

Spinal ependymoma presenting with visual and hearing impairment – An unusual sequelae

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ABSTRACT

Superficial siderosis of the central nervous system is a rare complication of recurrent bleeding in the neuraxis which may present with various symptoms. We report a case of spinal tumor presenting with superficial siderosis with involvement of vision and hearing along with ataxia. This is a rare presentation of superficial siderosis. The pathogenesis of selective involvement of these cranial nerves is discussed.

Key words: Recurrent sub arachnoid hemorrhage, gradient echo imaging, myxo papillary ependymoma, CNS superficial siderosis

INTRODUCTION

Spinal tumors with recurrent bleeds present occasionally as superficial siderosis (SS) of the central nervous system, characterized by deposition of hemosiderin in leptomeninges and cranial nerves, followed by gliosis, neuronal loss and demyelination.^[1-3] Sensorineural deafness, cerebellar ataxia and myelopathy form the classical clinical triad.^[4] MRI of the brain with gradient echo sequences is the gold standard for diagnosing this condition,^[5] the treatment of which is to surgically ablate the source of bleeding. We report an unusual case of superficial siderosis secondary to hemorrhage in a spinal myxopapillary ependymoma. In addition to the classical clinical presentation, this patient also had visual impairment, which is an uncommon feature.

CASE REPORT

A 57-year-old lady presented with a five-year history of blurring of distant vision, bilateral hearing impairment and gait ataxia. On evaluation, she was found to have best corrected visual acuity of 6/12 in both eyes with normal fields, fundi and bilateral high tone sensorineural loss of

45 dB. Visual evoked potentials showed a prolonged P100 latency of 119.1 ms [Figure 1a] and BAER showed mildly prolonged I-III interval of 2.51 ms [Figure 1b] suggestive of demyelination. MRI of brain revealed T2 hypointense coating of brainstem, cerebellum, cerebello-pontine angle cisterns [Figure 2a] and optic tracts [Figure 2b], which showed blooming on gradient echo imaging, suggestive of hemosiderosis. A diagnosis of superficial siderosis of CNS was made and further imaging was done to identify the source of bleeding. There was no contrast enhancing lesion in the brain. An MR angiogram was performed, which was normal. On imaging the spine, siderosis was found extending into the spinal canal. MRI of the dorsolumbar spine then revealed a heterointense enhancing intradural lesion [Figure 2c] at L 2-3 region, suggestive of a tumor with bleed. She underwent decompression of the tumor adherent to cauda equina roots, which was diagnosed as myxopapillary ependymoma with hemorrhage on histopathology [Figure 2d]. At two-year follow up, she had no further visual deterioration though ataxia had mildly worsened.

DISCUSSION

Myxo-papillary ependymomas have a propensity to bleed, but presentation as CNS siderosis is rare with only 15 cases reported in literature.^[3,4] The other lesions causing SS are oligodendroglioma, astrocytoma, paraganglioma,^[2,4] spinal and cranial vascular malformations, cranial or cervical trauma, brachial plexus injury,^[6] previous surgery especially in posterior fossa and bleeding disorders.^[7] In cases of recurrent sub-arachnoid hemorrhage,

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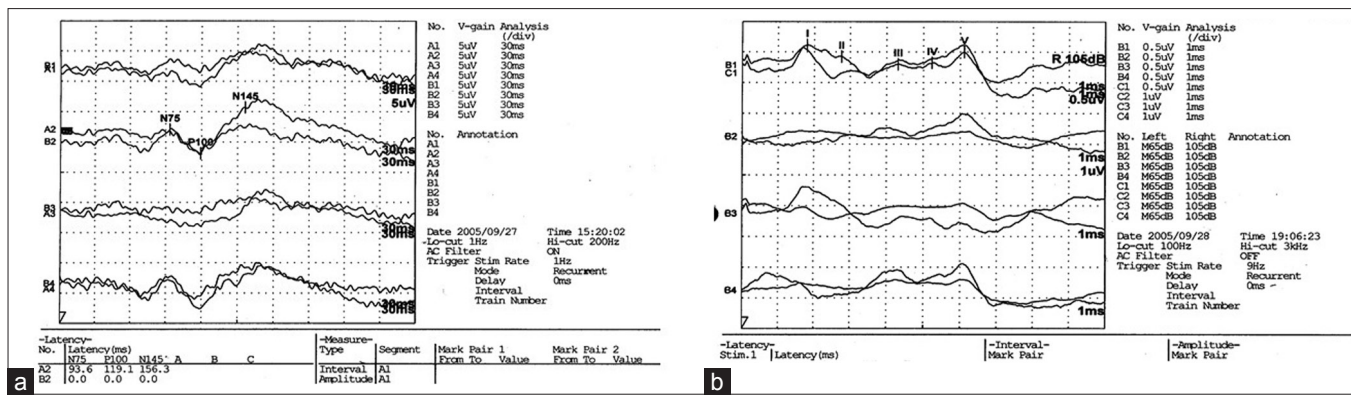


Figure 1: (a) Visual evoked potential (VEP) showing prolonged P100 latency of 119.1 ms, (b) Brainstem auditory evoked response (BAER) showing mildly prolonged I-III interval of 2.51 ms

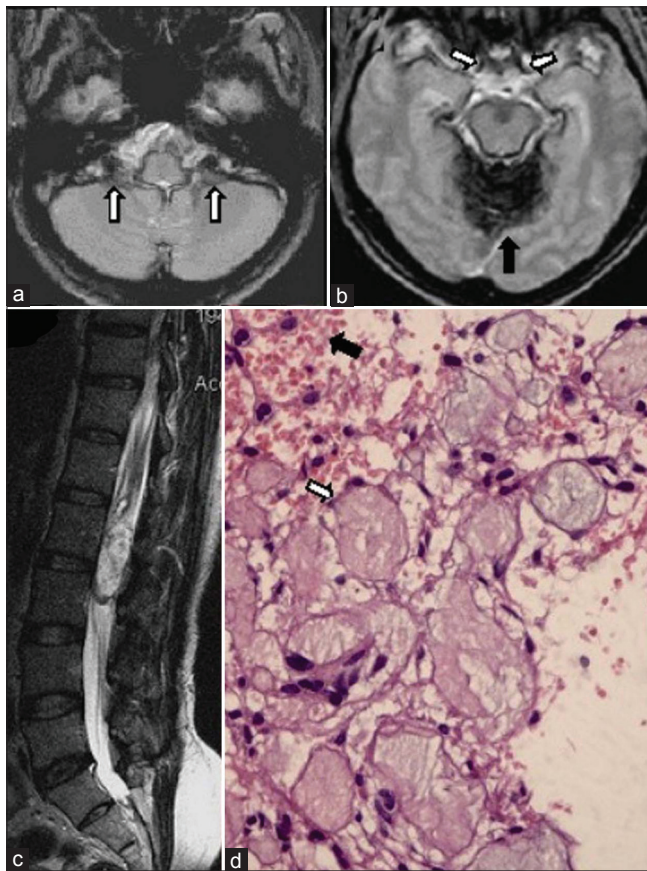


Figure 2: (a) MRI of brain gradient echo axial sequences with arrows showing blotches of hemosiderin in cerebello-pontine angle cistern with blooming, (b) MRI of brain T2W axial images showing optic tracts (white arrows), midbrain and superior vermis (black arrow) lined with hemosiderin, (c) MRI of dorso lumbar spine T2W sagittal images showing a heterogenous well circumscribed lesion at L2-3 level, (d) Histopathology (H and E stain) showing myxo-papillary ependymoma (white arrow) with bleed (black arrow)

central myelin and glial cells phagocytose hemosiderin and biosynthesize ferritin, which is in turn toxic to them.^[1] The triad of CNS superficial siderosis consists of progressive sensorineural deafness, cerebellar ataxia and pyramidal signs. However, other cranial nerves can also be rarely involved. Selective involvement of

olfactory, optic and vestibulocochlear nerves is due to the central myelin and glia covering these nerves in their cisternal course unlike the other cranial nerves. Despite evidence of coating of the optic tracts with hemosiderin, presentation with visual deterioration has been exceedingly rare, with only two reported cases, where both revealed other associated pathologies and normal VEP.^[4,5] Clinically, impairment of smell is often not noticed and mild peripheral visual deterioration is often overlooked due to the sparing of the central macular fibers.^[1] On comprehensive ophthalmologic examination, no other pathology responsible for the visual impairment could be identified. The patient also had prolonged p100 in VEP. Hence, this symptom has occurred as a direct result of the SS as evidenced on MRI. As suggested in all the previous case reports, a high index of suspicion is required for the diagnosis of CNS superficial siderosis. Characteristic MR imaging is the presence of hypointense rim around the neural structures, superior vermis and anterior cerebellum on T2 weighted sequences indicative of hemosiderin deposition, with blotches of hemosiderin in the cerebellopontine angle cisterns. MRI with gradient echo sequence shows classical blooming in these areas and has to be done in all cases of multiple cranial nerve involvement to rule out this condition. Identification of the source of the bleed is usually a challenge and spinal imaging should be included for completion. In about half of the patients, the source of bleeding remains unknown. These patients usually have a progressively disabling neurological course.^[4,6] The only available treatment is surgical ablation of the source of bleeding. However the clinical effects of disease can progress even after that.^[8] Our patient had a worsening of her gait on follow up, though she was stable in her cranial nerve symptoms.

CONCLUSION

This case highlights the rare clinical presentation of visual deterioration occurring primarily due to superficial

siderosis. It underscores the role of gradient echo MR imaging for accurate diagnosis. Careful search for and early surgical excision of the causative lesion is recommended.

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