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Case Report

Eight-and-a-half syndrome in pontine cavernoma: a rare presentation

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ABSTRACT

Cavernous vascular anomalies are varied in form, and cavernomas are very rare lesions commonly found in the supra-tentorial region. Eight-and-a-half syndrome includes horizontal gaze palsy, internuclear ophthalmoplegia, and facial nerve palsy, which is rare in clinical practice. Etiologies are commonly infarct and demyelination. Cavernomas are space-occupying lesions that can lead to life-threatening complications due to haemorrhage, and cavernomas resulting in "eight-and-a-half syndrome" are extremely rare. We report a case of pontine cavernoma presenting as "eight and half syndrome," which worsened systemically, but with timely intervention, the patient recovered completely with good follow-up and an intact permanent tarsorrhaphy.

Keywords: Horizontal gaze palsy, Internuclear ophthalmoplegia, Facial palsy, Cavernoma, Tarsorrhaphy

INTRODUCTION

One-and-a-half syndrome is a rare supranuclear condition characterised by ipsilateral conjugate horizontal gaze palsy and internuclear ophthalmoplegia due to involvement of the paramedian pontine reticular formation and medial longitudinal fasciculus in pons. If facial nerve fasciculus is involved, it results in the rarest eight-and-a-half syndrome. Etiologies of this rarest entity are ischemia/demyelination, and space-occupying lesion contributing to it is very rare.[1] Cavernoma is a vascular malformation seen in less than one per cent of the population and which bleeds rarely. We report a rare case of pontine cavernoma with eight-and-a-half syndrome. Patient underwent resection of the pontine lesion and subsequently underwent permanent lateral tarsorrhaphy for the facial palsy, and on follow-up, she was doing well. To best knowledge, eight and half syndrome is a very rare condition, and the vascular malformation of the pons that contributes to it makes it extremely uncommon.

CASE REPORT

A 38-year-old young female presented with a one-month history of left upper and lower limb weakness associated with headache. Bulk and tone were normal, with powers of the upper and lower limbs of 4/5 and normal pain, temperature, and pressure sensations. Ophthalmological examination revealed visual acuity in both eyes of 6/6 (Snellen's) with right-side horizontal gaze palsy with end gaze nystagmus of opposite gaze and right-side inadequate lid closure with poor Bell's phenomenon and deviation of the angle of the mouth to the left and weakness of right-side facial muscles suggesting facial palsy, from which the diagnosis of eight and a half syndrome was made. Imaging revealed a well-defined lesion in the right side of the pons, which was hyperintense on T1 with mixed signal intensity on T2-weighed images with mass effect on the pons, along with features suggestive of a developmental venous anomaly draining into the left transverse sigmoid sinus. On the same day, the patient deteriorated and was intubated, with imaging revealing a large bleed in the pontine region, for which she underwent surgical intervention with decompression and removal of the lesion, which was revealed to be a cavernoma with fresh and old haemorrhage on histopathological examination. The patient improved systemically over time, but she still had horizontal gaze palsy with facial nerve involvement on the right side, for which she underwent right eye lateral

permanent tarsorrhaphy with no further worsening. She underwent periodic imaging to look for any evidence of rebleeding.

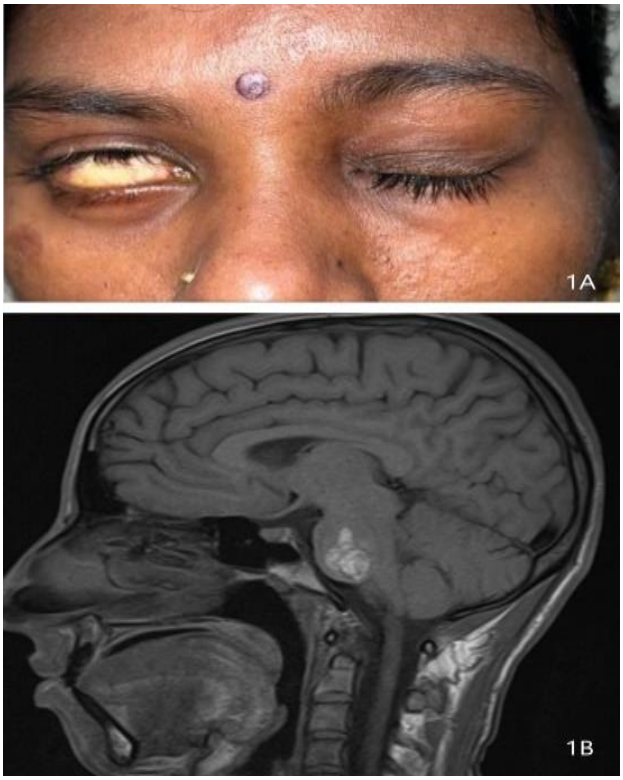

Figure 1 (A and B): Clinical photograph of patient showing right side inadequate lid closure due to facial nerve palsy. Sagittal view of MRI brain of well-defined hypertense lesion in brainstem (Cavernoma).

DISCUSSION

Conjugate horizontal gazes are supranuclear in origin, and the centre for them is the paramedian reticular formation in the pons close to the facial colliculus, which sends signals to the ipsilateral abducens nucleus and the contralateral medial longitudinal fasciculus. Internuclear ophthalmoplegia associated with facial palsy is called "eight and half syndrome," for which pontine infarct is the commonest etiology. The condition was first described by Eggenberger in 1988 and is caused by vertebral basilar disease.[2] Cavernomas are cavernous vascular malformations composed of abnormal capillaries with hemosiderin deposits and gliosis; the majority are supra-tentorial.[3] Cavernomas are slowly growing lesions that, in many cases, are asymptomatic; bleeding is reported rarely and results in stroke. Space-occupying lesions contributing to the "eight-and-a-half syndrome" are very rare, and the aetiologies like infarction and demyelination have a variable prognosis.[4,5] The choice of intervention in cavernoma is microsurgical and depends on various factors, including the location of the lesion and its accessibility, size, severity, and patient choice. Facial nerve palsy leads to complications, especially neuroparalytic keratitis, due to inadequate lid closure. Management in reversible cases includes artificial tears and taping of the lids until the lid closure recovers; in irreversible cases, tarsorrhaphy is required.[6] We report a case of pontine cavernoma in a patient who presented with internuclear ophthalmoplegia with associated facial nerve palsy and worsened systemically due to bleeding from the cavernoma, for which emergency surgical intervention was carried out, and the patient had a good recovery post-operatively and is doing well with permanent tarsorrhaphy. To our knowledge, eight and half syndrome is very rare, and a space-occupying lesion contributing to it is extremely rare.

CONCLUSION

Horizontal gaze palsy, facial nerve palsy, and inter-nuclear ophthalmoplegia contributing to eight and half syndrome due to pontine cavernoma are very rare. Bleeding and re-bleeding in cavernomas can be life-threatening and require continuous follow-up.

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