Case Report

Superficial Siderosis Due To Third Ventricular Cavernous Malformation: Case Report

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Abstract
Cavernous malformations can be seen anywhere in the central nervous system. However their existence in intraventricular area is very rare. Superficial siderosis is an extensive deposit of hemosiderin in the leptomeninges and pial surface. Superficial siderosis due to intraventricular cavernous malformation (CM) is very rare. Here we report a 50-year-old male with superficial siderosis due to third ventricular cavernoma. The patient had multiple hemorrhages due to CM. He had subtotal surgical removal of the lesion. Lately he developed ataxia, speech disorder and right hemiparesis. Gamma knife radiosurgery was performed for the residual CM. Short period of dexamethasone treatment partly solved the clinical complaints. The present case is the first report of superficial siderosis due to third ventricular CM.

Keywords: Cavernous malformation, superficial siderosis, third ventricle

INTRODUCTION

Cavernous malformations are benign vascular malformations, which constitutes approximately 5 to 10% of the central nervous system vascular malformations. CM's are present in about 0.4 to 0.8% of the population, according to the findings of autopsy series and large magnetic resonance imaging (MRI) based studies. Superficial siderosis of the central nervous system is a rare condition. Superficial siderosis is a radiological and pathological condition in which there are extensive deposits of hemosiderin in the leptomeninges, subpial region, and subependymal region in the brain, spinal cord and cranial nerves. This may cause
to progressive cerebellar ataxia, sensorineural hearing loss and pyramidal sign\(^7\). Intraventricular CM's are rare and its occurrence with superficial siderosis even more rare\(^11,12\).

Here we reported a patient with third ventricular CM and superficial siderosis.

**CASE PRESENTATION**

Fifty–year-old male had suffered right hemiparesis due to CM related intraventricular hemorrhage 16 years ago. He had multiple hemorrhages in the following years. He had subtotal removal of the CM two years ago. He had speech disorder and ataxia in the last 2 years. MRI revealed third ventricular popcorn like rounded lesion and the dimensions was 1x1 cm (Fig 1 and 2). T2-weighted MR images showed a rim of low signal intensity around the medulla, pons, midbrain and basal cortical surface (Fig 3a and b). Gamma knife (Elekta, Atlanta, Georgia, USA) radiosurgery was performed for the residual CM with 12 Gy dose to 50% isodose line. The patient received dexamethasone 4 mg/day for 12 weeks. His complaints were partly resolved. After 2 years of follow-up he had no new hemorrhage.

![Fig 1: Third ventricular cavernous malformation was irradiated with 12 Gy to 50% isodose line.](image1)

![Fig 2: Sagittal T1-weighted image demonstrates the cavernoma in the third ventricle.](image2)
DISCUSSION

CM's typically diagnosed during the 2nd and 5th decades of life. A single CM is found in 75% of sporadic cases and only 8% to 19% of familial cases. Clinical presentation is usually nonspecific and may include seizure, headache, paresthesia and memory loss.

Intraventricular CM is a rare pathological entity, constituting 2–10 % of all cerebral CM's. Intraventricular CM's may cause to hemorrhage, focal neurologic deficits and headache. Hydrocephalus may be seen due to hemorrhages or by directly due to obstructive effect of CM. Reyns et al. published their clinical series in 1999 and told that 44 % of the intraventricular CM's are located in third ventricle.

Superficial Siderosis (SS) of the central nervous system is a progressively deteriorating neurological condition. Common symptoms are sensori-neural hearing impairment, ataxia, myelopathy, urinary disturbance, headaches, and anosmia. Hemosiderin deposition along the leptomeninges, subpial tissues, and subependyma of the ventricles as a result of chronic, recurrent subarachnoid hemorrhage induces intracellular uptake of iron, leading to damage of neural tissue. Interestingly, cranial nerves I and VIII are the most compromised cranial nerves, because they remain within the CNS for a relatively long distance outside of the brain and containing siderosis- susceptible microglia.

Common causes of superficial siderosis are recurrent hemorrhage, trauma, hemorrhagic neoplasms and vascular malformations.

In the past superficial siderosis were diagnosed by autopsy. After the invention of MRI this may be revealed by MRI properly. MRI is very sensitive to hemosiderin deposits. It shows marked hypointensity on T2-weighted images. Multiple bleeding is the main reason of superficial siderosis. In the literature it is published that bleeding source has been reported in only 54-75 of cases. Calcifications may be seen macro and microscopically. Computed tomography may be enough to show cavernomas but not for superficial siderosis.

Fig 3: Axial T2-weighted MR images demonstrates rims of low signal intensity surrounding the brain stem (a) and basal cortical surface (b).
Unfortunately, there is no therapy to reverse the damaging effects of hemosiderin deposition in the brain. At the present, the aim of the therapy is to prevent the progression of the condition and preserve existing neurological function. The most commonly attempted therapy for the hemosiderin deposition in the brain is iron chelation using a medication such as desferrioxamine. However, only a single case report has described an improvement in symptoms or halting of the progression of the disease with the use of iron chelators such as Trientine\(^8\). Steroids were used in one patient with anti-Ri autoimmune disease\(^8\). For treatment, iron chelators have not proven beneficial, and ongoing studies are needed.

Intraventricular cavernous malformation with superficial siderosis is very rare. There are only two case reports in the literature. Hashimoto and Hoyt reported a patient with lateral ventricle CM related SS\(^2\). They did not mentioned about treatment in this patient. Hsu et al reported multiple CM's related SS in a patient\(^3\). There was a lateral ventricle located cavernoma in this patient. Most probably this was the reason for SS. No surgical or medical treatment was used for this case. Lately a case report published by Sabat, which is a 60-year-old woman with CM located in the left lateral ventricle\(^11\). The patient had surgical removal of the CM and had dexamethasone 4 mg/day for 6 weeks. Her complaints have considerably improved.

The present case report is the first case report of superficial siderosis due to third ventricle CM. Gamma knife radiosurgery helped the patient to prevent from new bleedings. Also short course of dexamethasone relieved the complaints partly.

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