INTRAMEDULLARY CAVERNOUS MALFORMATIONS

E. CHABERT (1), X. MORANDI (2), M.P. CARNEY (1), L. RIFFAUD (2), C. LOUAIL (1),
B. CARSIN-NICOL (1)

(1) Service de Neuroradiologie,
E-mail : echabert@univ-rennes1.fr
(2) Service de Neurochirurgie, CHRU Pontchaillou, 35033 Rennes.

SUMMARY

Five cases of intramedullary cavernous malformations were retrospectively reviewed. There were 4 women and one man ranging in age from 30 to 67 years. Thoracic spinal cord was involved twice and cervical cord in three cases. Four of them underwent surgery : two improved, one remained stable and symptoms worsened in one. Clinical, radiological features and surgical management are discussed in the light of the follow-up and literature analysis. The role of T2* weighted sequence in MR diagnosis of intramedullary cavernomas is emphasised.

Key words : angioma, central nervous system, arteriovenous malformations, MR imaging, cavernoma.

INTRODUCTION

Cavernous angiomas or cavernomas are vascular malformations that may affect any tissue of the body. Within the central nervous system, the spinal cord is less frequently involved than the brain. However, with the extent of use of magnetic resonance imaging (MRI), intramedullary cavernomas (IC) occur more frequently than previously presumed. In a recent review, 117 cases were reviewed by Zegvaridis and coll. [23].

The aim of this study is to present 5 cases of symptomatic IC which four were histologically proved, to describe their clinical, MR findings, outcome and compare them to the literature.

PATIENTS AND METHODS

Between 1992 and 1998, diagnosis of IC was made in five patients hospitalized in our hospital. There were 4 women and a man ranging in age from 27 to 67 years with an average age of 43 years. Four of them underwent surgery. Three cavernomas were located in the cervical cord and two in the thoracic cord. The duration of pre-operative symptoms ranged from 8 months to 7 years. Features of relevant clinical, therapeutic and follow-up data are summarised in table I.

MR imaging was performed on a 1.5-T unit (Signa ; GE Medical systems, Milwaukee). A phased array surface coil was used, T1-weighted (TR/TE : 420/18) and T2-weighted (TR/TE : 3000/114) spin-echo images in a sagittal plane were obtained. Axial T2*-weighted images were also obtained with a gradient-echo sequence (TR/TE/FA : 500/20/20). In 3 cases, injection of gadopentate dimeglumide was injected intravenously (0.1 mmol/kg).

CASE REPORTS

Case 1

This 67-year-old woman experienced acute gait disturbance without objective sensory-motor deficit. Six months later, an acute deterioration occurred with a spastic paraparesis (strength 3/5) and bladder
TABLE I. — Clinical features and follow-up.

<table>
<thead>
<tr>
<th>Case</th>
<th>Location</th>
<th>Age/sex</th>
<th>Clinical course</th>
<th>Surgical intervention</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>T5-T6</td>
<td>67/F</td>
<td>Progressive paraparesis</td>
<td>Total removal 8 months after the onset</td>
<td>8 months: sensory loss disappearance bladder dysfunction</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Bladder dysfunction</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>C2</td>
<td>30/F</td>
<td>Post-traumatic neck pain and sensory-motor deficit of left arm</td>
<td>Total excision 1.5 year after the onset</td>
<td>3 years: stable</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Stepwise worsening of left side sensory deficit</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>T5</td>
<td>27/H</td>
<td>Spontaneous hematomyelia with immediate paraplegia and residual spasticity. Progressive sensory-motor deficit of the right leg</td>
<td>Total excision 7 years after the onset</td>
<td>1 month: post operative incomplete paraplegia</td>
</tr>
<tr>
<td>4</td>
<td>C6</td>
<td>37/F</td>
<td>Post traumatic left arm pain</td>
<td>No surgery</td>
<td>1 year: stable</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Hypaesthesia of the left side</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>C2</td>
<td>34/F</td>
<td>Acute neck pain and quadriplegia spontaneously regressive. Residual pyramidal syndrome</td>
<td>Total excision 3 years after the onset</td>
<td>6 months: residual spasticity of inferior limbs</td>
</tr>
</tbody>
</table>

**FIG. 1.** — a-b Case 1. Sagittal fast spin echo T2 (figure 1a) and axial T2* (figure 1b) weighted images demonstrate a round intramedullary mass with areas of high and low signal intensity within the lesion (figure 1a) and pronounced low intensity signal in the T2* sequence (figure 1b).
dysfunction. MRI demonstrated a well-defined multicystic intramedullary lesion at T5-T6 surrounded by a marked hypointense rim on the T2* sequence (figures 1a, 1b). No enhancement was observed after Gadolinium injection. Diagnosis of IC was made on the basis of the typical mulberry configuration of the lesion. Surgery was carried out 8 months after the onset of symptoms and disclosed a lesion consisting of multiple cystic cavities containing old blood without interposition of the normal cord. Follow-up showed a complete disappearance of sensory deficit with partial strength improvement (strength 4/5).

Case 2

The second case is a 30-years old woman. Her clinical history started after a car crash. Immediately after the trauma, she presented with left hand pain and weakness complicated few weeks later by sensory motor deficit of the left leg, examination revealed a diminution of nociception in the left from C5 to T1 and proprioception on all of left side. MRI showed a 1 cm-diameter intramedullary lesion at C2. The signal was mixed on both T1 and T2 sequences. Medullar angiogram was normal. Surgical removal performed one year after the onset of symptoms disclosed an intramedullary lesion characteristic of cavernoma on histopathological examination.

Case 3

This 27 years-old-man was first hospitalised for a spontaneous rapidly worsening paraparesis. Initial MRI detected an intramedullary high-signal-intensity lesion on both T1 and T2 weighted MR-images at T6 suspected of hematoma. A posterior laminectomy performed could not visualise the lesion. A MRI performed 5 months later was negative (T2*-sequence was not available at that time), the diagnosis of Multiple Sclerosis was then considered. Four years later the patient was re-hospitalised for a motor deficit of the left leg and hypaesthesia of the right side below T10. MRI showed a right-sided T5 nodular mass of the spinal cord with a mixed signal intensity on T1 and T2 sequences surrounded by a rim of low-signal-intensity (figures 2a, 2b). At that time, the patient refused sur-
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Case 4

The clinical course of this 37 years-old woman began with a left arm pain and numbness in C7 territory following a car accident. Hypaesthesia of the right side was demonstrated at neurological examination. Plain radiographs initially performed were normal. MRI performed 4 years after trauma showed a small T1 high-signal-intensity area at C6, slightly hypointense on the FSE T2 sequence and clearly demonstrated on the T2* sequence as a round, low-signal-intensity mass (figures 3a, 3b). Because of the hypointensity on the T2* sequence, this lesion was strongly suspected of a cavernoma. The patient refused surgery and had no complication on one year follow-up.

Case 5

This 35 years-old woman experienced spontaneous acute cervicalgia following few hours later by a quadriplegia. Within a week, she recovered normal strength with only persistent sensory deficit. A high signal intensity mass in both T1 and T2 weighted images suspected of hematomyelia was first disclosed at C2. This lesion which resolved on successive MRI allowing to depict more accurately a typical cavernoma at the site of the hematoma (figures 4a, 4b). Surgical intervention was carried out three years after the onset of symptoms and histopathological examination confirmed the diagnosis of cavernoma.

Discussion

Cavernomas exhibit the same morphological, histological and radiological characteristics whatever its location along the neuraxis. It is a well circumscribed lesion composed by juxtaposed vascular cavities filled with blood and variable-age blood products.
The distribution reported in the literature is 55 % for the thoracic cord, 40 % for the cervical cord and 5 % for the lumbar conus medullaris [23]. Three out of five patients of our series underwent MR examination of the brain which was normal. The review of literature revealed nine cases of simultaneous intracranial and spinal lesions [1, 2, 5, 12, 16, 19]. Therefore it is advisable to perform a cranial MR screening in patients harbouring a IC. If simultaneous localisation is found, it is worthwhile to perform a familial screening [5].

Clinically, IC are often disclosed after a progressive history of paraparesis and paresthesiae. Acute discovery with hematomyelia has been also documented [7]. In case of superficial masses, subarachnoid haemorrhage can be encountered [11, 18] and was not seen in our series. Ogilvy [3, 14] proposed four patterns of symptoms evolution with histopathologic correlation:

Type 1: acute episodes of stepwise neurological deterioration
Type 2: slow progression of neurological deterioration
Type 3: acute onset with rapid decline
Type 4: acute onset of mild symptoms with gradual decline

In our series, two patients presented with a type 1 evolution (cases 3 and 4), one patient with a type 3 (case 5) and two others a type 4 (cases 1 and 2).

In two cases (cases 2 and 4) clinical symptoms occurred after a trauma, which probably induced haemorrhage leading to its diagnosis. But curiously association of trauma and IC has never been reported in the literature and was seen in two out of our five patients.

**Radiological features**

MRI is the best imaging modality to explore and diagnose intramedullary cavernomas. A typical lesion on MRI is a well-circumscribed lesion with areas of high and low signal intensity within the lesion on both T1 and T2 sequences, the lesion is surrounded by a complete rim of hypointensity on T2 weighted sequence, becoming more hypointense in T2* sequence, which is proportional to the field intensity.
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Surgical management

Because of the low frequency of IC, it is difficult to predict the course of the lesion. However in all but one patient (case 4) in our series and in most of cases reported in the literature, patients experience worsening of clinical symptoms leading to surgical intervention. According to Zevgaridis [23] the average bleeding rate of IC can be estimated at 1.4 % per lesion and per year which is similar to the bleeding risk of supratentorial cavernomas [18]. However due to the size of the spinal cord a slight augmentation of mass effect caused by a bleeding may lead to dramatic worsening [18]. Analysis of surgical results demonstrate it’s benefits: 66 % improved, 28 % unchanged and 6 % further worsening [23]. This calls for surgical excision in every symptomatic IC. For asymptomatic IC, surgical intervention is more controversial since no data concerning their natural history has been published.

In one case (case 3), surgical exploration performed 3 days after the onset of the symptoms, was negative. A second operation performed six years later disclosed the lesion. It suggests that the cavernoma at the beginning must have been very small. Two other cases of first negative surgical exploration have been reported [23, 22]. These examples confirm that the bleeding risk doesn’t correlate to the size of the cavernomas. T2* sequence may be very useful to disclose small lesions. In addition, in case of small lesion, intraoperative ultrasound can be very helpful to localise the lesion [10].

CONCLUSION

The widespread availability of magnetic resonance imaging has dramatically increased the frequency of intramedullary cavernomas diagnosis. For enigmatic spinal cord lesions posing a diagnostic dilemma we suggest to include a T2* sequence in addition to the standard sequences. This is specially advantageous in the work up of small symptomatic lesions where the detection of haemosiderin increases the confidence with which a probable IC can be suggested. With atypical imaging characteristics the detection of haemosiderin on T2* sequence helps to include IC in the list of differential diagnoses.

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REFERENCES


