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RESEARCH ARTICLE

SUBEPENDYMAL GIANT CELL ASTROCYTOMA IN TUBEROUS SCLEROSIS

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ABSTRACT

Subependymal Giant cell astrocytoma (SEGA) are benign tumors of glioneuronalorigindistinct from astrocytoma and is common cerebral neoplasm in tuberous sclerosis with presence of tubers and subependymal glial nodules in majority of patients. Tuberous sclerosis is a genetic disorder resulting in benign lesions in multiple organs including the brain, skin, eyes, heart, kidney and lung with systemic manifestations including seizures, cognitive impairment, dermatologic abnormalities. Large subependymal giant cell astrocytoma leads to occlusion of foramen of Monro with hydrocephalus. Subependymal giant cell astrocytoma (SEGA) patients usually present with symptoms of elevated intracranial pressure from obstructive hydrocephalus.

INTRODUCTION

Tuberous sclerosisis autosomal dominant genetic disorder (1). Tuberous sclerosis complex (TCS) leads to development of benign hamartoma in several organs, including the brain, skin, eyes, kidney, heart, lungs and liver. Estimated prevalence of tuberous sclerosis is 1/ 6800 and 1/ 15000.SEGAs occur in 1.7-26% of patients with tuberous sclerosis(2-4).The main structural brain lesions seen in tuberous sclerosis are subependymal nodules (SENs), subependymal giant cell astrocytoma (SEGA) and cortical tubers (5). SEGAs usually arise from subependymal nodules in the wall of lateral ventricle of patients with tuberous sclerosis (6). The main difference between SENs and SEGAs is size (with cutoff size ranging from 5-10 mm) and location: SEGAs are typically at the caudo-thalamic groove while SENs are located in the ependyma of the lateral ventricles along the caudate nucleus. SEGAs will grow, whereas SENs remain stable in size (7).

Case Summary

A 26-year-old female came with complaint of headache, multiple episodes of projectile vomiting, diplopia and giddinessassociated with tingling numbness in bilateral upper and lower limbs since one year. No history of head trauma, diplopia, seizures, loss of consciousness.

Imaging Findings

Axial plain CT Scan revealed a well defined slightly hyperdenselesion measuring approx. 15 mm (Transverse) x 14 mm (Anteroposterior) x 15 mm (Cranio-caudal) at level of foramen of Monro on right side of midline causing obstructive dilatation of right lateral ventricle (Figure 1a) showing multiple small hyperdense foci of calcification. Asmall 2 mm calcified lesion was noted in relation to body of left lateral ventricle, subependymal in location (Figure 1b).Left lateral ventricle, 3rd and 4th ventricle appeared normal.

Plain and contrast enhanced MRI Scan of the brain showed a well defined lesion measuring approx. 15 (Transverse) x 14 (Anteroposterior) x 15 mm (Craniocaudal) was noted at level of foramen of Monro on right side of midline causing obstructive dilatation of right lateral ventricle. It was heterogeneously hypointense on T1WI (figure 2a), heterogeneously hyperintense on T2WI and FLAIR (figure 2b and c) with foci of blooming on GRE suggestive of calcification (figure 3a) with no restricted diffusion on DWI (figure 3b). Contrast study showed moderate heterogeneous enhancement (figure 4 a-c). Small hyperintense foci were noted in left frontal, right parietal subcortical white matter on T2WI and FLAIR –suggestive of tubers/ hamartomas (figure 5 a,b).

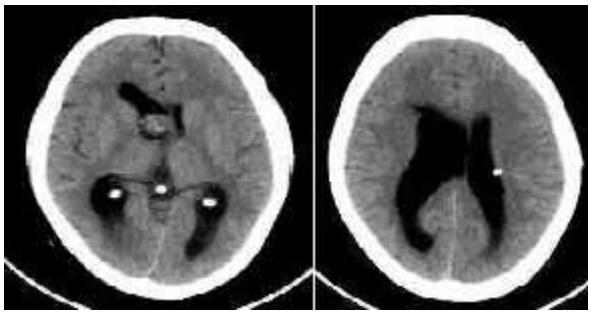

Figure 1 (a,b) Plain CT Brain Axial

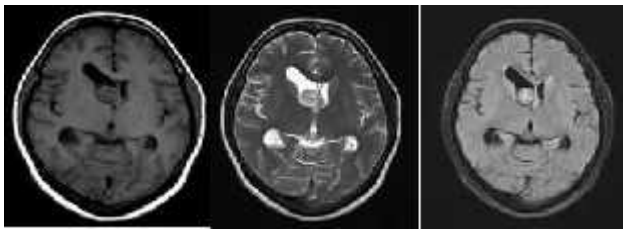

Figure 2 (a,b,c) MRI Brain- Axial- T1WI (a), T2WI (b),FLAIR (c),

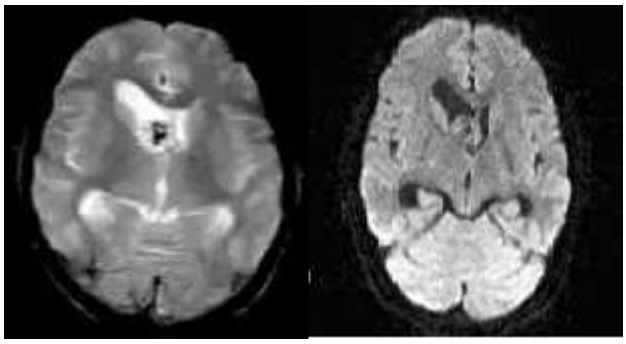

Figure 3 (a,b) MRI Brain- Axial – GRE (a), DWI (b),

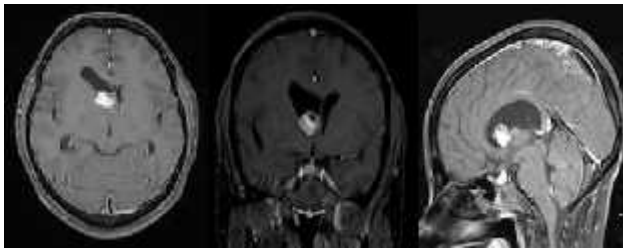

Figure 4 (a-c) T1FS Contrast- Axial(a), coronal(b) and sagittal(c)

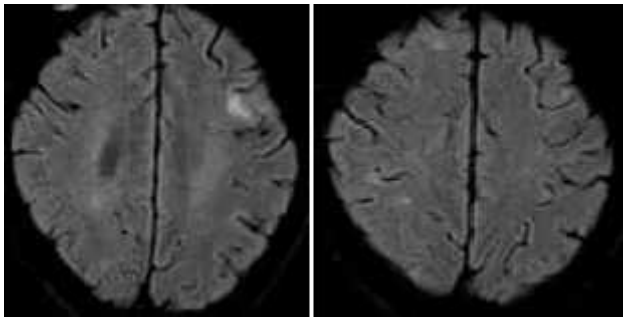

Figure 5 (a,b) MRI Brain- Axial - FLAIR

DISCUSSION

Cortical tubers develop in 85% of patients. Cortical tubers are collection of giant cells, dysmorphic neurons, and gliosis(8). Subependymal nodules are collections of abnormal, swollen glial cells these can calcify and progress into subependymal giant cell astrocytomas, which are indistinguishable from SENs histologically but distinguishable based on larger size and potential for mass effect compared to that of SENs [9,10]. The radiographic appearance of SENs on noncontrast CT show small calcified foci along the walls of the lateral and third ventricles and on MRI it shows hyperintensity on T1WI and isointense to hyperintense on T2WI (11). The most common location of SEGAs is the foramen of Monro, causing obstructive hydrocephalus. SENs can transform into SEGAs due to presence of Intermediate cells in SENs and SEGAs(12).SEGAs show more intense contrast enhancement on CECT and are larger tumors (>1 cm**)** compared to SENs(13). There are increased chances of malignant transformation in masses located at the foramen of Monro. The imaging findings between subependymal giant cell astrocytoma and subependymal nodule is based on its incomplete calcification, enhancement with gadolinium, and size of more than 1 cm (14). Faint subependymal nodule calcification can be completely missed on MRI. Subtle lobulations or asymmetry on the ventricular margins should prompt radiologist to exclude subependymal calcified nodules. These can be better demonstrated on plain CT or gradient echo sequences. Rarely cystic lesions may be found in subcortical and white matter location in cerebral hemisphere in tuberous sclerosis. These are due to cystoid brain degeneration. (8, 14).

CONCLUSION

The proximity to the foramen of Monro, incomplete calcification, enhancement on MRI and large size (>1 cm) make SEGA the likely diagnosis. Most SEGA's will show enhancement after contrast administration; however, a growing subependymal lesion in the absence of enhancement should be considered as SEGA.

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