

Content available at: <https://www.ipinnovative.com/open-access-journals>

IP Archives of Cytology and Histopathology Research

Journal homepage: <https://www.achr.co.in/>

## Case Report

# Anaplastic oligodendroglioma WHO grade 3: Rare brain tumor

Sunil Vitthalrao Jagtap<sup>1,\*</sup>, Shubham S Jagtap<sup>2</sup>, Neha Ghadge<sup>1</sup>, Dhvani Mavani<sup>1</sup>,  
Nidhi Goswami<sup>1</sup>

<sup>1</sup>Dept. of Pathology, Krishna Institute of Medical Sciences Deemed University, Karad, Maharashtra, India

<sup>2</sup>Dept. of Medicine, Krishna Institute of Medical Sciences Deemed University, Karad, Maharashtra, India



### ARTICLE INFO

#### Article history:

Received 17-04-2023

Accepted 22-06-2023

Available online 27-07-2023

#### Keywords:

Oligodendroglioma

CNS grade 3 tumors

Histopathology

Anaplastic oligodendrogliomas

### 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 heterogeneously 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 its clinical, Histopathological and radiological findings.

This is an Open Access (OA) journal, and articles are distributed under the terms of the [Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License](https://creativecommons.org/licenses/by-nc-sa/4.0/), which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: [reprint@ipinnovative.com](mailto:reprint@ipinnovative.com)

## 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).<sup>1,2</sup>

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

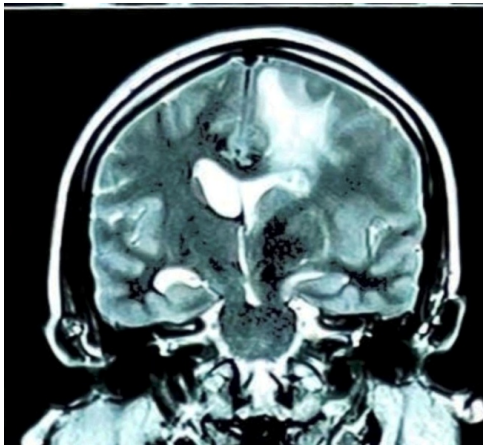
We present a 43-year female having complaint of headache, tingling numbness, giddiness. Which was of 2 year duration, off and on, and was increased last 15 days. She had history of head injury 10 year back. There was no history hypertension, epilepsy. The other systemic examination was normal.

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 heterogeneously hyperintense on T2WI with suppression on FLAIR and shows no diffusion restriction

\* Corresponding author.

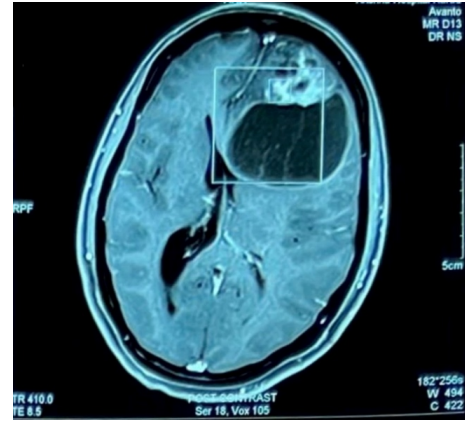
E-mail address: [drsvjagtap@gmail.com](mailto:drsvjagtap@gmail.com) (S. V. Jagtap).

or blooming with homogenous enhancement of walls. Solid component appeared to hypointense on TWI and heterogeneously 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 hyperintense 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 appeared 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 heterogeneously 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 gliar neoplasm. The biopsy was advised and its correlation.

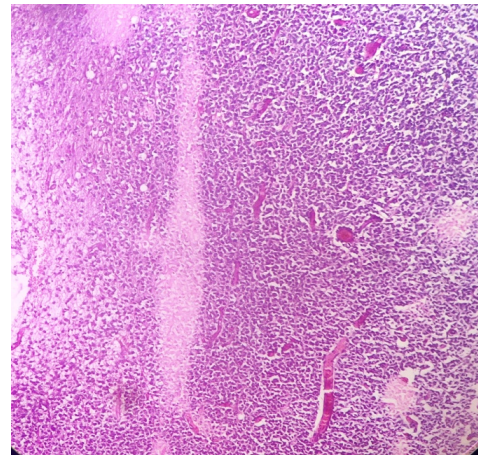


**Fig. 1:** MRI brain shows a large mass in left frontal region of brain.

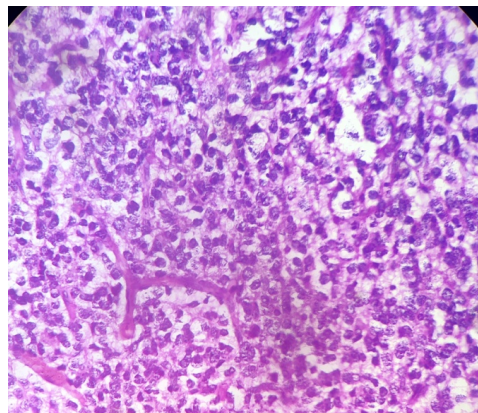
On microscopic features tumor was cellular with diffusely growing pattern with high cellularity. Tumor cells were round with distinct cell borders and having round nuclei with small prominent nucleoli and with open bland chromatin. Tumor cells shows moderate nuclear atypia. Tumor showed increased mitotic 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.<sup>3</sup> The oligodendrogliomas are located in cerebral hemispheres, mostly at frontal lobe which constitute about 50-60%, followed by parietal and temporal lobes.<sup>3</sup> 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 manifestations are signs and symptoms of increased intracranial pressure, cognitive changes, focal neurologic deficits, or hemiparesis.<sup>4</sup> These are incidentally 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.<sup>5</sup>

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.<sup>6</sup>

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.<sup>2</sup>

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.<sup>7</sup>

The oligodendrogliomas on histologic grades are mostly WHO CNS grade 1 or 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) criteria are along with above microscopic features, tumor shows increased mitotic activity  $\geq 6$  mitoses per 10 HPF, microvascular 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 multinucleated cells are noted.<sup>8</sup>

On immunohistochemistry tumor cells are positive for Olig 2, GFAP, Neu N EMA, MAP2, while the GFAP is variably positive.<sup>9</sup>

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.<sup>10,11</sup> 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 histopathological 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.<sup>12</sup> 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.

## References

1. Wesseling P, Van Den Bent M, Perry A. Oligodendroglioma: pathology, molecular mechanisms and markers. *Acta Neuropathol.* 2015;129(6):809–27.
2. Louis DN, Perry A, Reifenberger G, Deimling A, Figarella-Branger D, Cavenee W, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol.* 2016;131(6):803–20. doi:10.1007/s00401-016-1545-1.
3. Ostrom QT, Patil N, Cioffi G, Waite K, Kruchko C. CBTRUS Statistical Report: Primary Brain and Other Central Nervous System Tumors Diagnosed in the United States in 2013-2017. *Neuro Oncol.* 2020;22(12 Suppl 2):iv1–96. doi:10.1093/neuonc/noaa200.
4. Jagtap SV, Rajput SR, Jagtap S. Oligodendroglioma presented as hemiparesis. *Int J Scientific Res.* 2017;6(12):282–3.
5. Smits M. Imaging of oligodendroglioma. *Br J Radiol.* 2016;89(1060):20150857. doi:10.1259/bjr.20150857.
6. International Agency for Research on Cancer. *Classification of Tumours Editorial Board.* 2021;.
7. Komori T. Pathology of oligodendroglioma: An overview. *Neuropathology.* 2017;37(5):465–74. doi:10.1111/neup.12389.
8. Figarella-Branger D, Mokhtari K, Dehais C, Carpentier C, Colin C, Jouvet A, et al. Mitotic index, microvascular proliferation, and necrosis define 3 groups of 1p/19q codeleted anaplastic oligodendrogliomas associated with different genomic alterations. *Neuro Oncol.* 2016;18(6):888–90. doi:10.1093/neuonc/nov085.
9. Wesseling P, Van Den Bent M, Perry A. Oligodendroglioma: pathology, molecular mechanisms and markers. *Acta Neuropathol.* 2015;129(6):809–27.
10. Jagtap SV, Gupta SO, Jokhi CD, Jagtap SS. Pleomorphic xanthoastrocytoma. *Int Surg J.* 2017;4(11):3789–91. doi:10.18203/2349-2902.isj20174910.
11. Van Den Bent M, Chang SM. Grade II and III Oligodendroglioma and Astrocytoma. *Neurol Clin.* 2018;36(3):467–84.
12. Cairncross JG, Wang M, Shaw EG. Chemotherapy plus radiotherapy (CT-RT) versus RT alone for patients with anaplastic oligodendroglioma: Long-term results of the RTOG 9402 phase III study. *J Clin Oncol.* 2008;30(15). doi:https://ascopubs.org/doi/abs/10.1200/jco.2012.30.15\_suppl.2008b.

## Author biography

**Sunil Vitthalrao Jagtap**, Professor  <https://orcid.org/0000-0002-4844-7624>

**Shubham S Jagtap**, Assistant Lecturer  <https://orcid.org/0000-0002-8304-8581>

**Neha Ghadge**, Assistant Lecturer

**Dhwani Mavani**, Assistant Lecturer

**Nidhi Goswami**, Assistant Lecturer  <https://orcid.org/0000-0002-4342-4028>

**Cite this article:** Jagtap SV, Jagtap SS, Ghadge N, Mavani D, Goswami N. Anaplastic oligodendroglioma WHO grade 3: Rare brain tumor. *IP Arch Cytol Histopathology Res* 2023;8(2):112-115.