Direct seventh-eighth cranial nerve involvement by brain stem glioma – a case report

Sanjay M. Khaladkar, Aarushi Gupta, Arijit Ghosh, Poorvi Sharma, Pooja Karanjule
Department of Radiodiagnosis, Dr. D.Y. Patil Medical College and Research Center, India

ABSTRACT
Brainstem gliomas constitute 10-20% of all pediatric CNS tumors. Brain stem astrocytomas are usually fibrillar.
Based on their appearance they can be focal, exophytic, cervico-medullary and focal tectal gliomas. Malignant gliomas are characterized by the infiltrative growth of malignant cells and they grow along the white matter tracts. Rarely, an extension of a tumor may be seen along the cranial nerve, simulating a nerve sheath tumor. We report a case of a brainstem glioma in a 5-year old female patient, extending along left seventh-eighth cranial nerves into the left internal auditory canal.

Keywords: Brainstem, Glioma, Cerebello-pontine angle, seventh–eighth nerve complex.

INTRODUCTION
Brainstem gliomas (BSG) constitute 10-20% of all pediatric CNS tumors. They generally occur in childhood with the average age of diagnosis in 7-9 years with no gender predisposition [1, 2]. Prior to the era of modern imaging, all brainstem gliomas were regarded as a single pathological entity and their prognosis was considered poor. In 1969 MATSON summarized that BSG must be considered as a malignant tumour regardless of specific histology as their location renders them inoperable [3]. A classification system was later introduced to identify tumors that benefitted from surgery. With the advent of MRI, morphological patterns of brainstem gliomas evolved. These help in predicting tumor behavior and in the management protocol.

A CASE REPORT
A 5-year old female patient presented with the loss of balance, headache, deviation of mouth to the right side, double vision for the past 2 months, and vomiting, neck pain for the past 8 days. MRI of the brain was performed with contrast. It showed a large diffuse lesion involving pons and medulla, causing pontine expansion. It was heterogeneously hypointense on T1WI (Figures 1, 7), heterogeneously hyperintense on T2WI and FLAIR (Figures 2, 3, 4). Extension of the lesion was noted in the left cerebellopontine (CP) angle cistern and along the left seventh and eight nerve complex into the left auditory canal (Figures 2 d, 4 b, 6). Posteriorly, it was extending into the floor of the fourth ventricle (Figures 1, 2, 3). No proximal obstructive hydrocephalus was noted. The dorsal portion of the mass was extending along the fourth ventricle and in the left CP angle cistern, and showed a mild restricted diffusion with low ADC values (Figure 5). The contrast study showed a heterogeneous enhancement in the dorsal and cisternal portions of the mass (Figure 8). The remaining portion of the mass showed no significant enhancement. MR spectroscopy revealed elevated choline relative to the NAA signal (Figure 9). Radiotherapy was suggested, which was refused by parents. The patient eventually died after two months.
Figure 1: Sagittal T1WI – showing a diffuse hypointense lesion in the medulla and pons with cervicomedullary kinking.

Figure 2: Axial T2WI – showing a diffuse hyperintense lesion in the medulla and pons with extension in the left paramedullary, parapontine and left cerebellopontine cisterns, and with extension along the left seventh-eighth nerve complex into the left internal auditory canal.
Figure 3: Axial FLAIR – showing a diffuse hyperintense lesion in the medulla and pons with extension in the left paramedullary, parapontine and left cerebellopontine cisterns.

Figure 4: Coronal T2WI – showing a diffuse hyperintense lesion in the medulla and pons with extension in the left paramedullary, parapontine and left cerebellopontine cisterns, and with extension along the left seventh-eight nerve complex into the left internal auditory canal.

Figure 5: Axial DWI and ADC – showing a subtle restricted diffusion in the left lateral portion of the mass with low ADC values.

Figure 6: Axial CISS – showing a diffuse hyperintense lesion in the medulla and pons with extension in the left paramedullary, parapontine and left cerebellopontine cisterns, and with extension along the left seventh-eight nerve complex into the left internal auditory canal.

Figure 7: Axial T1WI fat saturated (Pre-contrast) – showing a diffuse hypointense lesion in the medulla and pons extending into the left paramedullary cistern.
Figure 8: Post-contrast Axial T1WI fat saturated (a), Coronal FLAIR (b) – showing a diffuse enhancement in the lateral portion of the mass extending to the left parapontine and left cerebello-pontine cisterns.

Figure 9 – MR Spectroscopy in the enhancing portion of the mass shows raised choline and reduced NAA, with increased choline creatinine ratio.
DISCUSSION

Brain stem astrocytomas are usually fibrillary in contrast to cerebellar astrocytomas, which are usually pilocytic. MRI is useful in detecting the epicenter of a tumor, expansion of involved portion of the brainstem, predicting its biological behavior. Depending on the appearance, they can be focal, exophytic, cervicomedullary and focal tectal gliomas. Diffuse brainstem gliomas are generally more than 2 cm in size during the time of presentation, and are characterized by a diffuse infiltration and swelling/hypertrophy of the brainstem. The epicenter of a lesion is usually in the pons. Rostral or caudal tumor extension may be seen. Focal tumors are defined as a demarcated lesion in the midbrain, pons or medulla, either solid or cystic, and have well defined margins in the MRI. They are usually histologically benign (Grade I or II), rarely anaplastic gangliogliomas and PNET have been reported. There is a lack of infiltration and edema. Exophytic tumors (dorsally exophytic brainstem gliomas) arise from subependymal glial tissue. The bulk of tumor resides in the fourth ventricle. Hence, there is a relatively late onset of symptoms [4-6]. Malignant gliomas are characterized by infiltrative growth of malignant cells and they grow along the white matter tracts [7]. The cranial nerve nuclei are within the brain parenchyma and have glial cells, which extend into the root entry zone and proximal cisternal segments with the gradual replacement of Schwann Cells of more than 1-9 mm [7,8]. Hence, infiltrating growth is seen along the white matter tracts. The extension of a tumor may be seen along the cranial nerve, simulating a nerve sheath tumor, intracranial perineural spread of head and neck tumor, leptomeningeal spread of tumor [7].

Before 2014, 11 unique cases of gliomas directly infiltrating cranial nerves have been reported in the literature with 8 cases detected in imaging [8-10]. 8 additional cases of pathologically confirmed gliomas with imaging findings indicating direct involvement of cranial nerves have been identified by M.C.Mabray [7]. The imaging technique in identifying the intra-axial origin of a disease is the involvement of the pons and medulla, deep to the root entry zone of the cranial nerves by a lesion contiguous and matching the cranial nerve involvement in signal characteristics, and enhancement patterns, and expansion of the brainstem. Expansion of the adjacent pons or midbrain, thickening of the root entry zone and the cisternal segment of the cranial nerve having similar signal characteristics and enhancement patterns on MRI are the key features in differentiating intra-axial origin of a tumor with the involvement of cranial nerves from the nerve sheath tumors, diffuse neoplastic, infectious or inflammatory leptomeningeal diseases. Cranial nerves, which can be involved, are the oculomotor nerve and the trigeminal nerve apart from the vestibulocochlear nerve and the facial nerve. Intrinsic tumors of the brainstem are astrocytoma (the most common), PNET, lymphoma, ganglioglioma and oligodendroglioma [11]. Non-neoplastic lesions arising from the brainstem are tuberculomas, cavernous malformations, haemangioblastomas and epidermoids.

CONCLUSION

Brainstem gliomas directly infiltrating cranial nerves is a rare but known condition. This entity should be known in order to differentiate it from the nerve sheath tumor, intracranial perineural spread of head and neck tumor, and leptomeningeal spread of a tumor. These tumors can cause cranial neuropathy. Identification of intraparenchymal, intraaxial origin of a cranial nerve involvement by glioma is the key to its correct diagnosis.
REFERENCES


