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A Case Report on: GANGLIOGLIOMA

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Abstract:- Gangliogliomas are rare tumors of the central nervous system(1-1.5% of all primary brain tumors). They can occur anywhere in the central nervous system but are most commonly located in the temporal lobe(75%) and are mainly found in children. A 50-year-old male presented with features of headache,dizziness and altered behaviour. plain CT images shows large intra-axial predominantly cystic lesion with eccentric hyperdense solid component without calcification. Contrast-enhanced magnetic resonance imaging (CEMRI) of the brain showed well defined intense and heterogenously enhancing solid and cystic mass lesion in the left high frontal region with mass effect and midline shift.

We report a case of ganglioglioma in 50 years old male in the left frontal lobe (10%),which is rare location which was diagnosed by CT, CEMRI and confirmed at histopathology.

Keywords:- *Ganglioglioma, frontal lobe, cortical based tumor.*

I. INTRODUCTION

A 50-year-old male presented with features of headache,dizziness and altered behaviour for 3 month, which aggrevated for the past 10 days. Systemic examination was within normal limits. GCS was E5V5M5, pupils were bilateral normal in size and reacting. No history of fever and seizures. No history of motor, sensory deficits. Heterogenously enhancing solid cystic mass lesion measuring approximately 5.7×5.2x4.7 cm in the left high frontal lobe. Plain CT images show large intra-axial predominantly cystic lesion with eccentric hyperdense solid component within the left high frontal lobe.There is significant perilesional vasogenic edema.Mass effect in the form of effacement of left lateral ventricle and midline shift towards right side. Contrast-enhanced magnetic resonance imaging (CEMRI) of the brain showed the lesion appears hypointense on T1WI and hyperintense on T2/T2 FLAIR images with dependent fluid-fluid level. S/o Intracystic hemorrhage(rare).

The eccentric solid component within the lesion abuts the adjacent dura and shows multifocal T1W1 hyperintensities and predominantly hyperintense on T2/T2 FLAIR images with incomplete T2WI/ FLAIR hypointense rim. No significant diffusion restriction noted within the lesion. Significant blooming noted in the entire solid component on SWI images with fluid-fluid level.

Based on the Imaging findings, the following differentials for a cortical based tumor were considered-

- Oligodendroglioma
- Pilocytic astrocytoma
- Ganglioglioma
- Pleomorphic xanthoastrocytoma

Intraoperatively, a cystic mass lesion with reddish brown nodule was seen in the left frontal lobe. Complete tumor excision was done. Histopathology revealed ganglioglioma.

Histopathology -Shows fragments of tissue with large areas of hemorrhage and congested vascular channels. Moderately cellular areas shows glial and neuronal elements. These areas shows round to oval cells interspersed with ganglion cells.These cells are round to polygonal, focal clustering and binucleated. No microvascular proliferation and necrosis seen.Mitoses are very occasional. Dural tissue identified. Admixed mild lymphomononuclear infiltrate seen. Focally eosinophilic granular bodies identified.

II. IMMUNOHISTOCHEMISTRY

*Chromogranin: Positive in few ganglion cells

*CD 34 : Negative in ganglion cells-highlights vascular endothelial cells.

*Ki 67 : 5%

Idh –not done

The immediate post-operative period was uneventful.

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Fig. 1 Fig. 2 Fig. 3

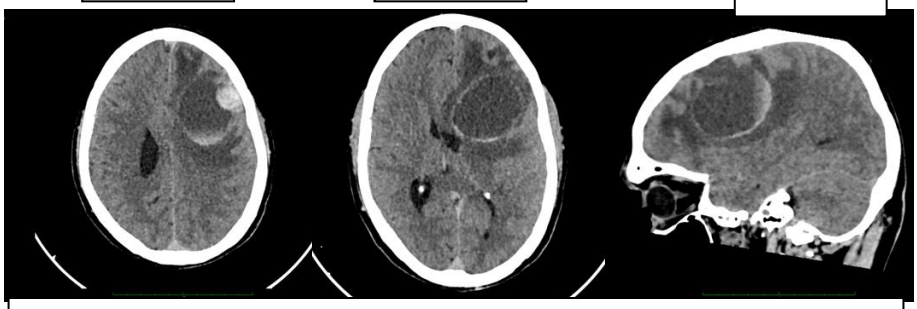

FIG 1-AXIAL NCCT IMAGE SHOWS LARGE INTRA-AXIALMIXED SOLID AND CYSTIC LESION, PREDOMINANTLY CYSTIC LESION WITH ECCENTRIC HYPERDENSE SOLID COMPONENT WITHIN THE LEFT POSTERIOR FRONTAL LOBE.

FIG 2 &FIG 3-AXIAL AND SAGITTAL NCCT IMAGES SHOWS THERE IS SIGNIFICANT PERILESIONAL EDEMA AND MASS EFFECT IN THE FORM OF EFFACEMENT OF LEFT LATERAL VENTRICLE AND MIDLINE SHIFT TOWARDS RIGHT SIDE.

Fig. 4 Fig. 5 Fig. 6

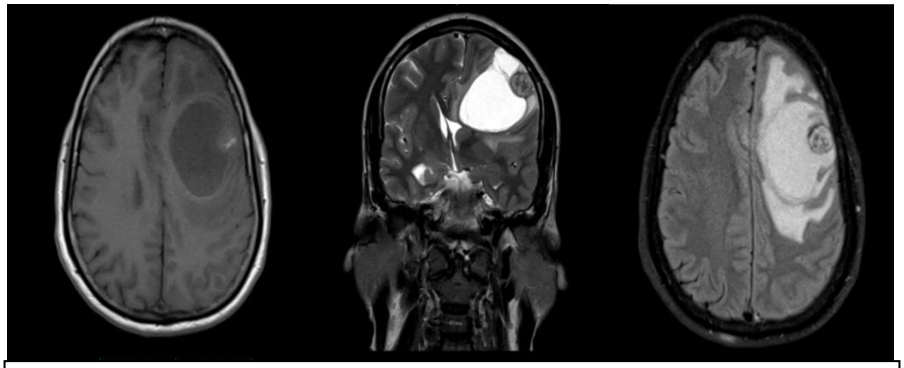

FIG 4- PLAIN AXIAL T1WI IMAGE SHOWS PREDOMINANTLY HYPOINTENSE CYSTIC LESION WITH ECCENTRIC HYPERINTENSE SOLID COMPONENT.

FIG 5- ON PLAIN CORONAL T2WI IMAGE SHOWS THE CYSTIC COMPONENT APPEARS HYPERINTENSE WITH ECCENTRIC HYPOINTENSE SOLID COMPONENT, WHICH IS ABUTS THE ADJACENT DURA. THERE IS MASS EFFECT SEEN TOWARDS THE RIGHT SIDE IN THE FORM OF EFFACEMENT OF LEFT LATERAL VENTRICLE.

FIG 6- ON AXIAL T2 FLAIR IMAGE SHOWS CYSTIC COMPONENT OF THE LESION SHOWS INCOMPLETE SEPARATION WITH HETEROGENOUSLY HYPOINTENSE SOLID COMPONENT. THERE IS SIGNIFICANT HYPERINTENSE SURROUNDING VASOGENIC EDEMA SEEN.

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Fig. 7

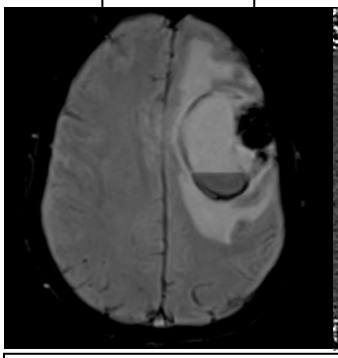

Fig. 8

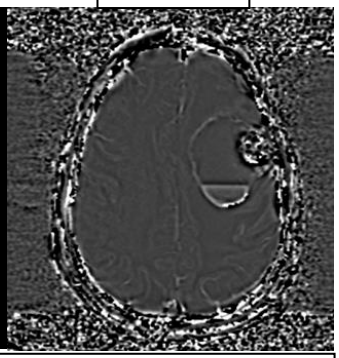

FIG 7 ON SWI IMAGE,THE SOLID COMPONENT SHOWS BLOOMING.

FIG 8 ON PHASE CONTAST IMAGE SOLID COMPONENT APPEARS HETEROGENOUSLY HYPERINTENSE SHOWS SIGNIFICANT BLOOMING NOTED IN THE ENTIRE SOLID COMPONENT.

Fig. 9

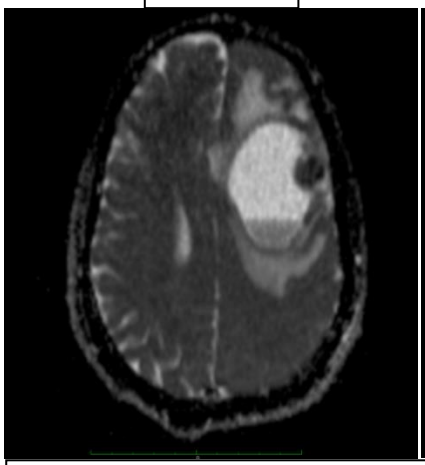

Fig. 10

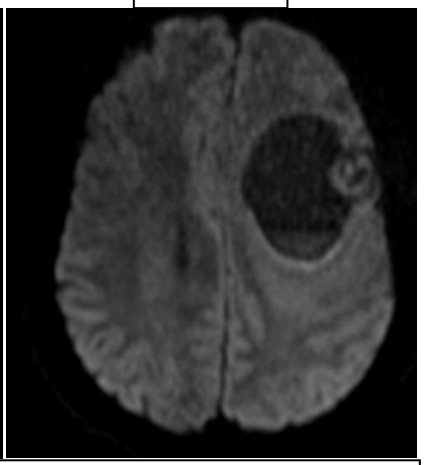

FIG. 9 & FIG. 10: ON DWI AND ADC THERE IS NO SIGNIFICANT DIFFUSION RESTRICTION NOTED WITH IN THE LESION.

Fig. 11 Fig. 12 Fig. 13

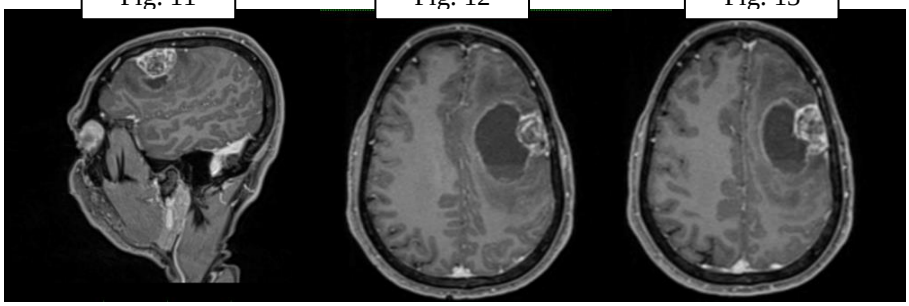

FIG 11 & FIG 12 -ON POST CONTRAST SAGITTAL AND AXIAL T1WI HETEROGENEOUS ENHANCEMENT IS SEEN WITHIN THE SOLID COMPONENT.

NO ENHANCEMENT IS SEEN WITHIN THE CYSTIC COMPONENT.

III. DISCUSSION

Ganglioglioma is an rare tumor of the central nervous system.ganglioglioma is a well differentiated,slow growing tumor composed of dysplastic ganglion cells and neoplastic glial cells. The age of presentation of these tumors varies from 2 months to 70 years.predominantly a tumor of childen and young adults;80% of patients are younger than 30 years.peak presentation is 15-20 years. The majority of gangliogliomas occur in the temporal lobe (> 70%) [7], The next most common site is the frontal lobe(10%) . The most common presenting signs and symptoms are seizures (temporal lobe and other supratentorial locations), followed by headache, dizziness, ataxia (posterior fossa), and progressive weakness (spinal cord). The typical of ganglioglioma is a benign, calcified tumor in the temporal lobe of a child with seizures [3]. On CT, the picture is of a circumscribed solid mass or cyst with a mural nodule. On MR imaging, gangliogliomas are isointense to hypointense on T1- weighted images, are hyperintense and heterogeneous on T2-weighted images, and can contain solid, cystic, and calcified components. Enhancement after administration of gadolinium has also been found to vary from no enhancement to marked, heterogeneous enhancement [2, 12]. Histopathologically, gangliogliomas are benign, well differentiated neuroepithelial tumors. The typical feature is a combination of neuronal and glial cell elements, which may exhibit marked heterogeneity [13]. On IHC, the glial component is positive for GFAP, S-100 protein, and vimentin, whereas the neuronal component is reactive for synaptophysin, MAP 2, NeuN, and neurofilaments [14, 15]. Histopathologic differential diagnoses comprise both high-grade and low-grade neoplasms, such as diffuse astrocytomas, oligodendrogliomas, dysembryoplastic neuroepithelial tumors, pilocytic astrocytomas (PAs), and pleomorphic xanthoastrocytomas (PXAs) [7]. Mitotic figures in ganglioglioma are rare, and MIB index varies from 1.1 to 2.7%. CD 34 is present 70–80% in the neuronal component of ganglioglioma but is less common in anaplastic variants [15]. IDH1 and P53 should be done to rule out diffuse glioma. Anaplastic gangliogliomas typically demonstrate malignant transformation of the glial component with hypercellularity, vascular proliferation, and necrosis high mitotic labeling indices (Ki-67) [16–18]. Anaplastic transformation is more common in the pediatric population and has been associated with previous subtotal tumor resection and radiotherapy [19]. Gross total resection should be attempted whenever possible with the preservation of neurological function. Some studies have advocated the role of gross total resection along with adjuvant radiochemotherapy in achieving good survival rates [20, 21].

IV. CONCLUSION

Gangliogliomas are rare tumors with frontal lobe as rare location. Imaging may vary, and histology along with IHC is needed for the diagnosis. Gross total resection along with adjuvant therapy improves outcome.

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