Case Report:
Third Ventricle Anaplastic Oligodendroglioma: A Case Report

Tamajyoti Ghosh[1*], **Subir Dey**[1]

1. Department of Neurosurgery, Armed Forces Medical College, Pune, India

Use your device to scan and read the article online

Citation Ghosh T, Dey S. Third Ventricle Anaplastic Oligodendroglioma: A Case Report. Iran J Neurosurg. 2021; 7(3):153-158. http://dx.doi.org/10.32598/irjns.7.3.6

doi: http://dx.doi.org/10.32598/irjns.7.3.6

Article info:
Received: 17 May 2021
Accepted: 02 Jun 2021
Available Online: 01 Jul 2021

Keywords:
Oligodendroglioma, Third ventricle, Cerebral ventricle neoplasms

ABSTRACT

Background and Importance: Anaplastic Oligodendroglioma (ODG) constitutes 24% of all pediatric ODG. The mean age of presentation of ODG is 12±6 years. They are most common in frontal and temporal lobes; however, rare cases of intraventricular ODGs are reported. Most commonly they arise from the anterior part of lateral ventricles. Third ventricle ODG is extremely rare and only a few cases of lateral and third ventricle anaplastic ODG are reported. ODGs infiltrate locally to meninges and rarely have leptomeningeal spread. Thus, ODG forms a differential diagnosis of pediatric intraventricular tumor.

Case Presentation: Here we present a case of a 15-month-old male child with raised intracranial pressure due to obstructive hydrocephalus. The patient was detected to be COVID-19 RT–PCR (Reverse Transcriptase Polymerase Chain Reaction) positive in the preoperative period and underwent emergency Right-sided Ventriculo Peritoneal (VP) shunt. His contrast MRI (Magnetic Resonance Imaging) Brain showed a 50×24×39 mm heterogeneously enhancing mass epicenter at third ventricle and extending to lateral and fourth ventricle with spinal drop metastasis. Preoperative differential diagnosis of Ependymoma was made and definitive surgery was done once the child recovered from COVID-19. However, his biopsy specimen pathology and Immunohistochemistry (IHC) were suggestive of anaplastic oligodendrogliomas and the child responded well to chemotherapy.

Conclusion: Intraventricular ODG is an extremely rare pediatric tumor. Patients usually present with obstructive hydrocephalus. Contrast MRI findings are nonspecific and help in detecting leptomeningeal spread to the spine. IHC and chromosomal analysis are important diagnostic and treatment prognostication tools. These tumors have a high recurrence and poor prognosis despite gross total resection.

*** Corresponding Author:**
Tamajyoti Ghosh, MD.
Address: *Department of Neurosurgery, Armed Forces Medical College, Pune, India*
Tel: *+98 (910) 1483164*
E-mail: *tamajyoti@gmail.com*

Highlights

- Anaplastic Oligodendroglioma (ODG) are rare in pediatric age. They are generally parenchymatous in location.
- Only few cases of intraventricular oligodendrogliomas are reported which are observed in adults. Pediatric intraventricular oligodendroglioma is extremely rare.
- Radiologically, our pre-operative diagnosis was ependymoma. The patient underwent gross total resection of the tumor.
- Microscopic examination of specimen supported by IHC and chromosomal analysis accurately diagnosed the tumor as anaplastic ODG.
- The patient was treated with adjuvant chemotherapy. He had recurrence within 6 month.

Plain Language Summary

Oligodendrogliomas (ODG) are rare in pediatric age unlike in adults. They arise from oligodendrocytes which are found in brain parenchyma hence they are typically found in those regions. Sometimes subependymal embryonic nest cells may differentiate and give rise to intraventricular ODGs which are extremely rare and only few case reports are present among adult population. Diagnosis is difficult preoperatively due to non-specific radiological features. With the advent of newer molecular and chromosomal analysis techniques, we are able to detect such tumors more accurately and formulate further plan of management. ODGs have good response to chemotherapy but anaplastic ODGs have poor prognosis and they tend to spread along CSF to spine.

1. Background and Importance

Oligodendroglioma (ODG) constitutes 0.9% to 4% of all brain tumors [1]. It is relatively rare in the pediatric age group constituting 6.5% of cases with a Mean±SD age of 12±6 years [2]. ODG is most common in frontal and temporal lobes; however rare cases of intraventricular ODG have been reported [1, 3, 4]. Most commonly, intraventricular ODG arises from the anterior part of lateral ventricles [5]. ODG in the third ventricle is extremely rare and only a few cases of anaplastic ODG in the lateral and third ventricles have been reported in the literature [6-8]. ODG infiltrates locally to meninges [9] and rarely has leptomeningeal spread [9, 10]. Anaplastic ODG constitutes 24% of all pediatric ODGs [11].

2. Case Presentation

Here we present a case of 15-month-old male child with normal milestones who presented with vomiting, irritability, and gait imbalance for the last two months. The patient was conscious and oriented but irritable. His pupils were bilaterally 3 mm reacting. Fundoscopy was suggestive of grade II papilledema. His neurological examination was within normal limits. His anterior and posterior fontanelles were closed. We evaluated him with contrast (magnetic resonance imaging) brain MRI with whole spine screening. It showed a 50×24×39 mm well-defined lobulated heterogeneously enhancing intraventricular mass epicentered in the third ventricle floor extending to the lateral and fourth ventricle with spinal drop metastasis dorsal 12 levels. Preoperatively, his COVID-19 RT-PCR (Reverse Transcriptase Polymerase Chain Reaction) was positive. Given the raised Intracranial Pressure (ICP) features and the patient's COVID-19 positive status, he underwent right sided medium pressure Ventriculoperitoneal (VP) shunt. The patient's irritability and gait imbalance improved postoperatively. His Cerebrospinal Fluid (CSF) study was normal and CSF tumor markers (Alpha-Fetoprotein [AFP], beta-Human Chorionic Gonadotrophin [beta-HCG], and Lactate Dehydrogenase [LDH]) were negative. The patient was planned for definitive surgery later and underwent a left anterior transcortical approach with gross total tumor excision. Intraoperatively, tumor was greyish red, vascular, soft, and suckable, occupying the third ventricle extending into the lateral ventricle and postero-inferior into the aqueduct of Sylvius. The brain tumor interface was ill-defined. The intraoperative frozen section was suggestive of features similar to ependymoma. A ventriculostomy catheter was placed in the third ventricle to facilitate CSF drainage. After the operation,

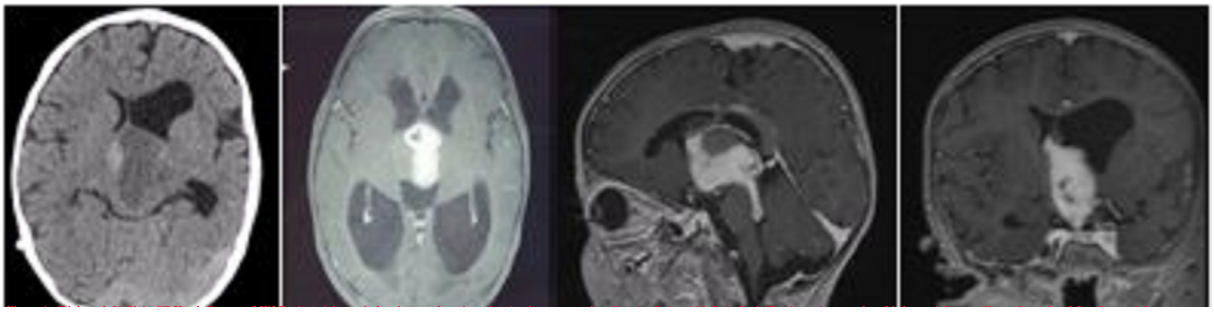

Figure 1. Plain CT scan head, contrast MRI brain axial, sagittal and coronal sections showing homogenously enhancing lesion with foci of calcification epicentered at third ventricle extending to lateral and fourth ventricle (Right to left)

the patient developed right-sided hemiparesis. He was extubated on Postoperative Day (POD) 2. The External ventricular Drain (EVD) was removed on POD 4. There were no features of diabetes insipidus. Post-operative contrast MRI showed gross total excision of the tumor with minimal residual in sella and no infarct. The patient developed multiple spikes of fever along with tachycardia from POD 7. On POD 10, the patient developed irritability, dystonic movements of the right upper and lower limb. Then we started injection of benztropine 1 mg for the patient twice daily after ruling out meningitis, to which he responded well. Histopathological Examination (HPE) suggested anaplastic oigodendroglioma- Glial Fibrillary Acidic Protein (GFAP) positive, olig-2 positive, INI1 retained, Isocitrate Dehydrogenase 1 (IDH1) mutant, synaptophysin positive, chromogranin negative, Epithelial Membrane Antigen (EMA) focal membranous positive, Ki67 20%, S100 positive, Alpha Thalassemia/ mental Retardation X (ATRX) retained, P53 non-mutant, and 1p13q co-deletion positive. Histone gene and BRAF mutation were negative. The patient was given postoperative chemotherapy after 4 weeks comprising lomus-

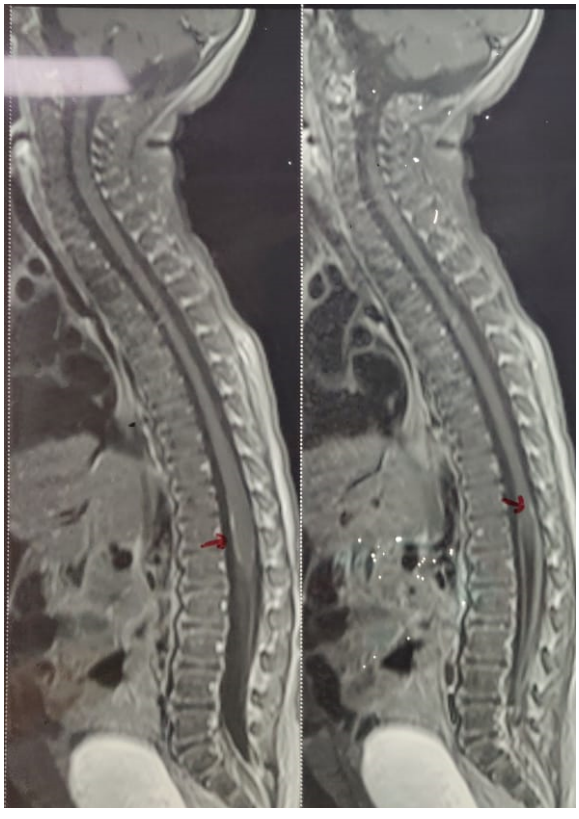

Figure 2. Contrast MRI screening whole spine shows epidural metastatic deposits at dorsal vertebra 12

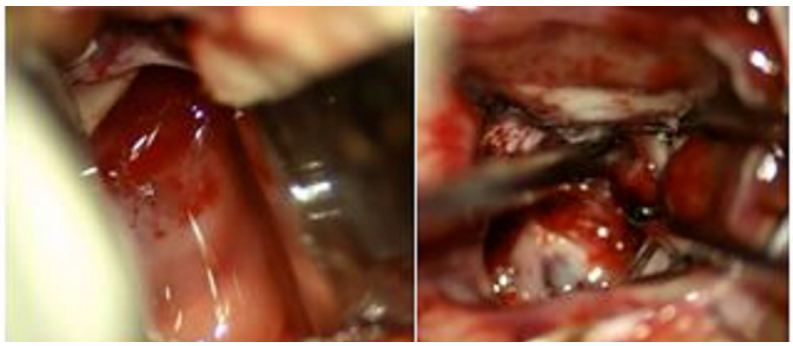

Figure 3. Intraoperative microscopic view of the tumor (Right to left)

View of third ventricle after complete tumor excision

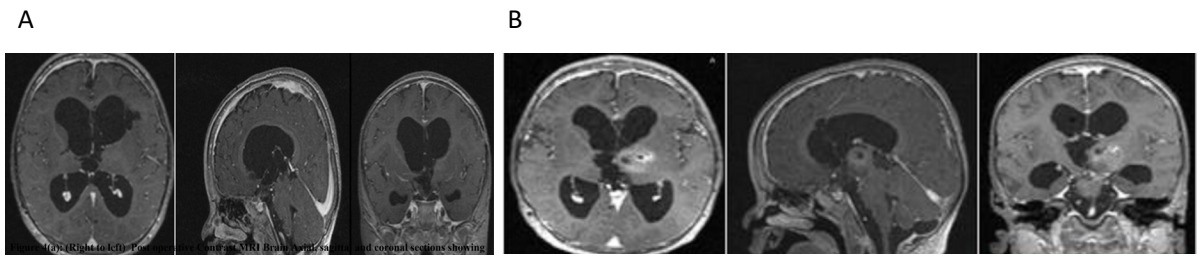

Figure 4. MRI brain axial

A: Postoperative contrast MRI brain axial, sagittal and coronal sections showing no residual lesion in postoperative site (Right to left)

B: Six-month postoperative contrast MRI brain axial, sagittal and coronal sections showing no residual lesion in postoperative site with heterogeneously enhancing lesion in left thalamus and subthalamus (Right to left).

tine 110 mg/m^2 orally on day 1 followed by procarbazine 60 mg/m^2 orally from day 8 to day 21 along with the injection of vincristine 1.4 mg/m^2 intravenously on day 8 and day 29 (a total of 4 cycles). The patient tolerated chemotherapy well. On follow-up, the patient was conscious and oriented. However, there is a regression of developmental milestones. His six-month follow-up contrast MRI shows no residual tumor but a new heterogeneously contrast-enhancing lesion in the left thalamic and subthalamic region, which can explain his dystonic movements during the postoperative period (Figures 1, 2, 3, 4, 5, 6, and 7).

3. Discussion

Anaplastic oligodendroglioma is more common in the older age group [12]. Its five-year median survival period is 20% [13]. The intraventricular location of ODG is an extremely rare entity. In his literature review of 33 studies involving seventy patients, Zada et al. mentioned only 3 patients of anaplastic intraventricular ODG [1]. Similarly, Sherif M. Elwatidy et al. in their analysis of 42 cases of the lateral and third ventricle tumors reported only a single case of ventricular anaplastic oligodendroglioma in adults [6]. Few isolated case reports of intra-

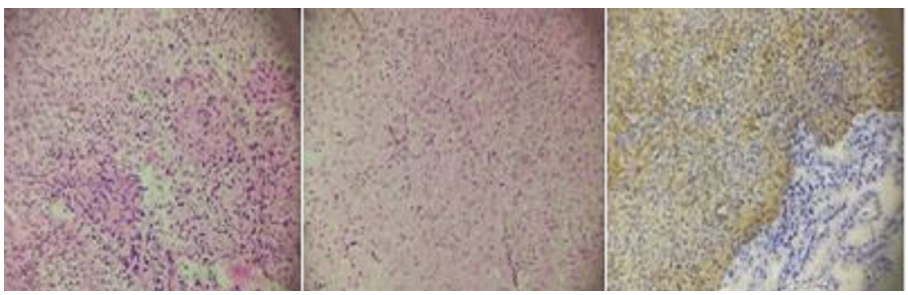

Figure 5. Hematoxylin & Eosin (H & E) showing characteristic chicken wire pattern and EMA focally positive on Immunohistochemistry (IHC) (Right to left)

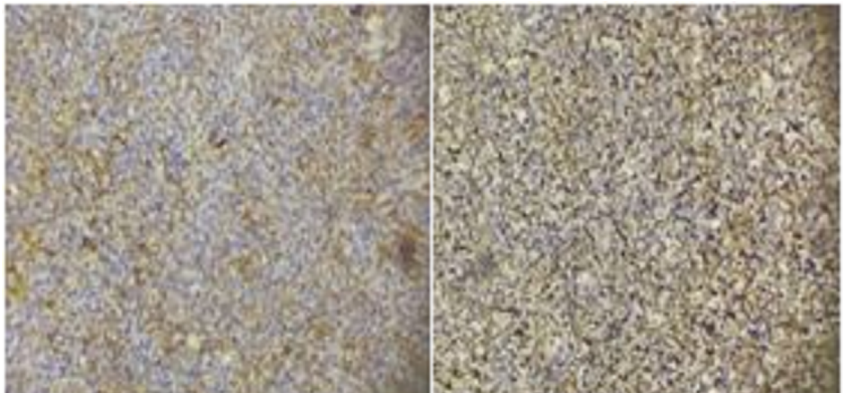

Figure 6. IHC showing synaptosin and GFAF positive (Right to left)

ventricular oligodendrogliomas exist in the adult population [7, 8]. No case report was present in the pediatric age group. High-grade ODG is known for its recurrences. Packer and colleagues [13] mentioned a young adult with a high- grade ODG in the fourth ventricle who developed a frontal lesion 21 months after surgery.

Similarly Zada et al. also reported a 22-year-old female who underwent gross total resection of lateral ventricle anaplastic ODG extending to the third ventricle and 21 days later was readmitted with recurrence on the same site [1]. Unlike lobar ODG, where a patient presents with seizure, intraventricular ODG presents with raised ICP features [1, 3-5, 8]. Histology of intraventricular oligodendroglioma is consistent with parenchymal location. [3, 5]. Our case showed features of a high- grade lesion such as vascular proliferation and focal necrosis. On Immunohistochemistry (IHC), the neoplasm was Epithelial Membrane Antigen (EMA) focally positive, and chromogranin A negative. CSF was negative for tumor markers, thereby ruling out other common pediatric intraventricular neoplasms such as ependymoma, germinoma and lymphoma. IHC is important as anaplastic ODG shares similar H&E features with other tumors like central neurocytoma and ependymoma [14]. Moreover, most of the case reports of intraventricular ODG were from the pre-IHC era; thus the possibility of other lesions being diagnosed as ODG could not be ruled out. The transcortical intraventricular approach is the most common surgery seen among intraventricular anaplastic ODG by Zada et al., and all 3 cases had postoperative recurrences and died [1]. In his study of 200 cases, Shaw et al. found CSF dissemination in 14 out of 165 cases of oligodendroglioma, but no other details were provided [15]. Beck and Russel in 1942 mentioned a case of a 29-year-old male with a left posterior ventricular tumor with diffuse spread over the spinal cord [10]. Pitt et al. reported a case of the fourth ventricle oligodendroglioma with spread along the whole length of the spinal cord up to the midbrain [16]. Natale et al. mentioned a 67-year-old male with intraventricular ODG presenting with cauda equina [17].

4. Conclusion

Intraventricular anaplastic oligodendroglioma is an extremely rare tumor in pediatric age, and our case is probably the only documented pediatric case. Patients

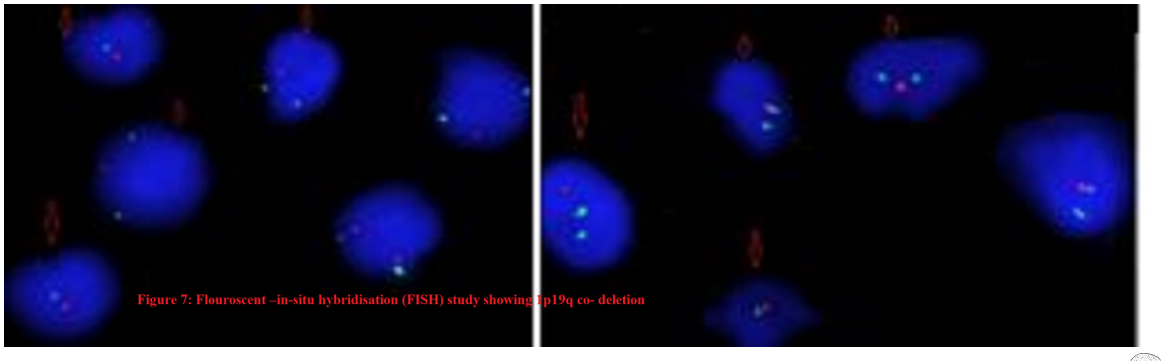

Figure 7. Fluorescent–In-Situ Hybridization (FISH) study showing 1p19q co-deletion

usually present with signs of raised intracranial pressure due to hydrocephalus. Radiological features are non-specific and sometimes may mimic ependymoma. Immunohistochemistry plays a pivotal role in accurate diagnosis and further characterization of the tumor. Chromosomal analysis helps in confirming the diagnosis and predicting treatment outcomes. Gross total resection is the treatment of choice followed by adjuvant chemotherapy. Patients with intraventricular ODG have a propensity for spinal drop metastasis. These tumors have a high recurrence rate and poor prognosis; thus frequent postoperative follow-up is necessary.

Ethical Considerations

Compliance with ethical guidelines

Written consent has been obtained from the subjects.

Funding

This research did not receive any grant from funding agencies in the public, commercial, or non-profit sectors.

Authors' contributions

Conception and design: Tamajyoti Ghosh, Subir Dey; Data collection: Tamajyoti Ghosh; Data analysis and interpretation: Tamajyoti Ghosh; Drafting the article: Tamajyoti Ghosh; Critically revising the article: Subir Dey; Reviewing submitted version of manuscript: Tamajyoti Ghosh; Approving the final version of the manuscript: Tamajyoti Ghosh, Subir Dey.

Conflict of interest

The authors declared no conflict of interest.

References

[1] Zada G, McNatt SA, Gonzalez-Gomez I, McComb JG. Anaplastic intraventricular oligodendroglioma: Case report and review of the literature. Surgical Neurology. 2009; 71(6):693-700. [DOI:10.1016/j.surneu.2007.11.013] [PMID]

[2] Lau CSM, Mahendraraj K, Chamberlain RS. Oligodendrogliomas in paediatric and adult patients: An outcome-based study from the Surveillance, Epidemiology, and End Result database. Cancer Management and Research. 2017; 9:159-66. [DOI:10.2147/CMAR.S117799] [PMID] [PMCID]

[3] Atasoy C, Karagulle A, Erden I, Akyar S. Primary ologodendroglioma of the lateral ventricle: Computed tomography and magnetic resonance imaging findings. Journal of Ankara Medicine School. 2002; 55:39-44. [DOI:10.5455/gulhane.23374]

[4] Kikuchi K, Kowada M, Mineura K, Uemura K. Primary oligodendroglioma of the lateral ventricle with Computed tomographic and positron emission tomographic evaluations. Surgical Neurology. 1985; 23(5):483-8. [DOI:10.1016/0090-3019(85)90243-5.] [PMID]

[5] Dolinskas CA, Simeone FA. CT characteristics of intraventricular oligodendrogliomas. American Journal of Neuroradiology. 1987; 8(6):1077-82. [PMID] [PMCID]

[6] Elwatidy SM, Albakr AA, Al Towim AA, Malik SH. Tumors of the lateral and third ventricle: Surgical management and outcome analysis in 42 cases. Neurosciences (Riyadh). 2017; 22(4):274-81. [DOI:10.17712/nsj.2017.4.20170149] [PMID] [PMCID]

[7] Agrawal A, Shanthi V, Reddy VU. Intraventricular anaplastic oligodendroglioma. Journal of Datta Meghe Institute of Medical Science University. 2018; 13(4):209-11. [DOI:10.4103/jdmimsu.jdmimsu_13_18]

[8] Romero Z, Mora-La Cruz E, Cardozo J. [Intraventricular oligodendroglioma. Description of 2 cases (Spanish)]. Investigación Clínica. 1996; 37(1):51-9. [PMID]

[9] Blumenfeld CM, Gardner J. Disseminated oligodendroglioma. Archives of Neurology and Psychiatry. 1945; 54:274-9. [DOI:10.1001/archneurpsyc.1945.02300100046004] [PMID]

[10] Beck DJ, Russell DS. Oligodendrogliomatosis of the cerebrospinal pathway. Brain. 1942; 65(4):352-72. [DOI:10.1093/brain/65.4.352.]

[11] Rodriguez FJ, Tihan T, Lin D, McDonald W, Nigro J, Feuerstein B, et al. Clinicopathologic features of paediatric oligodendrogliomas: A series of 50 patients. The American Journal of Surgical Pathology. 2014; 38(8):1058-70. [DOI:10.1097/PAS.0000000000000221] [PMID] [PMCID]

[12] Celli P, Nofrone I, Palma L, Cantore G, Fortuna A. Cerebral oligodendroglioma: Prognostic factors and life history. Neurosurgery. 1994; 35(6):1018-34. [DOI:10.1097/00006123-199412000-00003] [PMID]

[13] Packer RJ, Sutton LN, Rorke LB, Zimmerman RA, Littman R, Brunce DA, et al. Oligodendroglioma of the posterior fossa in childhood. Cancer. 1985; 56(1):195-9. [DOI:10.1002/1097-0142(19850701)56:13.0.CO;2-3]

[14] Yuen ST, Fung CF, Ng TH, Leung SY. Central neurocytoma: Its differentiation from intraventricular oligodendroglioma. Child's Nervous System. 1992; 8(7):383-8. [DOI:10.1007/BF00304785] [PMID]

[15] Shaw EG, Scheithauer BW, O'Fallon JR, Tazelaar HD, Davies DH. Oligodendrogliomas: The mayo clinic experience. Journal of Neurosurgery. 1992; 76(3):428-34. [DOI:10.3171/jns.1992.76.3.0428] [PMID]

[16] Pitt MA, Jones AW, Reeve RS, Cowie RA. Oligodendroglioma of the fourth ventricle with intracranial and spinal oligodendrogliomatosis: A case report. British Journal of Neurosurgery. 1992; 6(4):371-4. [DOI:10.3109/02688699209023798] [PMID]

[17] Natale M, Spennato P, Savarese L, Bocchetti A, Esposito S, Barbato R. Anaplastic oligodendroglioma presenting with drop metastases in the cauda equina. Clinical Neurology and Neurosurgery. 2005; 107(5):417-20. [DOI:10.1016/j.clineuro.2004.09.013] [PMID]