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Short Illustrated Review

Ependymoma of conus medullaris presenting as subarachnoid haemorrhage

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Summary

Subarachnoid haemorrhage (SAH) due to spinal ependymoma is very rare. We report a 37 year old man who presented with typical clinical signs of SAH. Lumbar puncture confirmed SAH but cerebral angiography was negative, and further diagnostic work-up revealed an ependymoma of the conus medullaris as the source of the haemorrhage. A comprehensive review of the literature was conducted. Only 17 patients with spontaneous SAH due to a spinal ependymoma have been reported since 1958. However, in cases of SAH and negative diagnostic findings for cerebral aneurysms or malformations, this aetiology should be considered and work-up of the spinal axis completed.

Keywords: Spinal ependymoma; subarachnoid haemorrhage.

Abbreviations

CT Computer tomography
SAH Subarachnoid haemorrhage

Introduction

Spontaneous subarachnoid haemorrhage (SAH) due to a spinal ependymoma is a very rare entity. Approximately less than 1% of SAH is caused by spinal lesions [1, 3, 8, 15]. Low-back pain and severe headaches are often the leading clinical features. We present a patient who had diagnostic work-up for intracranial SAH but was found to have an ependymoma of the conus medullaris as the source of haemorrhage. A thorough review of this very rare condition was performed.

Literature review and analysis

We searched the PubMed database (www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=PubMed) for "spinal ependymoma subarachnoid haemorrhage" and found 39 queries up to December 2006. Furthermore, we reviewed the references of the relevant literature. Twelve authors were found in the English and German literature, reporting about 17 patients with spinal ependymoma who presented with typical clinical features of SAH since 1958.

We reviewed the location, clinical presentation, the diagnostic evaluation and the time from onset of symptoms to diagnosis (Table 1). There were 13 males and 4 females; the median age was 31.35 years. All patients reported had histologically proven ependymomas. All tumours except one, in the lower thoracic spine, were located in the lumbar region. The most common symptoms were headaches, low-back, and leg pain. In 8 patients, headache was predominant compared to the backache. Cranial computer tomography was performed in seven patients. Of these, only one was positive and six were negative for blood. Lumbar puncture revealed bloody or xanthochromic cerebrospinal fluid in 15 patients. Cerebral panangiography was done in nine pa-

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Table 1. *Summary of cases with SAH due to spinal ependymomas*

Ref.	Patient no.	Gender	Age (y)	Location	Symptoms	Cranial CT for blood	Lumbar puncture	Cerebral panangiography	Time from onset to diagnosis
[11]	1	M	25	L 4 intradural	low-back pain, headache	not performed	xanthochromic	no bleeding source	7 months
[13]	2	M	22	L 5 intradural	low-back pain, headache	not performed	bloody	no bleeding source	3 months
[13]	3	M	74	L 1 intradural	papilloedema, vomitting, low-back pain, headache	not performed	xanthochromic	not performed	1 month
[13]	4	M	15	L 1-2 intradural	low-back pain, headache	not performed	bloody – xanthochromic	not performed	9 days
[7]	5	F	40	L 3-S 1 intradural	loss of vision, low-back pain, headache	not performed	xanthochromic	not performed	60 months
[6]	6	F	29	L 3 intradural	headache, leg pain	not performed	not obtained	no bleeding source	63 months
[6]	7	M	15	L 3 intradural	headache, low-back pain	not performed	unknown	no bleeding source	12 months
[6]	8	M	17	L 2 intradural	headache, low-back pain, sphincter distubance	not performed	bloody	no bleeding source	12 months
[14]	9	M	35	Th 9 intradural	headache, low-back pain	positive	bloody	no bleeding source	40 days
[8]	10	M	31	L 3-4 intradural	headache, photophobia, vomiting	negative	xanthochromic	no bleeding source	3 months
[12]	11	M	14	L 1-2 intradural	headache, nausea, vomiting, low-back pain	negative	bloody	no bleeding source	several months
[1]	12	M	23	L 1-2 intradural	headache, low-back pain	negative	bloody	no bleeding source	14 days
[15]	13	F	16	L 2-3 intradural	headache, vomiting, low-back pain	negative	bloody	just considered	10 months
[10]	14	F	65	L 2-3 intradural	low-back pain, headache, cauda equina, motor deficits	not performed	xanthochromic	not performed	12 months
[4]	15	M	36	L 2 intradural	low-back pain, headache, motor deficits	negative	xanthochromic	not performed	5 days
[4]	16	M	46	L 5 intradural	low-back pain, headache, motor deficits	negative	xanthochromic	not performed	10 days
[2]	17	M	30	L 2 intradural,	low-back pain, headache	not performed	bloody	not performed	unknown
Current report	18	M	37	L 1-2 intradural	headache, leg pain	negative	xanthochromic	no bleeding source	16 days

tients without any evidence of a source of bleeding. The median time from onset of symptoms to diagnosis was approximately one year (353.53 days).

Case report

A 37 year old man who complained of severe headaches, nausea, and vomiting for two days presented at an outside hospital. He also reported pain in both legs. The medical history was significant for an acute hearing loss two months previously. He received medication including, cortisone, and aspirin.

SAH (Hunt & Hess grade II) was suspected and a cranial CT scan was performed, which was negative for blood. A lumbar puncture with a three tube test, however, revealed a positive finding for xanthochromic cerebrospinal fluid.

The patient was referred to our department for further diagnosis and treatment. The patient's mental status was normal. There was slight nuchal meningism and also bilateral positive Lasègue sign. The neurological examination was otherwise normal. The day after admission a cerebral angiogram was performed, but a cerebral aneurysm or vascular malformation could not be found. Also, MRI of the cervical and thoracic spine showed no evidence of a bleeding source. Finally, MRI of the lumbar spine revealed a 3 cm intradural mass at the level of L1/2 (Fig. 1). Furthermore, multiple small lumbar and sacral masses were present. The patient underwent surgery for a suspected ependymoma. Intraoperatively, a

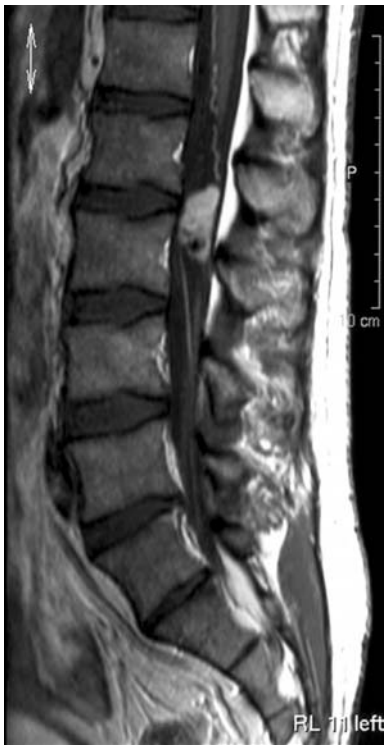

Fig. 1. MRI of the lumbar spine with ependymoma at the level of L1/2

highly vascular tumour was removed. The histological examination of the tissue confirmed the diagnosis of an ependymoma (WHO grade I). The tumour was resected without any sequelae.

Discussion

The vast majority of patients with acute onset of SAH are found to have intracranial lesions, most often aneurysms or arteriovenous malformations. In some patients SAH is caused by brain tumours, vasculitis, or secondary to infarction [17]. SAH due to spinal lesions, however, is very rare. Walton [17] analysed 312 patients with SAH and found the bleeding originating from intraspinal lesions in only 2 both of which were angiomas. Other authors report a rare incidence of SAH due to spinal origin, accounting for less than 1% of all cases [1, 3, 8, 15]. The most common aetiologies are spinal trauma, arteriovenous malformations, and aneurysms of spinal arteries [5]. Spinal cord masses like ependymomas, nerve sheath tumours, hemangioblastomas, metastasis, or meningiomas may also cause SAH. Therefore, the tumour histology seems to be an important factor for the incidence of bleeding. In relation to SAH of spinal origin the most frequent (60%) pathology is ependymoma [4].

Our patient presented with typical signs and symptoms of SAH. The diagnostic approach was according to the standard protocol, but cranial CT was negative for blood. Therefore, a lumbar puncture with the three tube test was done with a positive result. Cerebral angiography as well as cervical and thoracic MRI revealed no bleeding source. In view of persistent leg pain a lumbar MRI was performed and showed an ependymoma of the conus medullaris as the source of bleeding.

Also, the patient's past medical history revealed acute hearing loss. This may indicate preliminary bleeding from the lumbar ependymoma, since there are several descriptions of superficial cerebral siderosis caused by chronic subarachnoid haemorrhage entailing hearing loss, ataxia and other symptoms [9, 16]. However, the result of deposition of haemosiderin is a proliferation of microglia and astrocytes as well as axonal spheroid swelling. The eighth cranial nerve and the cerebellar cortex are described as particularly vulnerable to the ferritin metabolites [9, 16].

Spinal ependymoma presenting as SAH is very rare. Including the current example, only 18 patients have been reported so far. Nonetheless, this aetiology should be considered in the event of negative diagnostic findings for vascular malformations in a patient with SAH, and a work-up of the spinal axis should be completed.

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