

Anaplastic Oligodendroglioma in an Adolescent Male; A rare case Report and Review of the Literature.

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Received: January 2017

Accepted: January 2017

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ABSTRACT

Anaplastic oligodendrogliomas are rare paediatric brain tumours. They represent a very small proportion of childhood brain tumours. The literature about neuroimaging findings is scant. A correct diagnosis is important to plan the surgery or therapeutic approach. We report a rare case of an anaplastic oligodendroglioma in a 15-year-old male with a brief review of the literature.

Keywords: Anaplasia, Brain tumour, Oligodendroglioma.

INTRODUCTION

Oligodendrogliomas are the tumours of normal glial cells of brain called oligodendrocytes. They represent the third most common glioma overall, accounting for 2%–5% of primary brain tumours and 5%–18% of all glial neoplasms^[1-4]. The tumour accounts for less than 1% of paediatric central nervous system neoplasms^[5]. Most oligodendrogliomas manifest in the adult age groups with a peak incidence in the fourth and fifth decades^[6-8]. Patients with anaplastic tumours are usually slightly older (peak age, sixth and seventh decades) than those with well-differentiated oligodendrogliomas^[8,9]. A small percentage arises in children, accounting for a second smaller age peak at 6–12 years^[3,4,10-12]. Incidence of its anaplastic variant is further lower. We report a rare case of an anaplastic oligodendroglioma in a 15-year-old male with neuroimaging findings and brief review of the literature.

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CASE REPORT

A 15 years old male child presented to our hospital with complaints of seizures and headache for last one year. On examination no significant findings were seen. Hematological parameters were within

normal limits. Contrast enhanced CT scan head was done which revealed a large cortical based complex heterogeneous mass lesion measuring approx. 71 x 55 x 45 mm (AP X TR X CC) in left cerebral hemisphere in high fronto-parietal region. It showed cystic components of varying sizes with areas of internal haemorrhages, foci of coarse calcifications and heterogeneous post contrast enhancement. Perilesional mass effect was also evident in form of contralateral midline shift & effaced left lateral ventricle [Figure 1a, 1b].

MR imaging confirmed the presence of a large cortical based complex heterogeneous mass lesion measuring approx. 76 x 58 x 49 mm in left cerebral hemisphere in high fronto-parietal region, showed heterogeneous altered is to hypointense signal intensities on T1W sequence with subtle areas of T1W hyperintensities within suggestive of haemorrhages [Figure 2a]. On T2W and inversion recovery sequences there was presence of heterogeneous altered hyperintense signals with internal cystic components and internal fluid levels [Figure 2b, 2c] on ADC mapping lesion showed variable ADC values [Figure 2d]. Multiple foci of blooming were also noted within the mass lesion on SW and Phase sequences confirmed the presence of internal calcifications [Figure 2e, 2f]. On post contrast sequences mass lesion showed intense heterogeneous enhancement [Figure 2g]. Perilesional mass effect was also evident in form of contralateral midline shift & effaced left lateral ventricle [Figure 2a, 2b].

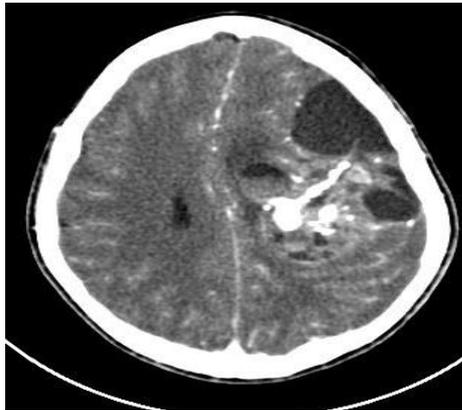
On the basis of CT and MRI brain imaging findings and location possibility of anaplastic

oligodendroglioma was kept with a differential of supratentorial P-NET.

Subsequently patient was operated, craniotomy was done under general anaesthesia with total excision of the mass lesion and confirmatory histopathology was done. Post-operative period was uneventful.

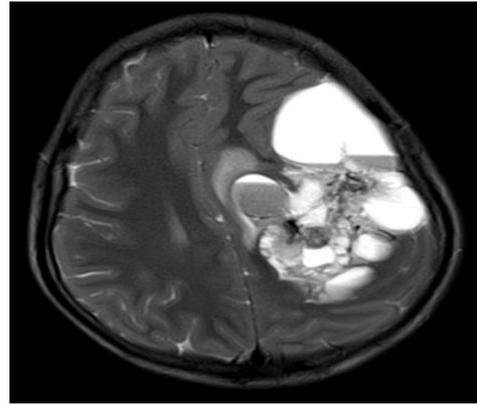


1a

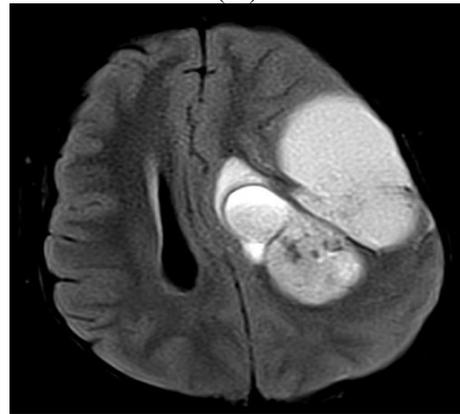


1b

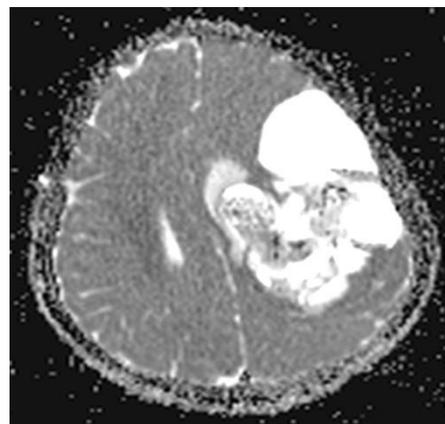
Figure 1: Axial plain and contrast enhanced CT scan head showing a large cortical based complex heterogeneous mass lesion in left cerebral hemisphere in high fronto-parietal region with cystic components of varying sizes, areas of internal haemorrhages, foci of coarse calcifications, perilesional mass effect in form of contralateral midline shift & effaced left lateral ventricle, (1a) and heterogeneous intense enhancement on contrast administration (1b).



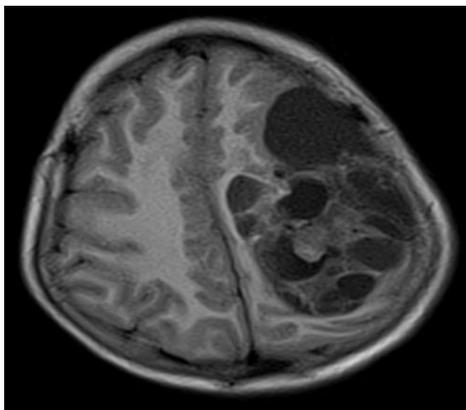
(2b)



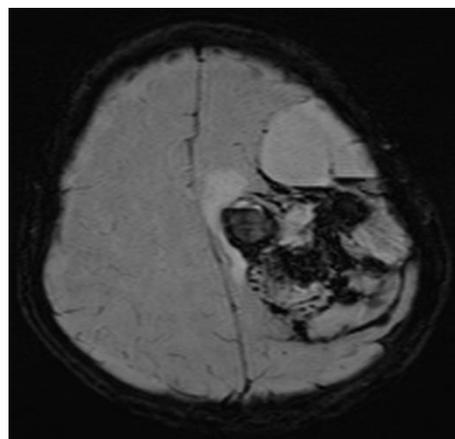
(2c)



(2d)



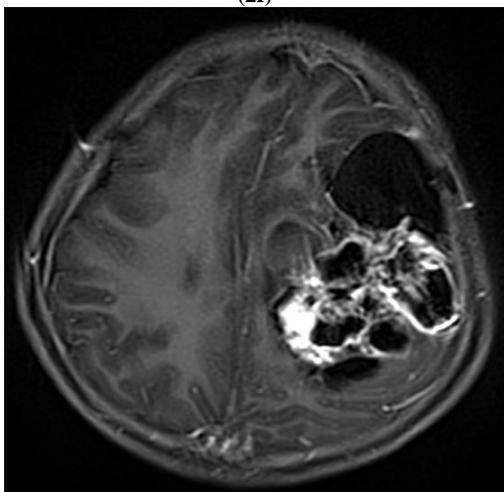
(2a)



(2e)



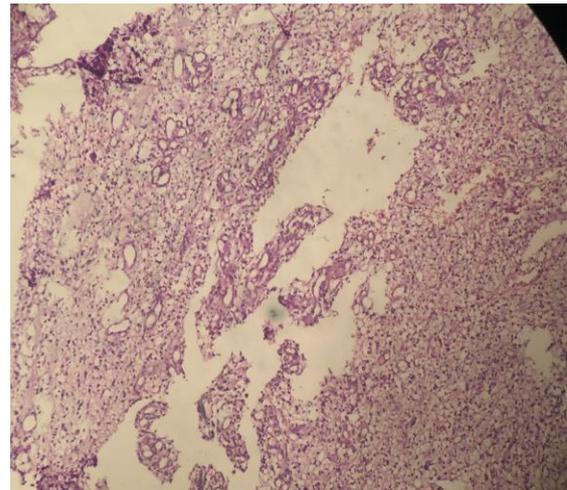
(2f)



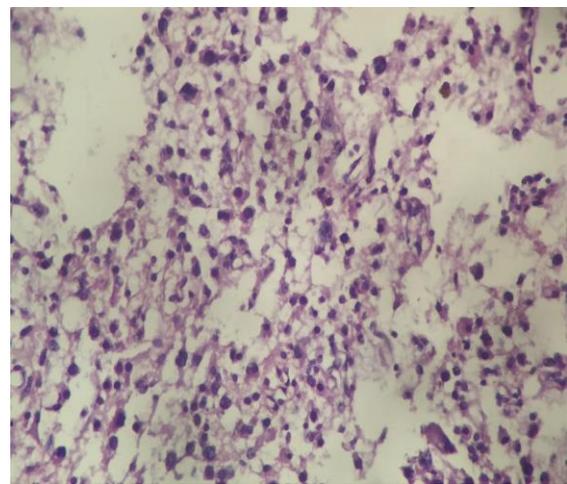
(2g)

Figure 2: MRI of the brain confirms the presence of a large cortical based complex heterogeneous mass lesion in left cerebral hemisphere in high fronto-parietal region, showing heterogeneous altered iso to hypointense signal intensities on T1W sequence with subtle areas of T1W hyper intensities within suggestive of haemorrhages (2a). On T2W and inversion recovery sequences showing presence of heterogeneous altered hyperintense signals with internal cystic components and internal fluid levels (2b & 2c). On ADC mapping lesion showing areas of variable ADC values (2d). Multiple foci of blooming within the mass lesion on SW and Phase sequences confirm the presence of internal calcifications (2e & 2f). On post contrast sequences mass lesion showing intense heterogeneous enhancement (2g).

On histopathology sections showed richly vascular tumour with cells having round to oval nuclei with foci showing nuclear pleomorphism and irregularity, clear cytoplasm. Exuberant calcification with anastomosing capillaries forming configuration of chicken wire were seen endothelial proliferation was evident. These features were consistent with anaplastic type oligodendroglioma –WHO Grade III [Figure 3a, 3b].



a



b

Figure 3: Photomicrographs (hematoxylin and eosin staining) showing richly vascular tumour with cells having round to oval nuclei with foci showing nuclear pleomorphism and irregularity, clear cytoplasm. Exuberant calcification with anastomosing capillaries forming configuration of chicken wire were seen endothelial proliferation consistent with anaplastic type oligodendroglioma –WHO Grade III (3a & b).

DISCUSSION

Oligodendroglioma in children is a rare tumour. Most oligodendrogliomas manifest in the adult age groups with a peak incidence in the fourth and fifth decades [6-8]. The tumors account for less than 1% of paediatric central nervous system neoplasms [13-15] with a child: adult ratio approx. 1:8 [16]. Incidence of its anaplastic variant is further lower.

The clinical presentation is often of several years duration with most patients presenting with seizures, reflecting the strong predilection of this tumour to involve the cortical gray matter [17,18]. These tumours characteristically show a cortical-subcortical location, most commonly in the frontal lobe. Due to their superficial location, there may be focal

thinning, remodelling or erosions of the overlying skull^[18].

At computed tomography (CT), about 60% of oligodendrogliomas are hypodense while 23% are isodense and about 6% are hyperdense in nature^[18]. Calcification, usually coarse in morphology, is noted in 20%–91% of cases^[19].

Occasionally, cystic degeneration and hemorrhage may be seen^[18,20]. However, the presence of necrosis, cystic degeneration, and hemorrhage, the usual imaging appearance of an anaplastic oligodendroglioma is more variable compared to that of an oligodendroglioma^[21].

Enhancement following intravenous contrast material administration is seen in 15%–20% of oligodendrogliomas and is associated with higher-grade tumors^[18-21]. In contrast to other low-grade gliomas, moderate enhancement is commonly seen and perfusion may be moderately increased^[16]. Enhancement in a previously non-enhancing, untreated tumour is suggestive of malignant transformation due to its high growth rate.

Calcification, vasogenic edema, and enhancement are less commonly noted in children with oligodendrogliomas than in adults^[22]. The tumour may not be visualized on CT images^[19].

MR imaging is superior to CT in defining the full extent of tumour involvement^[23]. The tumor is usually hypointense compared to gray matter on T1-weighted images and hyperintense compared to gray matter on T2-weighted images^[18]. Heterogeneity of this signal intensity is the rule. Less commonly, a large cyst-like pattern may be seen^[24]. Surrounding vasogenic edema is not common^[18]. Advanced MR imaging with the apparent diffusion coefficient (ADC) shows a characteristic but not pathognomonic difference between low-grade and high-grade glial neoplasms. Lower ADC values, indicative of water restriction and likely reflective of lowered extracellular hyaluronic acid, are noted in high-grade tumors compared to the higher ADC values seen in low-grade tumors^[25,26].

On imaging, these tumours characteristically show calcifications, SWI sequence differentiates haemorrhage from calcification and plays a pivotal role because calcification is diamagnetic, whereas most haemorrhagic by-products are paramagnetic. Due to the fact that calcium and iron have opposite magnetic susceptibilities, their phase deflections are opposite as well^[27-29].

The pattern of contrast enhancement seen at CT is also noted at MR imaging. “Dot-like” lacy enhancement is commonly seen, but many tumours may not enhance at all^[24]. The presence or absence of enhancement has even been utilized to some extent in the grading of these neoplasms by Daumas-Duport and colleagues^[25]. While the presence of enhancement tends to be noted more commonly in more aggressive oligodendrogliomas, however, the absence of enhancement does not always suggestive

of low-grade tumour. Histologic confirmation is always required^[27,28].

Histopathologically it is classified as two main types of tumours : well-differentiated oligodendroglioma and its anaplastic variant. Less commonly, neoplastic mixtures of both oligodendroglial and astrocytic components occur and are termed oligoastrocytomas, with both well-differentiated and anaplastic forms. Their hallmark molecular feature is codeletion of the 1p and 19q chromosome arms, which is not only of diagnostic but also of prognostic and predictive relevance^[16].

Total surgical resection is the treatment of choice with many patients experience a long post-operative disease free asymptomatic period. Especially in younger patients who have undergone complete or near complete total resection of the tumor, recommend no further therapy until there is evidence of recurrence^[29,30]. However, because of its infiltrative nature, the tumor may frequently be poorly delineated from the normal brain parenchyma, making complete resection impossible^[31].

In cases of partial resection, recurrence or metastasis, additional chemotherapy and radiotherapy are also indicated.

Advanced multi-detector CT and magnetic resonance imaging techniques play increasingly important roles in both pre- and postoperative assessment of these complex neoplasms.

CONCLUSION

MRI is superior to CT in assessing tumour extent and cortical involvement, whereas CT is most sensitive to calcification. Advanced and functional imaging techniques may aid in assessment of grading of lesion.

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How to cite this article: Saraswat S, Sharma S, Kumar A, Chandak S, Kumar A. Anaplastic Oligodendroglioma in an Adolescent Male; A Case Report and Review of the Literature. *Ann. Int. Med. Den. Res.* 2017; 3(2):RD14-RD18.

Source of Support: Nil, **Conflict of Interest:** None declared