Case Report
Spinal ependymoma presenting as subarachnoid hemorrhage and leading to superficial siderosis of the central nervous system: a case report

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Abstract: Superficial siderosis of the central nervous system is a very uncommon disorder. We report a rare case of spinal ependymoma in a 58-year-old man whose main complaints at presentation were recent lumbago and headache, and he was evaluated to confirm suspected spinal subarachnoid hemorrhage (SAH). After cerebrospinal fluid analysis and imaging studies the patient was diagnosed with SAH and superficial siderosis of the central nervous system (SSCNS). Following enhanced MRI scans and surgery the final diagnosis was spinal ependymoma with SAH and SSCNS. This is unusual case of SSCNS due to repeated small hemorrhage of a spinal ependymoma presenting as acute SAH. This case report provides a new factual basis for the pathogenesis of SSCNS.

Keywords: Subarachnoid hemorrhage, siderosis, ependymoma, magnetic resonance imaging, case report

Introduction
Superficial siderosis of the central nervous system (SSCNS) is a rare and slowly progressing disease which result by the hemosiderin deposition derived from continuous or recurrent bleeding into the subarachnoid space [1, 2]. Hemoglobin iron is deposited on the subpial layers which results in an increase of microglia and injury to nerve cells which can lead to cerebellar ataxia, sensorineural deafness, dementia, pyramidal signs, bladder disturbance, seizures, anosmia, and anisocoria [1-5]. The ruptured aneurysms, arteriovenous malformation, and traumatic injury are the main causes of SSCNS; In addition, there are more unusual sources of chronic bleeding, such as dural pathologies, tumors, and surgery [1, 4, 5].

Subarachnoid hemorrhage (SAH) is a clinical syndrome that can be caused by bleeding from an arteriovenous malformation or bleeding from the rupture of a cerebral aneurysm directly into the subarachnoid cavity [6]. SAH of spinal origin is a rare clinical entity, occurring in only 0.05%-0.6% cases [7]. It has characteristic clinical manifestations such as headaches, unconsciousness, back pain, and symptoms of nerve root irritation. The rates of missed and misdiagnosed spinal SAH are high, especially for patients showing no symptoms of spinal cord damage or patients testing positive only for meningeal irritation, both of which could be easily confused with intracranial SAH. It is reported that about 80% of spinal SAH patients had cerebral symptoms, with symptoms of headache accounting for 70% of these symptoms, and about 22% of patients showing symptoms of consciousness change [8].

Spinal ependymoma is a rare ependymoma occurring predominantly in the lumbosacral region, particularly the filum terminale. In rare cases spinal ependymoma has been found to be the cause of SAH [9-11]. We report here an unusual case of a patient with spinal ependymoma presenting as SAH which led to SSCNS.

Case report
This study was approved by Harrison International Peace Hospital. Informed consent was obtained from all individual participants included in the study.

The patient who had experienced lumbago for 7 days was accompanied by headache, nausea,
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Figure 1. Brain MRI on T2W1 demonstrated short T2 signals in the bilateral dentate nuclei, midbrain roof, and lateral fissure (A-C); SWI showed low signal in the bilateral dentate nuclei, midbrain roof, and lateral fissure. Therefore, it was considered to be the deposition of hemosiderin (D-F). There was no evidence of acute SAH found on FLAIR (G-I).

and vomiting for 4 days. Seven days previously, the lumbago occurred when the patient worked in a field, but he had no headache, nausea, vomiting, or limb movement disorder. Thus, his backache did not receive special attention. However, three days later the backache was exacerbated and headache occurred, mainly at the bilateral orbital and temporal sites. Then, the headache was gradually aggravated and was accompanied by nausea and vomiting, which led him to visit our hospital. The patient had a 20-year history of waist placeholder surgery and often suffered from lumbago. He also had a medical history of 5-6 years of walking instability and hearing loss. The results of physical examination showed that the patient was conscious and had fluent speech. However, the patient had hearing loss. His bilateral pupils were equally round and normal in size, were sensitive to light reflex, and his eyes moved freely. Horizontal and small nystagmus was visible. The bilateral frontal lines and nasolabial fold were symmetrical, and his tongue was in the center. Muscle strength and muscular tension of the limbs were normal. The bilateral tendon reflexes existed. The bilateral Babinski sign was negative, and neck resistance was suspected to be positive. Body examination revealed bilateral mild ataxia. Brain CT indicated that the intensity of the anterior median fissure was increased, and SAH could not be excluded. There was no abnormality found based on the result of laboratory tests of blood, urine, and feces. Lumbar puncture was performed and it was found that uniformly and consistently bloody cerebrospinal fluid (CSF) was discharged, and the measured pressure of CSF was greater than 300 mm H2O. Furthermore, measurements of pressure in the neck and abdomen were negative. The biochemistry of CSF was as follows: total protein more than 3358 mg/L; total number of erythrocytes 188.0×10^12/L. MRI, magnetic resonance angiography (MRA), susceptibility-weighted imaging (SWI) and magnetic resonance venography (MRV) of the brain showed: no acute SAH, widening of the longitudinal fissure, schizencephaly remove and subarachnoid space of the cerebellum with blurred edges. Therefore, the findings were considered to indicate the deposition of hemosiderin; no abnormality was found on MRA and MRV (Figure 1). Spinal MRI showed an occupying lesion in the cone region at the L2 level. MRI after contrast agent injection showed the abnormal enhanced shadow of a strip in L1-2 vertebral canals, but the boundary was clear (Figure 2A-C). The imaging diagnosis was spinal ependymoma associated to spinal SAH and SSCNS. Occupying lesions were removed by surgery. During surgery, it was found that the blood supply in the tumor was rich and closely attached to nerves. Histopathologic examination of the resected tumor was diagnostic for a World Health Organization (WHO) grade II e-
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In this case report we described a patient with lumbar ependymoma which not only led to clinically asymptomatic spinal SAH but also resulted in SSCNS. The main clinical presentation of SSCNS is including sensorineural hearing loss, pyramidal signs, cerebellar ataxia, and sensory and cognitive disturbances, seizures, headache, urinary symptoms, visual and olfactory complaints. Except above, anisocoria, lumbar backache, bilateral sciatica and neck pain, and extra-oculomotor nerve palsy, are also typically clinical feature of SSCNS [1-5].

The patient had complained of headache and back pain. Head CT showed high density in the anterior median fissure; SAH could not be excluded. The patient underwent lumbar puncture, and uniformly colored reddish CSF was discharged. The patient was then diagnosed with SAH. In order to further investigate the etiology of SAH, MRI+MRA+SWI examination of the brain was performed. The images obtained showed the low signal of T2-weighted imaging (T2WI) in the bilateral dentate nuclei of the cerebellum, longitudinal fissure, and lateral fissure, as well as the SWI signal. Hence, it was considered to indicate the deposition of hemosiderin. MRI is an effective method for diagnosis of SSCNS [2]: T2WI reveals a characteristic linear hypointensity due to hemosiderin deposition that is in contrast to the hyperintense signals of the CSF [12]. Compared with T2WI, SWI is more sensitive to hemosiderin and can more accurately detect hypointense signals in the brain [4], which provides more valuable imaging information for the diagnosis of SSCNS. Therefore, SWI and T2WI gradient echo are often used to diagnose SSCNS. This is consistent with the results we observed by MRI, and combined with the symptoms and signs of the patient, the diagnosis of SSCNS was clear. Most studies have shown that for the diagnosis of acute SAH, MRI fluid attenuation inversion recovery (FLAIR) and SWI sequences are more sensitive than CT for detecting traces of bleeding, and can be used as a supplementary means of diagnosis in CT-based examination for acute SAH [13]. Although the patient had complained of headache, nausea, vomiting, and meningismus, SWI and FLAIR did not provide the evidence for acute SAH. Head CT showing high density in the anterior median fissure indicated the deposition of hemosiderin, so the patient was confirmed to have SAH. Cerebral screening could not demonstrate the particular cause of SAH. Since our patient reported that he had back pain before the onset, we were alert to the possibility of spinal SAH. Headaches might be the initial symptom of spinal SAH, and the degree of headache could be as severe as that in intracranial SAH [8]. The diagnosis of spinal SAH is made mainly by spinal MRI and spinal angiography.

Discussion

In this case report we described a patient with lumbar ependymoma which not only led to clinically asymptomatic spinal SAH but also resulted in SSCNS.

Discussion

Figure 2. Lumbar vertebrae MRI shows occupying lesions occurred in the cone region of L2 level spinal cord, with low signal of T1 (A) and mixed T2 signal (B), while the enhanced scanning showed the abnormal enhanced shadow of a strip in L1-2 vertebrae, and the boundary was clear (red arrow) (C). Histopathologic examination of the specimen confirmed a World Health Organization (WHO) grade II ependymoma (magnification ×200, D).
In order to identify the source of bleeding, the patient was given an imaging examination of the full neuroaxis. The examination found an unusual T1 low signal of the cone region at the L2 level and a T2 mixed signal occupying lesion. Enhanced MRI showed the abnormal enhanced shadow of strip at L1-2 vertebrae, with clear boundary. On MRI, ependymomas without capsular rupture appear as well-circumscribed masses, which are hypointense on T1WI and hyperintense on T2WI. Homogenous enhancement following gadolinium administration distinguishes ependymomas from neurinomas of the cauda equina, which enhance heterogeneously [14]. Therefore, a preliminary diagnosis is often based on the ependymomas predilection site and imaging features. The patient was highly suspected of having an ependymoma based on imaging examination. The tumor was surgically removed, and the histopathologic examination of the resected tumor was diagnostic for a World Health Organization (WHO) grade II ependymoma.

Despite our case, there have been a small number of previous reports of ependymoma presenting as SAH. For example, a patient who presented with a history of severe thunderclap headache and who had SAH confirmed by lumbar puncture but negative results on cerebral angiography was found to have an anaplastic medullary ependymoma [10]. In another case, a patient with the clinical signs of SAH who had SAH confirmed by lumbar puncture but had negative cerebral angiography findings had an ependymoma of the conus medullaris [9]. There is also a report of a patient who presented with acute compression of cauda equina syndrome who was diagnosed with ependymoma causing SAH [11]. Taken together, lumbar puncture, cerebral and spinal imagings are of great value in the identification of ependymoma origin.

Conclusion

Subarachnoid hemorrhage of spinal origin is a rare clinical entity. The screening of SAH etiology should be emphasized, and when the patient is confirmed to have SAH but cerebral screening cannot demonstrate SAH and the particular cause, clinicians should be alert to the possibility of spinal SAH. Clinicians should be aware of and include SSCNS in the differential diagnosis of cerebellar-pyramidal syndromes. Early diagnosis and treatment can significantly improve the prognosis of patients.

Disclosure of conflict of interest

None.

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