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Cervicomedullary Ganglioglioma in a Child – A Case Report

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Key words: Ganglioglioma – Intractable seizure – Posterior fossa – Cervicomedullary junction – Spinal cord

Abstract: Ganglioglioma is a benign slow-growing neoplasm that most frequently occurs at the supratentorial region. Nevertheless, there are occasional reports of ganglioglioma occurring in the brainstem and spinal cord. Here we report a rare case of the craniocervical ganglioglioma. A 3.5-year-old male, presented with severe progressive quadriparesis, gait disturbance, and sphincter deficit. Physical examination demonstrated the quadriparesis, associated with positive Hoffman, Babinski, and clonus signs, and increased respond of deep tendon reflexes. Magnetic resonance imaging (MRI) demonstrated an ill-defined mass within medulla and upper cervical spinal cord, which was hypo to iso signal on T1, heterogeneous iso to hypersignal on T2 and demonstrated marked bright enhancement on T1 with gadolinium (Gad) injection. On surgery, the mass had a soft texture, ill-defined border, and grey to brown appearance. According to the frozen section report, and due to the absence of the tumour-neural parenchymal interference, only decompression of the tumour and expansile duraplasty were performed. The histopathology revealed ganglioglioma. On last follow-up 14 months after surgery, the patient was asymptomatic and neurological status was improved. The craniocervical MRI demonstrated the tumour that did not grow. Although it is rare, the ganglioglioma should be in the differentiated diagnoses of tumours with compatible clinical and radiologic features even in the unusual locations, especially in the pediatric and young patients. Safety surgical resection should be considered in these patients, whenever possible. In the case of partial resection, that is common in the tumours located within functionally critical structures, long close follow-up rather than radiation therapy is required.

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Introduction

Ganglioglioma (GG) is usually a benign slow-growing neoplasm that mainly affects older children and young adults (Mpairamidis et al., 2008; Park et al., 2008). GG contains both glial and differentiated ganglion cells and accounts for approximately 0.4% of all central nervous system (CNS) tumours and 1–4% of pediatric CNS neoplasms (Mpairamidis et al., 2008; Park et al., 2008). It occurs most commonly in the supratentorial region; mostly in the temporal lobe (up to 85%) and presents with a long-standing intractable seizures (Mpairamidis et al., 2008; Park et al., 2008). Nevertheless, there are occasional reports of ganglioglioma occurring in the brainstem, cerebellopontine angle (CPA), thalamus, optic nerve, and spinal cord (Shin et al., 2002; Westwood and MacFarlane, 2009). The tumour size is variable, typically between 2–3 cm in adults, and larger (typically more than 4 cm) in children (Davis and Joglekar, 1981).

Here we report a rare case of the craniocervical ganglioglioma in a child and our experience in its management.

Case report

A 3.5-year-old male, presented with severe progressive quadriparesis, gait disturbance and sphincter deficit (urine and fecal incontinence) from about 1 month ago. At first, symptoms began with paraparesis, and gradually upper limbs weakness and gait disturbance added to the initial complaint. Past medical, drug and present patient history demonstrated no positive findings. On physical examination, the muscle strength force of upper limb was 4/5 on proximal and 3/5 on distal muscles on both sides. Lower limbs demonstrated the muscle strength force of 3/5 in the proximal and 2/5 in the distal. The upper motor signs such as

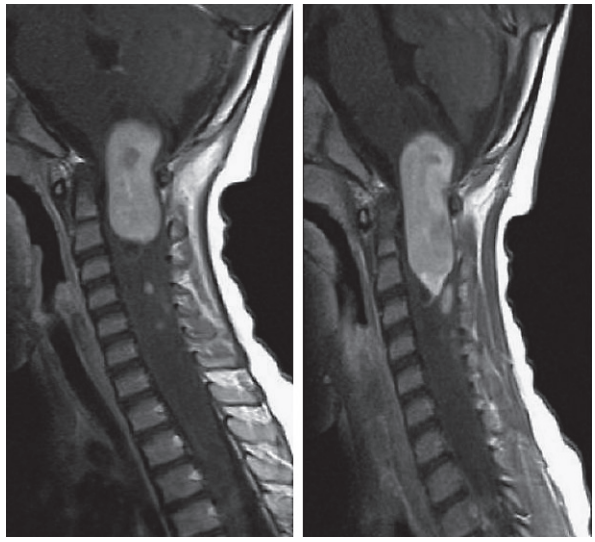

Figure 1 – The T1 with gadolinium injection image sequence demonstrates the tumour in cervicomedullary region.

Hoffman and Babinski sign, increased deep tendon reflexes (DTR), and clonus were positive on both sides. Moreover, the examination demonstrated the increased muscular tone and generalized spasticity. Magnetic resonance imaging (MRI) demonstrated an ill-defined mass within medulla and upper cervical spinal cord, which was hypo to iso signal on T1, heterogeneous iso to hypersignal on T2 and demonstrated marked bright enhancement on T1 with gadolinium (Gad) injection (Figure 1).

After whole spine and brain MRI evaluation, surgical intervention was planned. On Concord position, the head fixed on the Mayfield head holder. Suboccipital

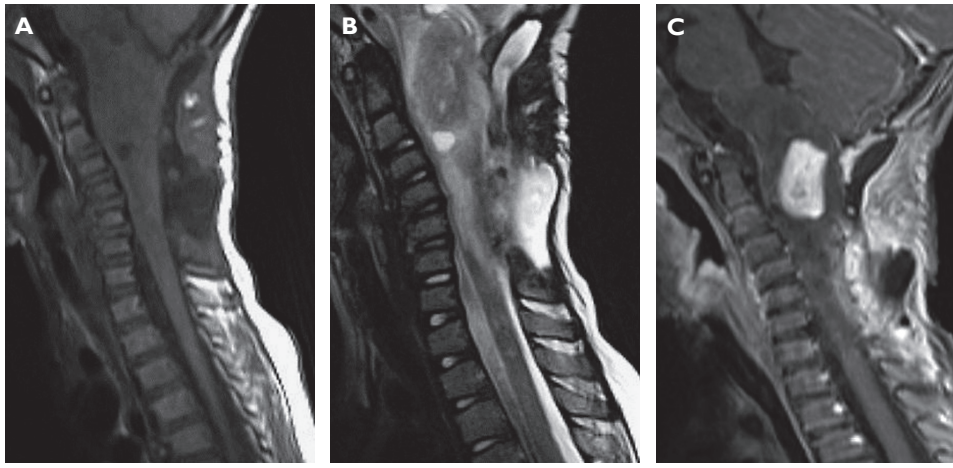

Figure 2 – The just postoperative magnetic resonance imaging demonstrates the tumour, which had only been decompressed (A – T1; B – T2; C – T1 after gadolinium injection).

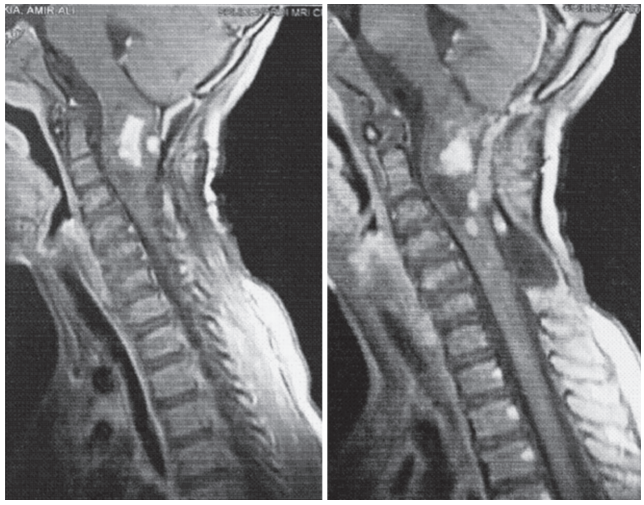

Figure 3 – The postoperative craniocervical magnetic resonance imaging fourteen months after surgery demonstrates no growth of the tumour.

craniectomy, C1 posterior arc laminectomy, and laminectomy of C2 to C6 performed. After opening the dura, posterior midline myelotomy performed. The mass had a soft texture, ill-defined border, and grey to brown appearance. A biopsy specimen sent for frozen section and reported a high-grade undifferentiated tumour. According to the frozen section report, and due to the absence of the tumour-neural parenchymal interference, only decompression of the tumour performed. Moreover, the dura was expanded via synthetic dural graft. The definite histopathologic examination was in favour of ganglioglioma (Figure 2).

After surgery, the neurological status partially improved, and the patient discharged, while he demonstrated a muscle strength force of 4/5 on all the limbs, he could to walk and showed improved sphincter control.

Due to benign nature of GG, and despite low volume surgical resection, the patient candidate for follow-up observation. On the last follow-up visit, 14 months after surgery, the patent has no complaint and neurological status improved. The craniocervical MRI demonstrated the tumour that did not grow (Figure 3).

Discussion

GG is a rare mixed glioneural tumour originating from neoplastic glial and neural cells. It consists 0.5–1.7% of all CNS neuroepithelial tumours, but up to 4% of CNS tumour in pediatrics. It is the most common tumour related to the intractable focal seizures (Mpairamidis et al., 2008; Park et al., 2008).

It has been seen throughout the CNS, but the majority of them occurred at supratentorial location (Gupta et al., 2014). At supratentorial location, the most common site is temporal, followed by frontal and medial parietal (I-Hao et al., 2003; Gupta et al., 2014). The posterior fossa is an uncommon site for GG, and brain stem and spinal cord are rare sites (I-Hao et al., 2003; Gupta et al., 2014).

GG of the brain stem is rare, is more common in pediatrics and young adult, and has an affinity to the medullary region. If it is diagnosed early, it has a more favourable prognosis than the cases with delay diagnosis (Blatt et al., 1995).

In the supratentorial location, the long-term intractable seizure is the most common symptom, but in the case of posterior fossa GG, symptoms include focal neurologic deficits (FND), cranial nerve (CN) deficits, and gait and speech disturbance (Kim et al., 2014).

GG located within cerebral lobe is well circumscribed, so it can be totally resected. Nevertheless, GG located in cerebellum and spinal cord has a poorer outcome, probably due to the site of the tumour or perhaps innate variance in biologic behaviour of the tumour (Gupta et al., 2014). Five-year event-free survival (EFS) for brainstem GG is five times lower than from supratentorial cases (Pan et al., 2016).

GG has a benign course with minimal malignant transformation potential, but in the cases with midline tumour, the prognosis is poorer. Despite this, it has a more indolent course in compared to the other brain stem intrinsic tumours (Blatt et al.,

1995). Because this, the differentiation from other tumours is important (Kim et al., 2014). Although, the malignant transformation potential is low, it is related to the glial component (Mpairamidis et al., 2008).

Most tumours are cystic (70%) associated with mural nodules (Davis and Joglekar, 1981). Clinical, radiological, and even surgical findings cannot easily differentiate ganglioglioma from other differential diagnoses. The most important differential diagnosis includes dysembryoplastic neuroepithelial tumour (DNET), oligodendroglioma, low-grade glioma, pleomorphic xanthoastrocytoma (PXA), astrocytoma, ependymoma, low-grade astrocytoma (grade II), oligodendroglioma, medulloblastoma, choroid plexus papilloma, and neurocysticercosis (I-Hao et al., 2003; Mpairamidis et al., 2008).

Posterior fossa GG demonstrated iso to hypo signal intensity on T1, hypersignal on T2, and a range of no enhancement to marked enhancement on T1 with Gad injection. Moreover, malignant GG demonstrated more enhancement (Kim et al., 2014).

Surgery in cases with GG of brainstem and spinal cord is restricted, because total resection is impossible (Davis and Joglekar, 1981). GG of the brainstem has a shorter duration of symptoms, higher mortality, and shorter progression free survival (PFS) in compare to supratentorial or cerebellum GG (Janjua et al., 2017).

Whenever possible, gross total resection should be tried in the case of GG, as it is possible in supratentorial location, it is less problematic in the cerebellum, but it is challenging in the case of brainstem GG (Janjua et al., 2017). The prognosis relates to the extent of resection (EOR) and more EOR caused to more survival (Pan et al., 2016).

The GG is not radiosensitive and role of radiotherapy is controversial, it can be even harmful. Despite increased local tumour control, some authors suggested that it can cause malignant transformation of GG, especially when the patient is young (Si et al., 2013). Moreover, chemotherapy has a little effect to control tumour progression and has more complications (Si et al., 2013). But chemoradiation is reserved for the cases with recurrence after surgery or suspicious to the adjacent parenchymal infiltration (Mpairamidis et al., 2008).

Janjua and the others surveyed 142 cases with brainstem GG within five years. The mean age was 11.4 years with a range of 1 day to 59 years old. The most majority of patients were below 20 years old. The most common symptoms include dysesthesia, motor weakness, and lower cranial nerves deficits. The severity of enhancement decreased with increased age but without any relation to the sex. In this study, all of the medullary tumours had contrast enhancement. The majority of tumours occurred in pons/medulla, followed by medulla and then the only pons tumours. The midbrain was the less common site of involvement. About one-third of the patients had cervicomedullary junction involvement (Janjua et al., 2017).

Our patient demonstrated an intraaxial mass within the cervicomedullary junction that showed marked enhancement. Although this case demonstrated the

typical imaging features of GG, the preoperative definite diagnosis was impossible. The follow-up demonstrated the indolent nature of GG in our case, although, the duration of follow-up is too short for the conclusion.

Conclusion

Although it is rare, the GG should be considered in the differential diagnoses of tumours with compatible clinical and radiologic features even in the unusual locations, especially in the pediatric and young patients. Whenever possible, safety surgical resection should be considered in those patients. In the case of partial resection, that is common in the tumours located within functionally critical structures, long close follow-up rather than radiation therapy is required.

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