—T1 and T2 weighted images, showing the intramedullary cavernous angioma at the C5 level as a low signal intensity lesion with small areas of high signal intensity, on both sequences. There is a slight expansion of the spinal cord.

C3 to C6 laminectomy was performed, without steroid administration due to previous gastrointestinal bleeding. On opening the dura, the CSF was clear and there were no abnormal vessels on the cord surface.

Following partial decompression by removal of a small amount of hemorrhagic fluid, a C5 median mielotomy revealed a dark-blue, lobulated, mulberry-like mass, which extended forwards, to be in contact with the anterior dura.

Total microscopical removal, using bipolar techniques, revealed a circumferential gliotic plane.

Postoperatively the patient was undisturbed, but twelve hours post surgery, he developed a dense tetraparesis. After CT scan exclusion of hemorrhage, intravenous steroids were introduced, and, with determined physiotherapy he then made a rapid recovery having achieved normal gait and sphincter function, with power MRC 5 in the legs and 4 in the arms, accompanied by normal tone and tendon reflexes. A C6-D12 suspended pinprick loss persisted.

Follow-up MRI indicated total removal.

Histological appearance was of a cavernoma (Fig. 2).



Fig. 2— Photomicrographic appearance of a cavernous angioma, showing large vascular spaces lined by a single layer of endothelium, mostly packed together, but with some neuronal intervening tissue. Parietal and intraluminal masses of fibrous tissue and hyaline collagen, some containing siderophages, are present, both in the vascular spaces and neuronal tissue (Verhoeff Van Giesson, x 40).

Discussion

Cavernous angioma is a relatively rare lesion ^{36,39,41,42,44}.

They represent 5% to 13% of intracranial vascular anomalies ³⁶. Most commonly described supratentorially, they have also been reported in the pineal gland ^{12,40}, IV ventricle ^{7,9,14,15,17,27,43}, cerebellum ³⁶, brainstem ^{5,7,14,16,17,24,33,35,36,40}, cranial nerves ^{21,34,37} and spinal nerves ^{11,28,42}.

They account for 3% to 16% of vascular spinal cord anomalies, which in his turn represent 6,2% to 7,5% of all intraspinal tumors ^{28,44}.

Usually located intramedullary, they may be extramedullary, either sub ²⁸ or extradural ¹⁸.

Thirty seven intramedullary cases, in 36 patients have been reported ^{2-4,6,10,13,19,20,23,25,26,32,39,40,44}. Thirteen of these were cervical ^{6,13,20,25,26,32,39,40,44}.

Together with arteriovenous malformations, venous angiomas and telangiectasias, cavernomas are classified as SNC vascular malformations ^{22,31}. Improved imaging tech-

niques, particularly MRI, are demonstrating cavernomas more frequently ^{2,6,8,18,26,29,33,44,45}.

Clinically the intramedullary cavernomas present in four clinical patterns ^{2,6,10,25,26,39,41,44}. The commonest, is discrete episodic neurological deterioration with intervening improvement. Second, slow and progressive neurological deterioration. Third, acute onset of symptoms with progressive deterioration over several days. Lastly, ictal onset of mild symptoms with chronic progressive deterioration lasting weeks to months ²⁶.

The symptoms reflect location complicated by further intramedullary or subarachnoid hemorrhage, thrombosis, and enlargement by capillary proliferation or hemorrhage inside the lesion ^{6,18,25,26,36,42,45}.

They may be accompanied by cutaneous hemangiomas, but this is a rare feature ³⁸.

Macroscopically they are well circumscribed, dark-blue, mulberry-like, vascular masses, usually with a surrounding gliotic plane ^{6,18,26,28,41,44}.

Histologically the cavernomas are characterized by large, closely apposed, irregular, sinusoidal vascular spaces, separated only by sparse collagen or fibrous tissue, usually without intervening neuronal parenchyma ^{26,33,36,45}. The presence of neuronal parenchyma does not preclude cavernoma. This could suggest a common pathology with telangiectasia ³⁰.

The vessel walls characteristically lack muscle component, and only scattered elastic elements may be found ³⁶. They are usually thin, but sometimes endothelium is packed by collagen and hyalinized connective tissue, to form thick, fibrotic vessels ^{26,36}.

Previous hemorrhage is revealed by the presence of cysts, hemosiderin-laden macrophages and cholesterol crystals ^{26,30,33,36,40,45}.

Recent or old intravascular thrombosis is common ^{26,30,36}, together with calcium deposits in the thickened vessel walls ^{26,30,33,36,39}.

The term «Hemangioma calcificans» has been used to describe a lesion, the bulk of which is composed by calcified tissue and metaplasia ^{24,36,39}.

A formal capsula as well as surrounding gliosis, may be manifest ^{26,36,45}.

With the increasing knowledge of the natural history of cavernoma, total removal should be considered in each case.

Biopsy and subtotal excision are not satisfactory methods for treating these lesions.

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