Idiopathic Superficial Siderosis: A Case Report

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Superficial siderosis of the central nervous system (SSCN) is a very rare disorder. The clinical syndrome of SSNC consists of sensorineural hearing loss, cerebellar ataxia and myelopathy. The clinical syndrome together with the typical appearance on magnetic resonance imaging (MRI) of hyposignal intensity along the leptomeninges in T2 sequence permit the diagnosis of SSCN. A 58 year-old man who has a history of chronic progressive hearing loss and gait instability for 5 years is presented. The neurological examination revealed bilateral sensorineural hearing loss, cerebellar ataxia and mild spasticity of the lower extremities. MRI showed classical superficial siderosis in the form of hyposignal intensity along the leptomeninges in T2 sequence. The prominent sites of hemosiderin deposition in this case were cerebellar vermis, trigeminal nerves, vestibulocochlear nerves, around the brain stem and spinal cord surface. Cerebrospinal fluid findings confirmed chronic subarachnoid hemorrhage but bleeding site could not be demonstrated. There is no specific treatment available for idiopathic SSCN.

Keywords : *Superficial siderosis, Hemosiderin, Central nervous system, Magnetic resonance imaging*

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Superficial siderosis of the central nervous system (SSCN) is caused by chronic subarachnoid hemorrhage with deposition of hemosiderin in the leptomeninges, subpial tissues of the brain, spinal cord and cranial nerves. SSCN is a rare neurological entity. The classical clinical syndrome consists of sensorineural hearing loss, cerebellar ataxia and myelopathy[1]. Previously, the diagnosis of this clinical condition used to require histopathological confirmation[2,3]. Now the clinical diagnosis of SSCN can be confirmed by magnetic resonance imaging (MRI)[4]. This case report draws attention to this rare complication of chronic subarachnoid hemorrhage, which can be recognized early by its clinical triad and MRI findings.

Case Report

A 58-year-old Thai male presented with a five year history of progressive bilateral deafness and a three year history of gait ataxia. The hearing impairment began in the right ear and was diagnosed as age-related hearing loss. Two years later, he developed gait unsteadiness and the right ear became almost completely deaf. The hearing difficulty of the left ear had been noted during this period. Past history revealed no other neurological disorders, craniospinal injury or operation of the brain and spinal cord. He had never experienced any episode of severe headache, nausea and vomiting which could indicate acute subarachnoid hemorrhage. No history of diseases or medications which would cause bleeding tendency was notified. He was not an alcoholic. There was no history of taking any ototoxic medications. Family history was unremarkable for neurodegenerative diseases. General physical examination was unremarkable. Neurological examination revealed a healthy, alert and cooperative patient. No stiff neck or Kernig's sign was detected. Mental status examination showed no evidence of dementia or any psychiatric problem. Audiogram revealed sensorineural hearing loss of both ears, which was more on the right and high tone frequency loss was more prominent than lower tone frequency loss. The muscle power appeared normal, whereas spastic tone of both legs was observed. Deep tendon

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reflexes were increased in both knees. Babinski's sign was absent. Mild finger to nose test impairment and dysdiadokokinesia were detected on both sides, which were more prominent on the left. He could not perform tandem gait due to instability. Gaze-evoked nystagmus was also observed. Olfactory nerves, other cranial nerves and the rest of the neurological examination were normal. Routine laboratory tests which included complete blood count, BUN, creatinine, liver function test and screening coagulogram were normal. Computerized tomographic (CT) scan of the brain with contrast enhancement was performed and appeared unremarkable.

MRI of the brain and whole spine were performed using sagittal SE T1 weighted images, axial SE T1 weighted images, FSE T2 weighted images, FLAIR images, coronal GRE T2 weighted images and also diffusion images for MRI of the brain. Sagittal SE T1 weighted images, FSE T2 weighted images with fat suppression, GRE T2 weighted images and axial SE T1 weighted, GRE T2 weighted images were sequenced for the whole spine. MRA and MRV of the brain were also performed using TOF-MOTSA technique and 2D-TOF technique respectively. MRI, MRA and MRV of the brain were performed without contrast at 1.5 Tesla with a Horizon General Electric Scanner. MRI of the spinal cord was performed without contrast at 1.5 Tesla with Megnetom Vision Plus Seimen Scanner. T1 weighted sequences of both brain and spinal cord were unremarkable and no evidence of tumor and abnormal vessels were observed. Diffused superficial siderosis manifested as marked hypointensity FSE T2 weighted images along the parenchymal surface of upper pons, superior aspect of cerebellar vermis and exiting position of trigeminal nerves and vestibulocochlear nerves on both sides (Fig. 1,2). The lower pons, cerebellar peduncles, surface of spinal cord and lumbosacral nerve root sleeves were also involved (Fig. 3). Both magnetic resonance angiography and venography (MRA and MRV) of the brain revealed no abnormalities which could be the source of central nervous system bleeding.

Lumbar puncture revealed a normal open pressure of 150 mm.H_2O and the cerebrospinal fluid (CSF) was xanthochromic. CSF analysis revealed 6400 red blood cells/ml and protein of 52 mg/dl. No organisms were detected by gram stain, AFB stain, Indian ink preparation and CSF culture. VDRL of the CSF was negative. No malignant cell or siderophage were detected from cytologic examination of the CSF. CSF iron and ferritin were 14 ug/dl and 131.7 ng/ml

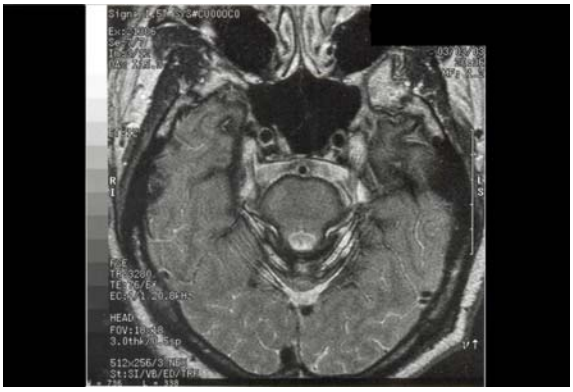

Fig. 1 T2 weighted MRI shows low signal intensity on the surface of pons, medial temporal lobes and cerebellar folia, and along the trigeminal nerves

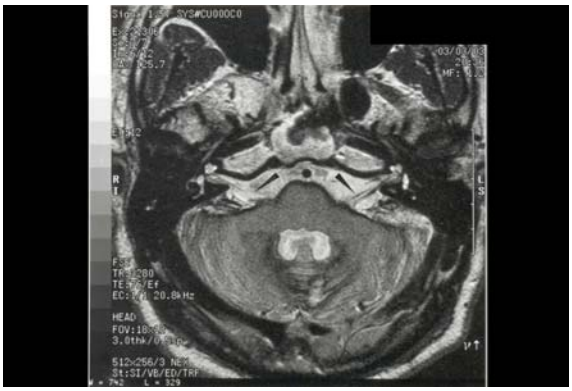

Fig. 2 T2 weighted MRI shows low signal intensity on the surface of pons and along the vestibulocochlear nerves (The arrows indicate the vestibulocochlear nerves)

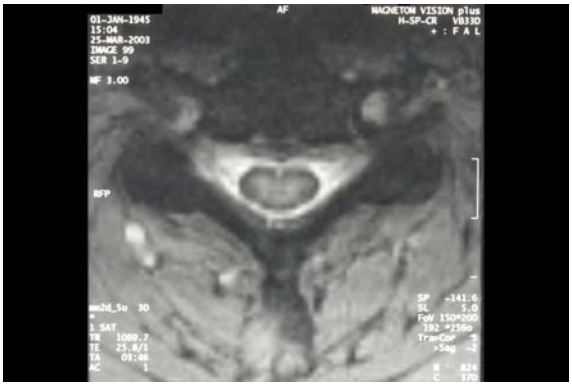

Fig. 3 T2 weighted MRI shows low signal intensity on the spinal cord surface

respectively. The clinical diagnosis in this case was idiopathic SSCN. Since there is no effective treatment for idiopathic SSCN, the patient was followed up at the outpatient department. After half a year of follow up the clinical status was stable.

Discussion

The main neurological findings in SSCN are sensorineural hearing loss (95%) cerebellar ataxia (88%) and pyramidal tract signs (76%)[1]. The pyramidal tract signs may be due to myelopathy or brainstem involvement[1]. The minority of cases have clear cut myelopathy e.g. transverse myelopathic syndrome, bladder disturbance or sensory level[1]. Other clinical syndromes include dementia, hydrocephalus, sciatica, neckache, oculomotor nerves, trigeminal nerves, olfactory nerves or facial nerves involvement[1]. This patient presented with a five year history of progressive sensorineural hearing loss. Later on he developed mild cerebellar ataxia. He also had mild spasticity of both legs which indicated the possibility of associated mild myelopathy. These clinical features are compatible with SSCN[1].

Prior to the introduction of modern neuroimaging technique, SSCN was diagnosed by meningeal biopsy or autopsy[1]. CT-scan of the brain has not been useful though it occasionally demonstrated meningeal enhancement and associated brain atrophy especially at the cerebellar vermis[5]. MRI is the investigation of choice for the diagnosis of SSCN, because it is sensitive and specific to the presence of hemosiderin[6]. The low signal intensity in T2-weighted images can be detected in all parts of the central nervous system; greatest at the superior cerebellar vermis, cerebellar hemisphere, around the brainstem, spinal cord and nerve roots[4, 6, 7]. In this case, the MRI T_2 weighted imaging showed marginal low signal intensity of the brainstem, vestibulocochlear nerves, trigeminal nerves, cerebellum, spinal cord and lumbosacral nerve roots. MRA and MRV failed to demonstrate abnormal cerebral and spinal vessels which might be the source of repeated bleeding in the central nervous system. He did not have any clinical or laboratory evidence of systemic bleeding tendency. CSF revealed only subarachnoid hemorrhage and increased protein level. There were no maglignant cells in the CSF. The CSF ferritin level was high (normal = 3.5 ± 0.55ng/mL)[8] which supported the evidence of siderosis involving leptomeningitis. The clinical and MRI, MRA, MRV findings permitted the diagnosis of idiopathic SSCN. The pathology of SSCN appears as marginal siderosis involving leptomeninges, granular ependymitis and obstructive hydrocephalus[1]. The common anatomical site of affected parts are usually associated to subarachnoid cisterns where there are high CSF volume and flow[1]. The most prominent sites are cerebellar vermis, the vestibulocochlear nerves, olfactory bulbs, crus cerebri and temporal cortex[1]. The other less preferential sites of involvement include cerebral cortex, cerebellar hemisphere, brain stem, spinal cord, other cranial nerves and spinal nerve roots[1]. Those sites of involvement reflect the clinical syndrome. The explanation of the predilection for some cranial nerves being affected by hemosiderin deposition such as olfactory nerves and vestibulocochlear nerves are the length of glial segment which extends from the central part of the nerve roots[9] and are susceptible to hemosiderin deposition together with the anatomical sites and distance where they lie in the subarachnoid space[1].

There is no definite treatment for SSCN except in patients who have definite sites of bleeding. The surgical treatment of SSCN consists of ablation of the bleeding source and shunting for associated hydrocephalus[10]. Iron chelating agents have not been proven to have any beneficial effect[10]. An attempt to reduce the oxidative toxic effect of heme-iron complex by using monoamine oxidase B inhibitor (selegiline) and vitamin C have been investigated[10].

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ภาวะสารฮีโมซิเดอริน สะสมบนผิวประสาทส่วนกลาง

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สารฮีโมซิเดอรินสะสมบนผิวประสาทส่วนกลาง เป็นภาวะที่พบได้น้อย ประกอบด้วยกลุ่มอาการ 3อย่างคือ หูหนวกตึง เดินเซที่เกิดจากสมองน้อยผิดปรกติ และอาการเกร็งหรือขาอ่อนแรงจากไขสันหลังผิดปรกติ การวินิจฉัยภาวะสารฮีโมซิเดอรินสะสมบนผิวประสาทส่วนกลางนั้นอาศัยกลุ่มอาการดังกล่าวและภาพเอ็กซ์เรย์คลื่นแม่เหล็กที่มีลักษณะสัญญาณต่ำในภาพ ทีสอง (T2) ที่บริเวณผิวระบบประสาทส่วนกลางและเส้นประสาทสมอง ซึ่งเป็นลักษณะเฉพาะ รายงานนี้กล่าวถึงผู้ป่วยชายไทยอายุ 58 ปีที่มีอาการหูหนวกตึง และเดินเซ จากประวัติการตรวจร่างกายและการตรวจด้วยคลื่นแม่เหล็ก เข้าได้กับภาวะสารฮีโมซิเดอรินสะสมบนผิวประสาทส่วนกลาง โดยพบความผิดปรกติอย่างชัดเจนบริเวณสมองน้อยส่วนเวอร์มิส เส้นประสาทสมองคู่ที่ 5 เส้นประสาทสมองคู่ที่ 8 รอบก้านสมองและประสาทไขสันหลัง ผลการตรวจน้ำหล่อไขสันหลังพบลักษณะที่เข้าได้กับการมีเลือดออกในเยื่อหุ้มสมองเรื้อรัง แต่ไม่พบตำแหน่งเลือดออก ในปัจจุบันยังไม่มีการรักษาภาวะสารฮีโมซิเดอรินสะสมบนผิวประสาทส่วนกลางที่ไม่พบสาเหตุเลือดออก