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Original Research Paper

Neurosurgery

EXOPHYTIC LOW GRADE BRAINSTEM GLIOMA – TOTAL EXCISION

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INTRODUCTION

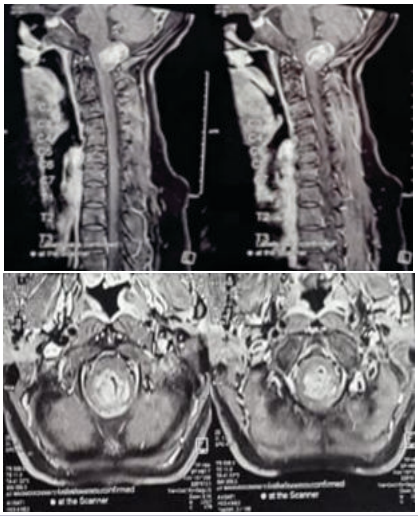
Low grade brainstem glioma frequently develop in the medulla. They often have an exophytic componenet (1,2,3). These lesions develop in young adults and children and are revealed by long tract involvement and lower cranial nerves dysfunction. Surgical resection is preferred for exophytic lesions as they have a better overall prognosis than midbrain and pontine lesions. It is found to be curative, allowing the control of the disease and avoiding adjuvant treatments. (2,3,4,5)

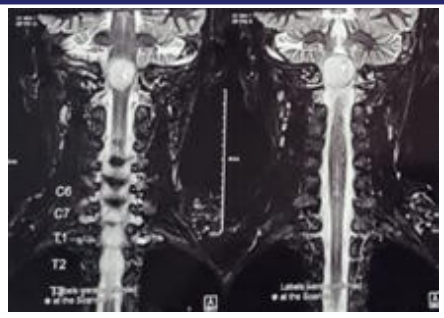
Case Report

A 55 year old lady presented with neck pain since 4 months. It was localized to the neck and non radiating. She had no difficulty in gripping objects in both the hands. She also complained of intermittent blurring of vision. She gave history of associated occipital headache. She complained of bilateral lower limb tightness.

There was no history of trauma , vomiting , seizures or any associated bowel or bladder complaints.On examination, she was conscious and oriented to time , place and person, and haemodynamically stable. She had no neuro deficits and all cranial nerve examinations were normal.

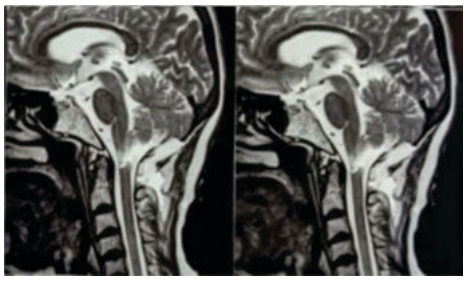
MRI was s/o well defined heterogeneously enhancing intradural extramedullary lesion at cervico-vertebral extending to adjoining posterior fossa causing mass effect on dorsal aspect of lower medulla and spinal cord with diffrential diagnosis either astrocytoma vs neurogenic tumour or ependymoma.





She was worked up and after thorough pre anaesthetic check up, she was taken up for midline suboccipital craniectomy with maximal safe total excision of tumor under GA. Suboccipital craniectomy was done along with C1 laminectomy. Tumor was identified which was firm, moderately vascular and suckable. Tumor was excised using CUSA. Gross total excision of tumor was done.

Post Operative MRI



- HPE report-
- Low grade astrocytoma
- IHC findings –
- P53 –negative
- Ki67-2.25%
- GFAP-Diffusely positive

DISCUSSION

Brainstem gliomas are heterogeneous, ranging from high-grade lesions that are rapidly fatal despite aggressive therapyto low-grade tumors that need little or no treatment(6). Prognosis and treatment depend upon histologic features and the location.

Focal brainstem gliomas typically arise in the midbrain and medulla, and typically are discrete, well-circumscribed tumors. Histologically, these tumors are most often pilocytic or diffuse astrocytomas, or rarely, gangliogliomas, all of which

are considered low-grade tumors(7). Evidence of locally invasive growth or edema may be present in tumors with nonpilocytic histology.

Around 80 percent of brainstem gliomas that occur in midbrain and medulla are low grade (World Health Organization [WHO] grade 1 and 2 tumors)(8,9). The WHO grading system for gliomas is based upon both molecular and histologic features(10). Low-gradeastrocytomas are characterized by occasional nuclear atypia and varying degrees of cellularity, although they lack other features of malignancy. The histology of grade 1 tumors (pilocytic astrocytomas)consists of Rosenthal fibers (composed of alpha-B crystallin) and biphasic microcystic areas interspersed between more compacted and cellular areas with extensive fibrillary processes(11). There may be a large macrocystic structure containing a mural nodule occasionally. Pilocytic astrocytomas can possess a vascular proliferation and few mitoseswithout elevation to a higher grade.

The location of the tumor generally reflect the signs and symptoms(12,13).

Small focal tumors of the midbrain or medullapresent insidiously, with a long history of localizing findings such as contralateral hemiparesis or an isolated cranial nerve deficit. Clinical features of raised intracranial pressure (ICP) are rare.

Dorsally exophytic tumors and focal tumors of the tectum, the dorsal aspect of the rostral or upper midbrain, typically present with vomiting, headache,and ataxia secondary to aqueductal occlusion causing hydrocephalus. Cranial nerve deficits are seen in around half of patients, but long tract signs are uncommon.

Medullary tumors may be associated with dysphagia, apnea, nasal speech and cranial nerve dysfunction. Patients may also have nausea, vomiting, ataxia and weakness. Rarely, a focal pontine tumor can produce auditory nerve and fascial nerve dysfunction.

Cervicomedullary junction tumors can cause quadriparesis or hemiparesis from upper motor neuron dysfunction, as well as loss of reflexes, muscle atrophy, and weaknessfrom lower motor nerve dysfunction. Hydrocephalus is rare, but patients may have cranial neuropathies, vomiting, headache, apnea and sensory deficits.MRI of the brain with and without contrast is the method of choice to image tumors in the brainstem, as many of these lesions are isointense on CT(13)

On MRI, focal low-grade brainstem gliomas most often appear as masses of limited size that may be cystic, well demarcated, noninfiltrating, and without associated edema. They are typically isointense or hypointense on T1-weighted images, hyperintense on T2-weighted images, and enhance uniformly and brightly with intravenous contrast(14).

Treatment of focal brainstem gliomas requires a consideration of the anatomic location of the tumor, which determines both symptoms and accessibility to surgical resection. The long- term prognosis of patients with brainstem low-grade glioma is excellent(15).

Surgical resection, radiation therapy (RT), and chemotherapy each can play a role in the management of patients with focal brainstem gliomas.

Although there are no randomized trials,for focal brainstem gliomas in surgically accessible locations, such as at the cervicomedullary junction in selected cases(16) and for dorsal exophytic lesions resection has become the preferred treatment. In patients for whom resection is contraindicated, stereotactic biopsy can provide important histologic and molecular information to guide further therapy.

Around 30 percent of resected tumors will progress and require further therapy. A second operation can be considered for patients with surgically accessible disease(17,18). For others, the most reasonable option is chemotherapy in children and focal RT in adults.

The approach to dorsal exophytic gliomas relies upon surgical resection whenever possible. Safe resection generally requires intraoperative guidance to achieve a maximal degree of tumor resection.Routine postoperative RT is not indicated and should only be considered for the rare patient with a high-grade lesion or for those with low-grade lesions who progress rapidly after initial resection(19). Later recurrences can be managed with reresection, RT, or chemotherapy.

Progression-free survival is seen in approximately 40 to 70 percent(20,21), and chronic disability is common. This disability results from both treatment-related morbidity and tumor-related neurologic damage resulting from compression of the brainstem, long nerve tracts, and cranial nerves(22,23).

For survivors of focal brainstem tumors, the duration of symptoms at presentation and the severity of the tumor-associated disability are the most important variables in predicting long- term neurologic outcome(24). Even patients receiving only surgery for a low-grade brainstem glioma can have significant functional impairment, indicating that all patients should be considered at risk(25).

CONCLUSION

Low grade brainstem gliomas are uncommon tumors. Most of these arise in the midbrain, medulla, or cervicomedullary junction.

On MRI, focal low-grade brainstem gliomas appear as lesions of limited size (<2 cm) that may be cystic, well demarcated, noninfiltrating, and without associated edema.

Whenever possible, histologic diagnosis should be supplemented by molecular diagnostics, which can provide important diagnostic, prognostic, and therapeutic information. Molecular targeted therapy is being studied in ongoing clinical trials and represents a rapidly emerging treatment option for low-grade glioma patients.

For patients with newly diagnosed focal brainstem gliomas other than tectal gliomas,surgical resection is suggested if it can be accomplished without excessive surgical morbidity.For patients with tumors that are unresectable or only partially resectable, radiation therapy (RT) offers an alternative to surgery.

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