Case Report

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A Rare Case Report of Frontotemporal Oligodendroglioma with Extraneural Orbitoethmoidal Invasion: Its Current Implications

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Abstract

Oligodendrogliomas generally remain confined to the nervous parenchyma throughout their entire course. However, there are exceptionally rare cases, which show either transdural extension or extraneural spread. In reviewing the world literature on suchspread from oligodendrogliomas, very few cases were found, and the exit door from the CNS was almost always through the cranial base, anentitity termed as metastatic anaplastic oligodendrioglioma. Here we present one such rare case of a frontotemporal oligodendroglioma (WHO grade 3) with primary orbito-ethmoidal invasion. Patient presented with right sided proptosis and nasal cavity mass lesion causing epistaxis post. MRI revealedtransduralextension of tumor with right orbital and nasal cavity infiltration through the ethmoidal bone. Atransnasal endoscopic excision of the nasal mass conformed to the immunocytogenetics of an oligodendroglioma. The reporting of such cases is important because firstly following the advent of IHC and the new WHO 2016 classification, the lines between low and high grade gliomas have become blurred. Secondly, the pathogenesis underlying the invasion of neuroepithelial tumors outside the dural structures calls for further research and may promote anti-invasive therapy in the future.

Keywords: Oligodendroglioma; Extraneural; Orbitoethmoidal; Immunohistochemistry.

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Introduction

Neuroepithelial tumors, oligodendrogliomas in particular, generally remain confined to the nervous parenchyma throughout their entire course. However, there are exceptionally rare cases, which show either transdural extension or extraneural spread. In reviewing the world literature on extraneural metastases from neuroepithelial tumors, very few such cases were found and the exit door from the CNS was almost always through the cranial base.

The existing anatomical structures in the central nervous system seem to facilitate the spread of malignant glioma cells to varying degrees, depending on the type of tumour. Outside and also within the CNS, definite barriers seem to exist. Theoretically, this might be due to non-permissivenes of surrounding tissues to glioma invasion, or that there is a real biological barrier as described by Laerum.[1] With progression, however, malignant gliomas may in rare cases both invade the meninges and metastasise to extracranial sites. Usually, this occurs after intracranial surgery or establishment of an intracranial shunt. We present here one such case of right frontotemporal oligodendroglioma with primary orbito-ethmoidal invasion.

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Case Findings

A 30 year old female, presented with 1 year history of recurrent headaches and seizures. She had no neurological deficits on examination, whatsoever. On imaging she was diagnosed to have 8 × 5 × 5 cm right side frontotemporal region SOL which was involving the right superior and middle temporal gyrus, perisylvian cortex and insular region with perilesional edema and mass effect. Erosion of the right posterior wall of frontal sinus was seen with infiltration of right frontal sinus right ethmoid sinus,right middle meatus and right posterior choanal space. Also right superior orbital wall was eroded with superior extraconal space infiltration with minimal mass effect (Fig. 1).

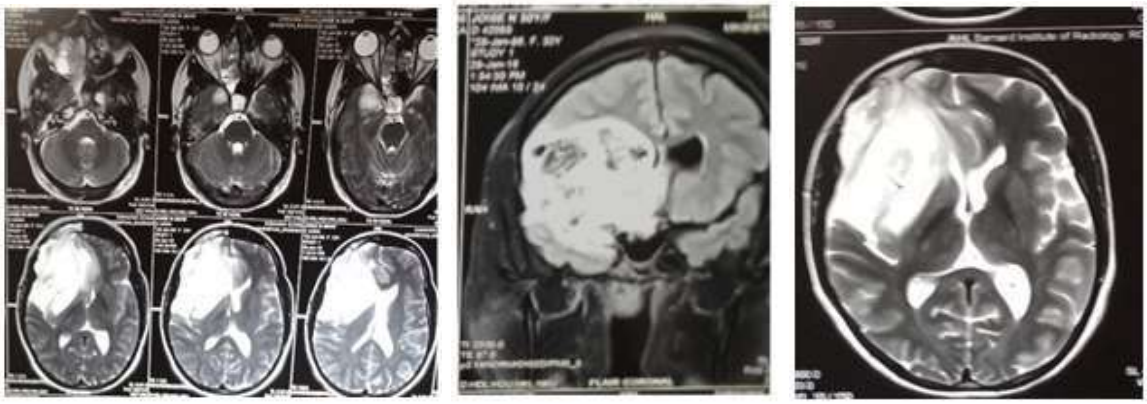

Fig. 1: Pre-op MRI

She underwent right frontotemporal craniotomy and near total excision of the tumor.The extracranial component was not excised, as the extension was minimal as she had no symptoms then. HPE reported as oligodendro-glioma.Patient received RT post-op for a year and was asymptomatic for a year.

However after 2 yrs. patient presented with complaints of epistaxis, right side nasal stuffiness and right sided retro orbital pain and mild to moderate proptosis; however vision was intact. Repeat MRI showed gliosis in the right frontotemporal region with some amount of residual lesion. However there was increased infiltration into the nasal and orbital cavities with tumor extending into the nasopharynx as well. CT orbit also showed erosion of the medial wall and extension into the right maxillary sinus, along with right frontal and ethmoidal sinuses (Fig. 2).

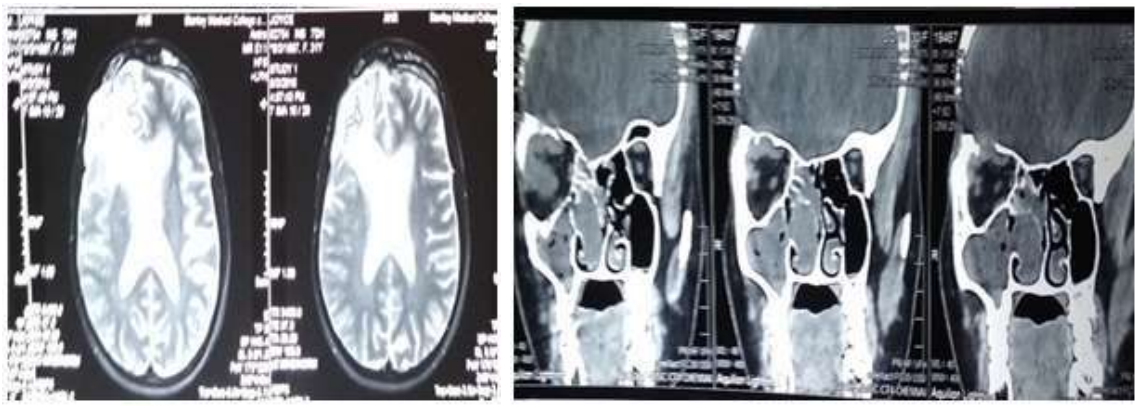

Fig. 2: MRI and CT PNS 2 yrs. Post surgery

On nasal endoscopic examination, there was a mass visible for which excision was attempted and biopsy proved as oligodendroglioma. IHC was performed which proved GFAP positive, IDH mutant type with ATRX retention and p53 positivity, confirming oligodendroglioma. Patientis now undergoing RT on regular follow up (Fig. 3).

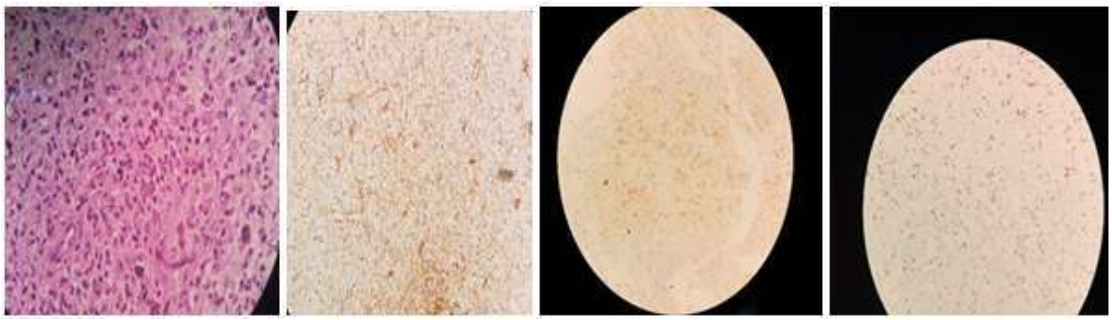

Fig. 3: HPE and IHC Slides HPE GFAP Positivity IDH Positivity ATRX Retention

Discussion

Oligodendrogliomas,like stated before, are infrequently associated with extraneural spread. Lubenstein[2] and colleagues in their review of 116 cases had outlined the mechanism of extracranial spread of primary CNS tumors of which ODG's constituted only 5.2%.

The first reporting of extraneural scalp metstasis of ODG was way back in 1950 by James and Pagel[3] in British Journal of Surgery,followed by a similar such case reported by Spataro and Sacks[4] in 1968. Both cases were recurrent cases with long case history and multiple surgeries, hence the surgical route for the dissemination of cancer cells was proposed. Furthermore a total of 61 cases of ODG extracranial spread have been reported by Li G,[5] *et al*. as described by Aydemir[6] and colleagues in their review paper on the mechanisms of spread of anaplastic oligodendroglioma. Since then many other cases of extraneural spread reported distant from the site of primary.

Most of them were distant metastasis to the bone and bone marrow,the other organs being involved are lymphatics, spleen, lungs, liver, breast etc. The reader is advised to go through the references at the end of article for more such case reports.[7-34]

The first case of transdural extension of gliomas was, however, reported in 1993 by Pompilii[35] *et al*. in the Journal of Neuro-oncology. This case was similar to the one describedhere, as in there was extraneural spread to the ethmoid sinus and orbital cavities,but following surgery.

Also Vahedi *et al*.[36] in the same year reported one more such case of ODG with recurrence and involvement of ethmoid sinus and left orbital cavity without loss of vision. However the proposed mechanism was raised ICP leading to spread of glioma cells through the cranial base under pressure. Some important observations were made;

1. Site of surgery was far from spread, hence surgery could not be directly responsible for the spread
2. An independence between degree of anaplasia and transdural spread was established since both tumors were low grade.
3. In each case of frontal glioma anterior cranial fossa and sinus involvement should be looked for in MRI.

Following this report, O.D Laerum[1] in his paper in 1997 in Acta Neuro Chirurgica discussed the mechanisms for primary invasiveness of ODG. He described in detail about the factors responsible for invasion like cell adhesion molecules NCAM and CD 44; growth factors like EGF, HGF and Glial motility factor; as well as biological barriers to the spread of invasion. However search is still on to find the genetic determinants of invasiveness and in future possibly to formulate anti invasive therapy to prevent the spread of these tumors.

Ali Akhavan[37] *et al*. in their paper in 2012 in BMJ reported one such case of anaplastic ODG with optic nerve involvement leading to visual loss. Further they identified the most unusual sites of anaplastic ODG involvement. Also it was noted that metastatic involvement was afterall, not so uncommon, and confirmed the 3 most possible routes of spread extraneurally,as originally proposed by Engelhard *et al*.[38] in 2003.

However now due the emergence of IHC and molecular markers and with the WHO 2016 classification the entire scenario has changed. Oligodendrogliomas are no more merely classified based on grades, but now IDH mutant and wild types have been described; with the mutant types having a better diagnosis. Where molecular studies are inconclusive, a separate class called NOS has been generated, thus paving the way for deeper molecular subclassification. Although the histologic subtype is still in vogue, the mportance with which it was previously associated has been diluted.

Terms such as anaplastic and oligoastrocytoma carry a lesser meaning today. Treatment modalities too are being developed targeting IDH subtype or based on 1p/19q codeletion or ATRX status.[39-40]

The need for reporting of one such casestems from the fact that, the previous theories need to be debunked and in the light of new evidence, further research should be encouraged to characterise the invasiness of neuro-epithelial tumors, especially oligodendrogliomas. Only then we can think of specifically targeting these factors and develop anti invasive therapies, which might prolong the lives of this subcategory of patients.

Conclusion

Neuroepithelial tumors, especially oligodendro gliomas, per se primarily do not spread extracranially or extraneurally. However, a review of literature shows that oligodendrogliomas too have, very rarely,such aggressive history of extraneural and transdural spread, an entity described as metastatic anaplastic oligodendroglioma. Our case is different from this series, in the sense there was only obvious local invasion; however the pathogenetics may stillremain the same. The aim of reporting one such case of ODG here is to better help characterise these lesions and in the light of new technology fuel research into the invasiveness properties of these subtype of tumors, ultimately to achieve a better patient care.

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