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Case Report

Anaplastic oligodendroglioma WHO grade 3: Rare brain tumor

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ABSTRACT

The anaplastic oligodendrogliomas are uncommon gliomas. It is clinically significant to know this entity as they have unique clinical, histopathological and molecular features. We present a 43 year female having complaint of headache, tingling numbness, giddiness. Which was of 2 year duration, off and on, and was increased in last 15 days. MRI brain (plain + contrast) showed a large heterogenously enhancing cortical based solid-cystic lesion with surrounding edema in left frontal lobe with mass effects and midline shift. On radiological features impression was suggestive of high grade glial neoplasm and advised biopsy. In our case on histopathology reported as anaplastic oligodendrogliomas WHO CNS grade 3 of left frontal lobe. Patient was treated with surgery, and chemotherapy. We are presenting this rare case of anaplastic oligodendrogliomas for it's clinical, Histopathological and radiological findings.

1. Introduction

Oligodendroglioma is an infiltrating glial tumor composed of cells resembling oligodendrocytes. The oligodendrogliomas differ from other glial tumors on clinical presentation,natural history,histopathological findings, and molecular pathogenesis. Recently various molecular pathogenesis and biomarkers for oligodendroglioma are studied which includes non-balanced translocation leading to 1p/19q codeletion, promoter hypermethylation of the MGMT gene, mutations of the IDH1/ IDH2 gene, and mutations of FUBP1 (on 1p) or CIC (on 19q).[1,2]

The anaplastic oligodendroglioma is a malignant brain tumor for which a treatment recommendations differ considerably. Anaplastic oligodendroglioma altogether sensitive to treatment with both chemotherapy and radiotherapy. For even in WHO grade 3 oligodendroglioma patients, the current standard treatment of surgery, radiotherapy and chemotherapy has increased overall median survival times.

2. Case Report

We present a 43-year female having complaint of headache, tingling numbness, giddiness. Which was of 2 year duration, off and on, and was increased last15 days. She had history of head injury 10year back. There was no history hypertension, epilepsy. The other systemic examination was normal.

Her MRI brain (plain + contrast) showed a large ill-defined multiloculated cortical based solid-cystic mass lesion measuring 6.3 × 5.6 × 4.6 cm, in left frontal region of brain (Figures 1 and 2). On signal characteristic showed a larger cystic component: appeared hypointense on T1WI and heterogenously hyperintense on T2WI with suppression on FLAIR and shows no diffusion restriction

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or blooming with homogenous enhancement of walls. Solid component appeared to hypointense on TWI and heterogenously hyperintense on T2WI/FLAIR with areas of cystic changes within. It showed patchy areas of mild diffusion restriction and multifocal peripheral and central areas of blooming on GRE (calcifications - confirmed on CT). On post-contrast T1WI, it showed heterogenous, predominantly peripheral enhancement with central non-enhancing areas. Surrounding non-enhancing T2WI hyper intense signal was noted extending into left frontal and temporal lobes suggestive of perilesional edema. The lesion was causing effacement of adjacent sulcal spaces and mass effect on left lateral ventricle leading to subfalcine herniation with focal midline shift measuring 10 mm towards right side. MR spectroscopy reveals increased choline peak and reduced NAA peak. Rest of the cortical sulcal spaces, basal cisterns and ventricular system appeared normal. Rests of the white matter and bilateral thalami appeared normal. Cerebellar peduncles appeaedr normal. Brainstem showed normal morphology and signal intensity. Pituitary gland, infundibular stalk, sella and parasellar structures were normal. The visualised parts of VII-VIII nerve complexes appeared normal in morphology and signal intensities bilaterally. Intracranial vessels and dural sinuses showed expected flow voids. On this findings impression given was large heterogenously enhancing cortical based solid-cystic lesion with surrounding edema in left frontal lobe with mass effects and midline shift and imaging characteristics as features suggestive of high grade glial neoplasm. The biopsy was advised and its correlation.

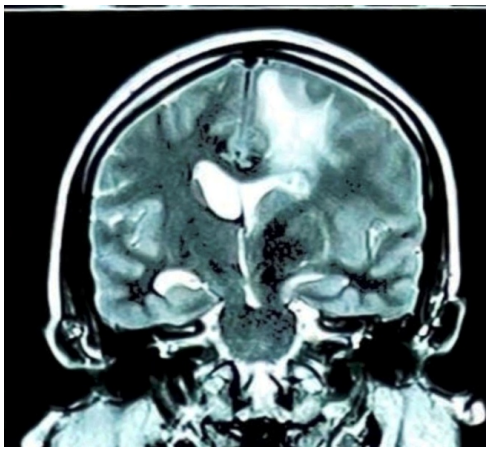

Fig. 1: MRI brain shows a large mass in left frontal region of brain.

On microscopic features tumor was cellular with diffusely growing pattern with high cellularity. Tumor cells were round with distinct cell borders and having round nuclei with small prominent nucleoli and with open bland chromatin. Tumor cells shows moderate nuclear atypia. Tumor showed increased mitotic activity > 6 mitoses per 10 HPF. The clear cytoplasm gives perinuclear

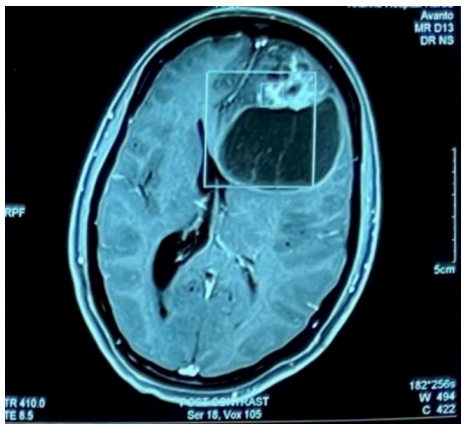

Fig. 2: MRI brain shows a large ill-defined multiloculated cortical based solid-cystic mass lesion measuring 6.3 × 5.6 × 4.6 cm, in left frontal region of brain.

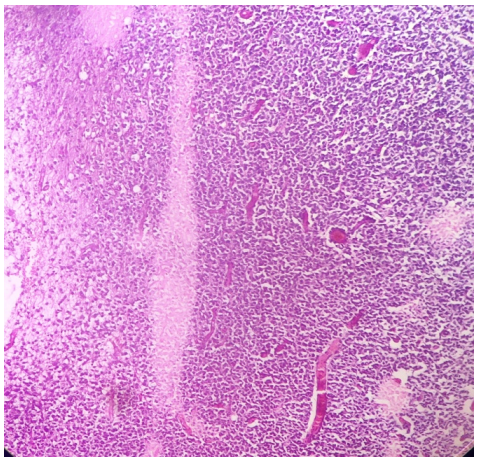

Fig. 3: Oligodendroglioma -monotonous tumor cells showing round nuclei with perinuclear halo (Haematoxyline & Eosin, x40).

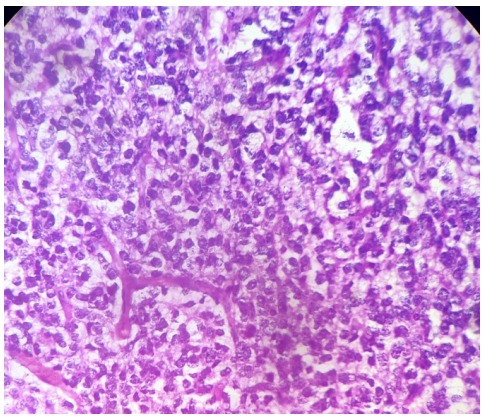

Fig. 4: Oligodendroglioma –tumor cells having round nuclei with perinuclear halo, cellular atypia and thin chicken wire like vasculature is noted.(Haematoxyline & Eosin, x100).

halos to form “fried egg” appearance ($). Tumor shows delicate vessels. Areas of microcalcifications were noted. In our case on histopathology reported as anaplastic oligodendrogliomas WHO CNS grade 3 of left frontal lobe. Patient was treated with surgery, and chemotherapy.

3. Discussion

Oligodendroglial tumors are the uncommon glial tumors. Oligodendrogliomas represent approximately 5-17% of intracranial gliomas. While anaplastic oligodendroglioma is comprising 0.5% of all intracranial neoplasms.[3] The oligodendrogliomas are located in cerebral hemispheres, mostly at frontal lobe which constitute about 50-60%, followed by parietal and temporal lobes.[3] In children these tumors are noted at an unusual sites such as the posterior fossa and the spinal cord. The seizures are the common clinical presentation in 2/3 of all cases when located at cerebral cortex. The other menifestations are signs and symptoms of increased intracranial pressure, cognitive changes, focal neurologic decifits, or hemiparesis.[4]These are incidentaly detected in 10% cases.

On neuroimaging: magnetic resonance imaging, oligodendrogliomas shows enhancing, occasionally well-circumscribed mass lesions. They have hyperintense or mixed signal on T2-weighted and fluid-attenuated inversion recovery (FLAIR) images. The focal Calcifications may be noted.[5]

On gross morphology these tumors are gray white, where as highly cellular grade 3 tumors are fleshy masses, but with out central areas of necrosis as seen in glioblastomas.

The World Health Organization (WHO) classification of central nervous system tumors, oligodendroglial tumors are defined by the molecular analysis of IDH1/2 hotspots and LOH1p/19q testing. Accordingly the oligodendroglioma are graded as, IDH-mutant and 1p/19q-codeleted, WHO CNS Grad 2, while oligodendroglioma, IDH-mutant and 1p/19q-codeleted, WHO CNS Grade 3 tumor.[6]

Oligodendroglioma NOS (not otherwise specified) suffix was introduced in the 2016 WHO classification for cases in which molecular information is not available or is insufficient for a more specific diagnosis. Without genetic testing for IDH-1/2 and LOH 1p/19q, the tumor is called oligodendroglioma, NOS.[2]

On microscopic features tumor is cellular with diffusely growing pattern or may shows circumscribed nodules with high cellularity. Tumor cells are round with distinct cell borders and having round nuclei with small prominent nucleoli and with open bland chromatin. Tumor cells shows moderate-to-marked nuclear atypia. The clear cytoplasm gives perinuclear halos to form “fried egg” appearance. Tumor may shows delicate, acutely branched capillary sized vessels - "chicken-wire" like appearance. Areas of microcalcifications are noted. In few cases mucin rich microcystic spaces, perineuronal satellitosis are findings.[7]

The oligodendrogliomas on histologic grades are mostly WHO CNS grade1or 2 are low grade. The WHO CNS 3 are considered as anaplastic oligodendrogliomas.

In our case it was anaplastic oligodendrogliomas WHO CNS grade 3.

In case of anaplastic (WHO grade 3) criterias are along with above microscopic features, tumor shows increased mitotic activity >= 6 mitoses per 10 HPF, microvacular proliferation i.e., marked hypertrophy and hyperplasia of endothelial cells and pericytes resulting in a multilayered aspect of the microvessel walls, and presence of necrosis. Occasional multnucleated cells are noted.[8]

On immunohistochemistry tumor cells are positive for Olig 2, GFAP, Neu N EMA, MAP2, while the GFAP is variably positive.[9]

The differential diagnosis of tumors with oligo-like morphology are diffuse astrocytoma, clear cell ependymoma, central neurocytoma, dysembryoplastic neuro epithelial tumor, metastatic clear cell carcinomas, pleomorphic xanthoastrocytoma.[10,11] This differentiation more difficult on small stereotactic biopsies.

The microscopic features for diffuse astrocytoma, these tumor shows oval to elongated, cigar shaped nuclei with variable cytoplasm. The tumor cells are in fibrillary background. In clear cell ependymoma on histolopathological findings, these tumor shows perivascular pseudorosette of surrounding brain and nuclear groove.

The current management of anaplastic oligodendrogliomas is a combination of surgery, radiotherapy and chemotherapy. In our patient surgical excision of tumor was done and adjuvant chemotherapy was given.

Cairncross JG et al, noted that patients with anaplastic oligodendroglioma, adjuvant chemotherapy with PCV (procarbazine [Matulane], lomustine [CeeNU], and vincristine) following standard radiation therapy delayed disease progression and increased survival.[12] The current standard treatment of surgery, radiotherapy and chemotherapy has increased overall median survival in anaplastic oligodendroglioma cases also.

4. Conclusions

The oligodendrogliomas differ from other glial tumors on clinical presentation, natural history, histopathological findings, and molecular pathogenesis. We present an uncommon glioma: Anaplastic oligodendrogliomas WHO CNS grade 3, a rare case for its clinical, histopathological and radiological findings. These findings have important prognostic implications for better treatment and patient care.

5. Conflicts of interest

There are no conflicts of interest.

6. Source of Funding

None.

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