

CAVERNOMA OF CERVICOMEDULLARY REGION PRESENTING WITH HEMIHYPESTHESIA

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ABSTRACT

Medullary cavernomas only contribute to 5% of brain stem cavernomas, various clinical presentations for medullary cavernomas have been described like intractable hiccups, dysphagia, hemiparesis, anorexia nervosa and hemihypesthesia. Exact pathomechanism for developing sensory symptoms has not been described earlier. Pressure effects due to changing dynamics in this cavernous malformation resulting in mass effect over the traversing sensory tract fibers is the possible mechanism. Differentials of cavernoma should be considered for any case presenting with hemihypesthesia. Surgery should be done only when there is appropriate indication, as surgery itself causes significant morbidity.

Keywords: cavernoma, cavernous angioma, medulla oblongata, hemihypesthesia

INTRODUCTION

Cavernomas are rare congenital benign angiographically occult vascular malformations of brain accounting for 5-15% of all brain vascular malformations. (1-3) Cavernomas more commonly occur in supratentorial brain parenchyma. (1,2,4) Brain stem cavernomas (BSC) constitute 10-15% of all intracerebral cavernomas and are more likely found in pons and midbrain (95% of cases), medullary cavernomas only contribute to 5% of BSC. (1,2,4) Various clinical presentations of medullary cavernomas have been described like intractable hiccups, dysphagia, hemiparesis, anorexia nervosa and hemihypesthesia. (1, 3-7) We present a case of a 51 year old male who presented to us with hemihypesthesia and we found the probable cause to be medullary cavernoma.

CASE REPORT

A 51 year old male came to neurology OPD with decreased sensation and altered perception for temperature on the left side of body which was happening for over a period of 6 months. He was non-alcoholic and non-diabetic. There was no history of recurrent headaches, diplopia, seizures, dysphagia, ataxia or intractable hiccups. On examination, his vitals were normal and laboratory investigations showed: CBC: Hb-13 gm/dl, total count -4500/mm³, Plts-4.5 lacs. Neurological examination: higher mental functions were well preserved, he was coherent, motor power in all limbs were 5/5, tone was normal. Cranial nerve examination revealed no abnormality. Sensory system examination revealed decreased light touch, deep touch, pain, temperature and vibration sensations over left half of body

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with more marked decrease in the left lower limb. MRI was performed to identify the cause of his hemihypesthesia (Fig. 1). It showed T1, T2, FLAIR hyperintense lesion in the lower medulla and cervicomедullary junction, with hypointense hemosiderin rim being well appreciated on T2 (Pop-corn appearance). On susceptibility weighted imaging, lesion was showing peripheral blooming. There was no significant contrast enhancement or restriction of the lesion. Another small similar appearing lesion was seen in the left superior frontal gyrus. Based on typical imaging features, diagnosis of cervicomedullary junction cavernous angioma was made. Patient was counselled and was advised to undergo surgery for the same as the lesion was in a critical location. However, the patient opted out of surgery temporarily pertaining to his family commitments and said that he will consider this option at a later date.

DISCUSSION

Cavernomas are lobulated, reddish purple, well circumscribed lesions composed of compactly packed angiogenically immature blood filled locules (caverns) lined by endothelial septae, fibrous adventitia and are devoid of elastic and muscle fibers (1,3,5). No interspersed brain parenchyma or neural elements can be found within the lesions. These well-defined lesions are surrounded by a hemosiderin rim and sometimes caverns can be filled with thrombi within (1,3,5). Approximately 90% of cavernomas are sporadic, remaining are familial (8). Mutations in chromosome 7 have been attributed for these malformations (3,5). Most cavernomas have benign acalical course. Supratentorial lesions present with seizures, infratentorial lesions especially brainstem lesions have been found to have more aggressive course as their propensity for

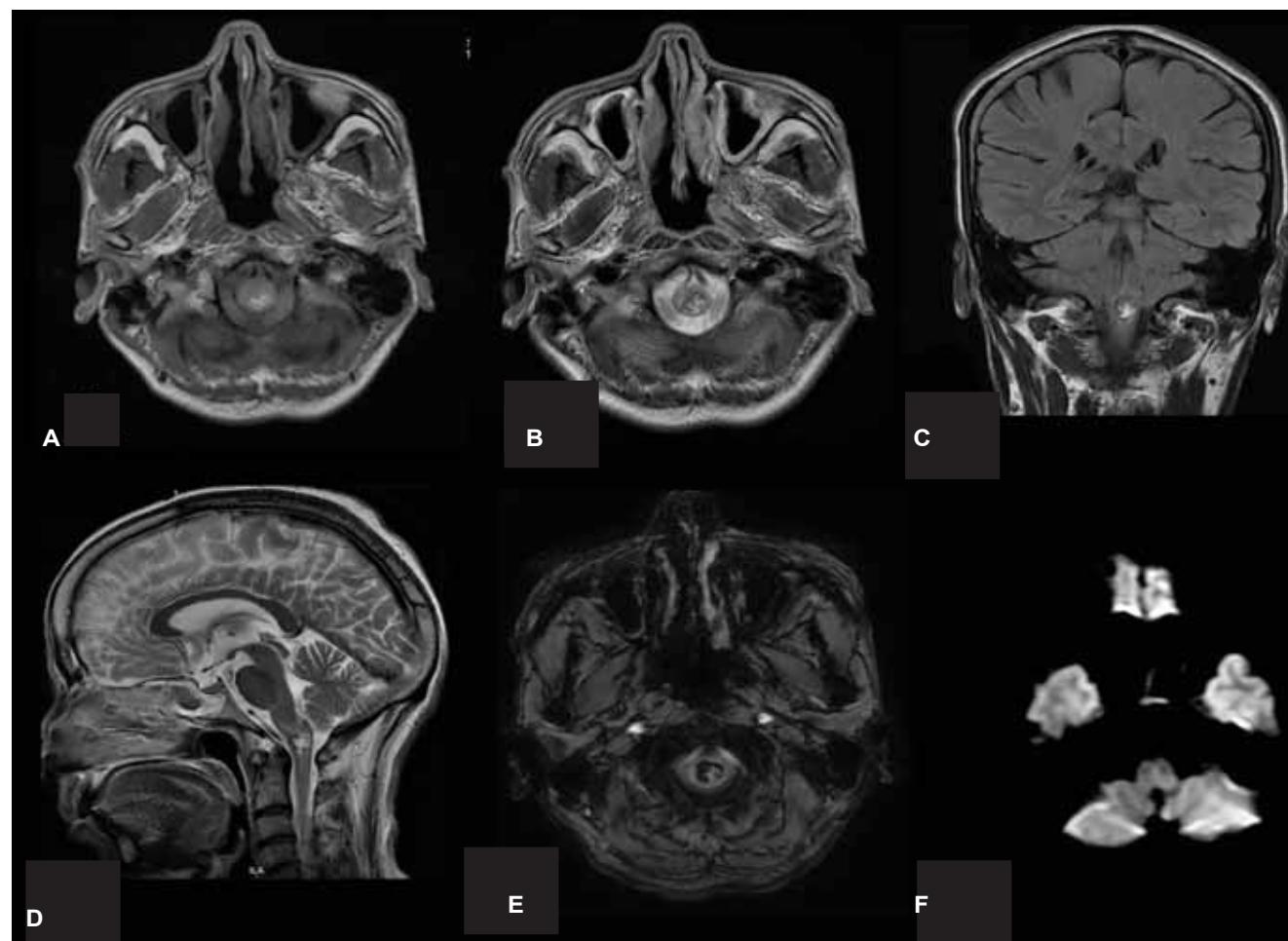


FIGURE 1. Axial T1W image (A), Coronal FLAIR image (C), showing hyperintense lesion in lower medulla and cervicomедullary junction. On T2 weighted axial (B) and Sagittal (D) images the lesion is hyperintense and is showing peripheral hypointense hemosiderin rim, perilesional vasogenic edema was also seen. On susceptibility weighted images lesion is showing peripheral blooming (E). There was no restriction seen on diffusion weighted image (F).

primary and recurrent bleed is much higher than ones in supratentorial location (1,3,5). Clinical symptoms of medullary and cervicomedullary cavernomas are variable. Medullary lesions causing hemihypesthesia and sensory symptoms have been described earlier. Hao li et al in their study had 5 cases of brain-stem cavernomas presenting with hemihypesthesia (1). K.H Lee et al reported a cervicomedullary cavernoma presenting with intractable hiccups their patient also had hemisensory symptoms (7). Similarly N. Fayed et al. reported one case of medullary and one case of cervicomedullary low grade astrocytoma presenting with hemisensory syndrome (9). Exactpathomechanism for developing sensory symptoms has not been described earlier. Pressure effects due to changing dynamics in this cavernous malformation resulting in mass effect over the traversing sensory tract fibers is the possible mechanism (4). Complications of brain stem cavernoma are mainly due to intralosomal thrombosis, bleeding, and progressive dilation of the caverns and rupture of endothelial septa between locules (1,5). It is also noteworthy that the time interval between primary and secondary bleeds is less for BSC (1,3,5). MR imaging is the diagnostic modality of choice to diagnose cavernous angiomas. Heterogeneous signal intensity lesion on T1 and T2 sequences with lobulated mar-

gins showing T2 hypointense hemosiderin rim which blooms on susceptibility imaging is classic appearance of cavernomas (3,7,10). Contrast MRI is not indicated in these lesions unless they show some atypical appearance (3). Once diagnosis is established it is very important to take a choice between conservative management and surgery it is a general tendency to resect the superficial cavernomas and observe the deep seated ones should aim in complete resection of tumor and it should avoid additional neurological compromise (11). Patient selection for surgery isutmost important as mishandling of these lesions in vital locations can cause significant morbidity (1).

CONCLUSION

Cavernomas of brain stem have more dynamic course than supratentorial cavernomas with chance of primary and recurrent bleeds being much higher in BSC. Medullary/Cervico-medullary cavernomas are rare and can present with variable clinical symptoms. Differentials of cavernoma should be considered for any case presenting with hemihypesthesia. Surgery should be done only when there is appropriate indication, as surgery itself causes significant morbidity.

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