Gliomatosis Cerebri  -  A case report

Clinical History: A 40 year old male presented with many months history of altered behaviour with excessive sleep. H/o trivial fall from chair one day prior to MRI scan.

Imaging Findings: Patient referred for plain & contrast enhanced MR imaging:

**T2Wi & FLAIR axial:** Diffuse lesion showing hyperintense signal on T2Wi & FLAIR in the deep periventricular white matter around right occipital horn, splenium of corpus callosum on both sides, subcortical white matter of right temporal and parietal lobes. It is seen to extend to lateral part of right thalamus and cerebral peduncle, right external and internal capsules.

**T1Wi Sag:** The splenium of corpus callosum is thickened.

**Post contrast T1Wi:** No contrast uptake is seen in the affected area.* Contrast uptake indicates worse prognosis.
MRI findings revealed:

- Poorly defined, diffuse lesion showing altered signal intensity in the deep periventricular white matter around right occipital horn, splenium of corpus callosum on both sides, subcortical white matter of right temporal and parietal lobes. It was also seen to extend to lateral part of right thalamus and cerebral peduncle, right external and internal capsules. Diffuse altered signal intensity was also seen in the dentate nuclei on both sides and in the dorsal part of the pons.

- The splenium of corpus callosum and involved deep white matter was expanded.

- No enhancement seen in these regions on post contrast study.

- No evidence of hemorrhage or calcification.

- Evidence of mass effect noted on right lateral ventricle with midline shift to left by 1.1 cm. No hydrocephalus.

**Imaging Diagnosis:** Findings compatible with Gliomatosis Cerebri.

**Discussion:**

Gliomatosis cerebri (GC) is a rare form of the commonest brain tumour, namely the glioma. The term was first used by Nevin in 1938. The WHO classification of brain tumours recognizes GC as a distinct clinicopathological entity among the neuroepithelial tumours of uncertain origin; the criterion for diagnosis is considered to be involvement of at least two lobes of the brain by small elongated cells without a cellular, centrally necrotic centre.

The incidence of GC is greatest in the third to the fifth decades. The disease is usually slowly progressive, although its clinical duration can vary from a few weeks to more than 20 years. Nonspecific personality and mental changes are the most frequent clinical manifestations, and clinical findings are disproportionate to the extent of the brain involvement, which may relate to the diffuse but generally nondestructive nature of the disease process. GC has a poor prognosis, and in the long term, no treatment has been proved effective.
MRI is significantly more sensitive than CT in both the diagnosis of GC and in determining its true extent. CT studies in these patients generally show, iso- to hypodense lesions, with a more or less diffuse mass effect, and minimal or no contrast enhancement; a definable mass is not present. Lesions that appear subtle or are not apparent on CT may be identified by MRI, as poorly defined iso- or hypointense areas on T1-weighted images, and hyperintense areas on T2-weighted images. Abnormalities are frequently observed in the basal ganglia, thalami and the hypothalamus, usually with infiltration of the process along the anatomic white matter pathways. Commissural structures, such as the corpus callosum, are frequently affected and expanded. Subpial and cortical extension of the disease may also be observed. Hyperintensity on T2-weighted images reflects tumor infiltration, but may also represent secondary destruction of myelin fibres which is typically observed in the white matter, corpus callosum, basal ganglia and thalamus, but is rare in the brainstem, cerebellum, and spinal cord. Owing to the infiltrative nature of GC, blood supply of the tumor is by pre-existing cerebral vessel, not requiring the formation of new vessels. Almost no angiogenesis is observed in GC, and this fact is demonstrated by analysis of the tumor microvasculature, which maintains the immunohistochemical characteristics of the blood-brain barrier. As a consequence, the tumor generally does not take up contrast. The presence of contrast uptake may suggest worse prognosis.

D/D:

Multiple sclerosis- Multiple lesions in typical location that often enhance and show no mass effect.
Leukodystrophy- Confluent periventricular white matter T2 hyperintensity.
Ischaemic lesions- No mass effect. Spares cortex. Associated with atrophy.
Inflammatory or infectious processes- Acute presentation. +/- Meningeal involvement.
Lymphoma- Intense enhancement. Iso/hypo intense on T2Wi.

Regards,

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N.B: This case is authentic and from the archives of Radiance Diagnostics. For any queries / suggestions/feedback write to us at radiance@radiancediagnostics.in