

CASE REPORT

'Intramedullary cervical spinal cord ganglioglioma, review of the literature and therapeutic controversies'

I Lotfinia¹ and P Vahedi²

¹Department of Neurological Surgery, Tabriz University of Medical Science, Tabriz, Iran and ²Department of Neurological Surgery, Tabriz University of Medical Sciences, Tabriz, Iran

Study design: Case report with comprehensive review of literature.

Settings: Department of Neurosurgery, Shohada Hospital, Faculty of Medicine, Tabriz University of Medical Sciences, Daneshgah Street, Tabriz, East Azerbaijan, Iran.

Report: A 22-year-old female presented with neck pain, which was worse at night and recumbency, and intermittent paresthesia in upper limbs. MRI revealed an intramedullary cervical tumour with a syrinx within the upper cervical cord and medulla. Total resection of tumour with laminoplasty and duraplasty was done. Pathology confirmed ganglioglioma of the spinal cord. A year after surgery, the patient has no complaint. Physical exam reveals no abnormality. MRI with contrast reveals no recurrence.

Conclusion: No general consensus exists on the management of intramedullary spinal ganglioglioma. MRI might be non-specific; however, some may be characteristic to differentiate it from other intramedullary tumours. The tumour responds optimally to resection, and every attempt should be made to perform a total surgical resection. The role of adjuvant therapy remains controversial.

Spinal Cord (2009) 47, 87–90; doi:10.1038/sc.2008.69; published online 17 June 2008

Keywords: ganglioglioma; intramedullary; cervical; chemotherapy; radiotherapy; imaging

Introduction

Ganglioglioma contains both neoplastic astrocytes and neural cells.¹ It is a rare tumour constituting 0.4–6.25% of all primary central nervous system (CNS) neoplasms.¹

Ganglioglioma may occur throughout CNS, predominantly in the supratentorial area. Spinal cord ganglioglioma is a very rare entity (1% of all intramedullary tumours).¹

Radiological characteristics and therapeutic interventions are matters of controversy.

Case report

A 22-year-old female presented with neck pain, paresthesia in the upper limbs, mild quadriparesis (4/5), increased deep tendon reflexes (3+) in four limbs and Hoffmann and Babinski signs.

Magnetic Resonance Imaging (MRI) revealed an intramedullary cervical tumour with a syrinx extending to the medulla (Figure 1). The tumour was of mixed signal intensities on T1 and hyperintense on T2. Homogeneous enhancement was evident (Figure 1).

Bilateral open door laminoplasty was performed. Dura was opened in the midline and the oedematous cord appeared.

After midline myelotomy, a dark red tumour with relatively few vessels appeared. Despite a well-defined border, the tumour was adhesive to the normal cord and resected gross totally under surgical microscope. Dura was closed tightly without myeloplasty. Laminoplasty was done applying titanium microplates.

Histopathological exam revealed ganglion cells with astrocytic components, in favour of ganglioglioma. (Figure 2). MIB-1 was positive.

Postoperatively, weakness became more severe in the right arm and leg (I/V and II/V, respectively). The patient was able to walk on the second day. After the fourth day, arm weakness gradually improved and she was discharged and referred to a physical therapist on the sixth day. Six months later, the patient was able to write and the force of the four limbs became 4/5. MRI with contrast showed no obvious mass lesion (Figure 3).

Discussion

Ganglioglioma was first introduced as a CNS neoplasm by Ewing in 1928. Only 1.1% of all spinal tumours show this pathology.

The most commonly affected areas are the temporal lobe and cerebellum and the least affected is the spinal cord with a predilection for the cervical region—(Table 1).

Correspondence: Dr P Vahedi, Department of Neurological Surgery, Imam Hospital, Tabriz University of medical Sciences, Tabriz, East Azerbaijan 5166614756, Iran.

E-mail: Payman.vahedi@gmail.com

Received 21 January 2008; revised 9 April 2008; accepted 7 May 2008; published online 17 June 2008

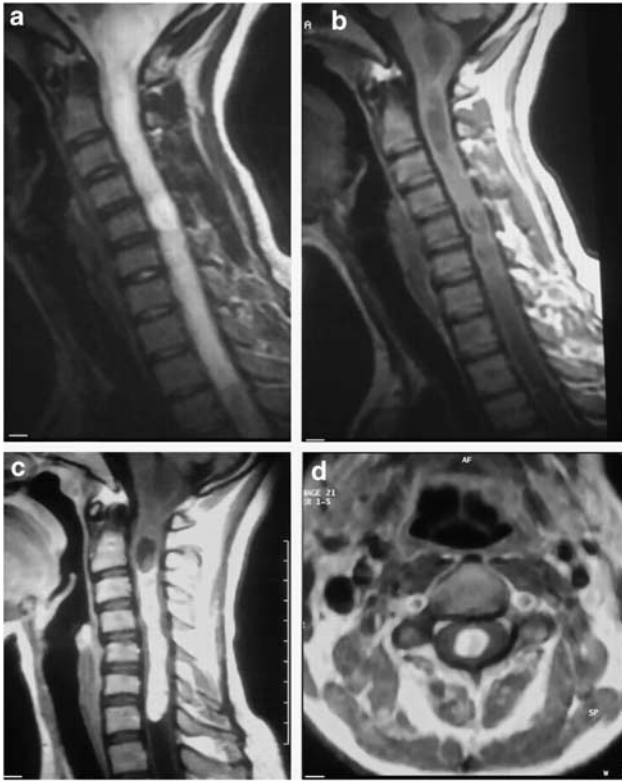


Figure 1 (a and b) Sagittal T1- and T2-weighted MRI demonstrating intramedullary tumour extending from C2 to C7. The lesion is iso to hypersignal on T1 and hypersignal on T2 images, respectively. (c and d). Sagittal and axial post-Gadolinium T1-weighted MRI delineating the extension of homogeneously enhancing intramedullary tumour from C2 to C7. A syrinx is also noted at the level of C2.

Children are more affected than adults. No gender predominance is seen for spinal tumours.¹

The most common presenting symptom is back pain and limb weakness. Spinal lesions are found earlier than cerebral lesions.

Radiological characteristics are not pathogenomic.¹ MRI is the diagnostic modality of choice. Ganglioglioma might be hyperintense, isointense, hypointense or heterogeneous on T1; it is mostly hyperintense on T2 and mostly enhances heterogeneously with contrast.

Total surgical resection remains the standard treatment. Preoperative radiological presumption of ganglioglioma is of clinical importance. Usually, more than eight segments of the spinal cord are involved, and its length is two times that of astrocytoma and ependymoma. Holospinal involvement, scoliosis and bone remodelling, mixed intensity on T1, significant cysts or syrinx, absence of oedema and enhancement within the tumour centre, hemosiderin or calcification and young age help in diagnosis.² The higher intensity of fluid contents of cysts, irregular margins with soft tissue compartment and gadolinium enhancement help in differentiating them from syrinx.

The literature only provides five cases of malignant ganglioglioma.³ This risk is 10% for intracranial gang-

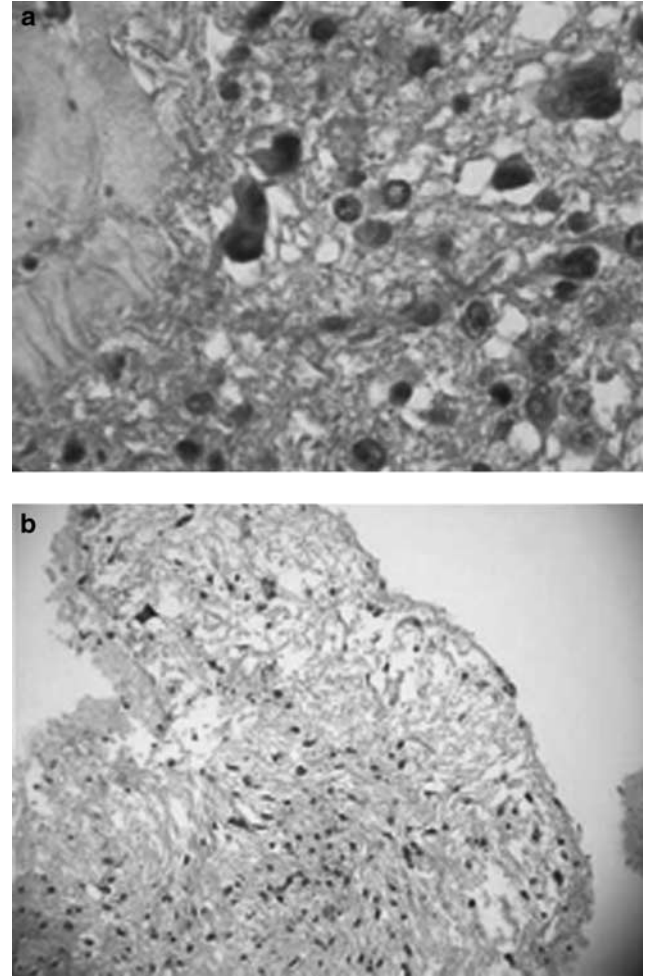


Figure 2 (a) Photomicrograph of ganglioglioma showing abnormal clustering, irregular distribution and cytotologic abnormalities of ganglion cells such as multinucleation. Delicate fibrillar material is evident Hematoxylin and Eosin original magnification $\times 40$. (b) Mature multinucleated ganglion cells with surrounding schwann-like cells. Hematoxylin and Eosin original magnification $\times 10$.

lioglioma, mostly within the glial component, but unclear for spinal tumours; however it is rare for cerebellar tumours.

That total surgical resection is the treatment of choice for ganglioglioma is accepted (Table 1). Early surgical intervention follows two main advantages: first, surgery with better functional status. Second, easier resection due to smaller size. Some argue that complete resection might not be feasible because no well-defined plane of cleavage could be identified. Hamburger *et al.*¹ believe that it carries a high morbidity rate even with microsurgical techniques.

The extent of resection is the main factor to determine the outcome; however, some disagree. Surgical outcome reports include recovery, no change in neurological status, and progression of neurological deficit.⁴ The recurrence is more in spinal ganglioglioma than in cerebral counterparts.⁴ Close follow-up is necessary. It is suggested that patients be seen every 6 months for 2 years postoperatively, and annually thereafter.⁵

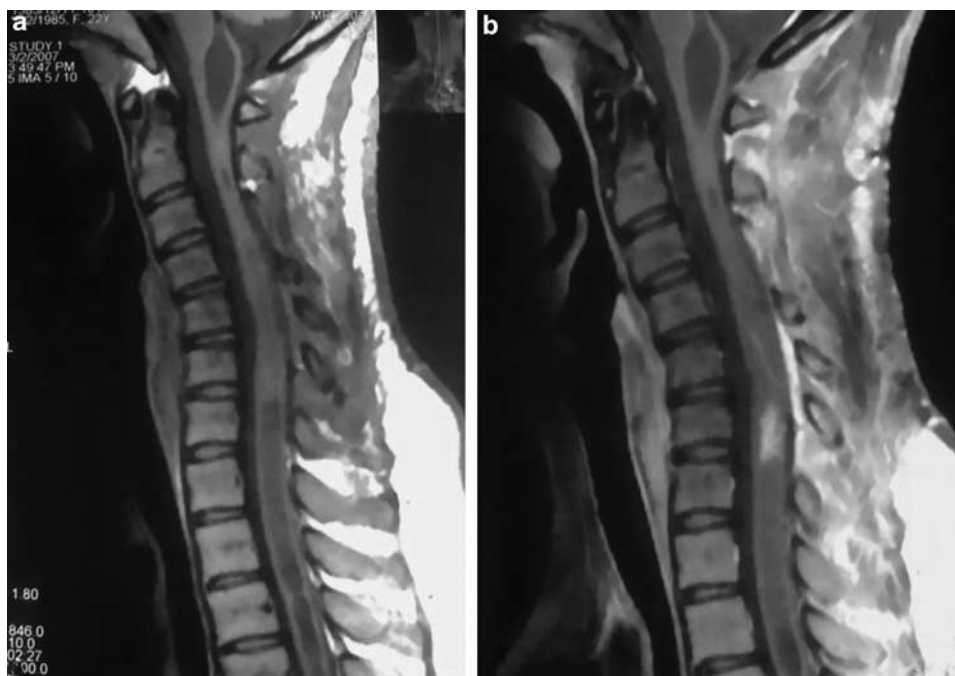


Figure 3 (a and b) Sagittal postoperative T1-weighted MRI with and without contrast showing a small remainder of tumour at the level of C7. Note the syrinx extending from the medulla oblongata down to the level of C2.

Table 1 Comprehensive list of reported cases of spinal intramedullary ganglioglioma with therapeutic options

Series/ref	Year	Age (year)/sex	Location	Symptoms/signs	Treatment	Recurrence
Pick <i>et al.</i>	1911	24/F	C1–C2	Singultus	None	—
Foerster	1924	24/M	C4–C6	Autopsy finding	None	—
Kernohan	1932	27/F	Cervical	Arm pain and weakness	Op, Rad	None
		34/F	Cervical	Arm leg and weakness	Op	None
Lichtenstein <i>et al.</i>	1937	20/M	Thoracic	Paraplegia, incontinence	None	—
Garrido <i>et al.</i>	1978	2.5/F	Cervicothoracic	Not mentioned	Op, Rad	None
		4/F	Cervicothoracic	Not mentioned	Op	None
		10/M	Cervicothoracic	Not mentioned	Op, Rad, Chemo	None
Bevilacqua <i>et al.</i>	1979	78/M	T3–T6	Paraparesis, gait problem	Op, Rad	5 Years
Albright <i>et al.</i>	1980	11/M	Holocord	LBP, gait problem	OP	None
Johannsson <i>et al.</i>	1981	12/F	C7–T6	Paraparesis	OP, Rad	Not mentioned
		80/M	Not mentioned	Autopsy finding	None	—
Nass <i>et al.</i>	1981	Child/F	Conus medullaris	Gait disturbance	OP	None
Wald <i>et al.</i>	1985	2.5/M	T10–T12	Gait disturbance	OP	None
Haridson <i>et al.</i>	1987	Child	Not mentioned	Not mentioned	Op, Rad, Chemo	13 months
Kitano <i>et al.</i>	1987	13/F	Holocord	Paraparesis	Biopsy, Rad	6 Years
Rodewald <i>et al.</i>	1987	17/M	T10–Cauda equina	Foot drop	Op, Rad, Chemo	Persistent
Pialat <i>et al.</i>	1987	25/F	C2–T8	Paraplegia, incontinence	Op	2 Years none
Bell <i>et al.</i>	1988	3/M	C2–C7	Hemiparesis	Op, Rad, Chemo	Persistent
Herrmann <i>et al.</i>	1988	41/M	T9–T12	Not mentioned	Op	Not mentioned
Russell <i>et al.</i>	1989	6 cases	Not mentioned	Not mentioned	Not mentioned	Not mentioned
Miller <i>et al.</i>	1990	5/M	Not mentioned	Hydrocephaly	Not mentioned	Not mentioned
Pereti-viton <i>et al.</i>	1991	7/F	T10–T12	Paraparesis	Op	5 and 6.5 Years
Haddad <i>et al.</i>	1992	7/F	Thoracic	Paraparesis	Op, Rad	Persistent
		8/M	Conus medullaris	Paraparesis	Op	None
Mickle	1992	3 children	Not mentioned	Not mentioned	Op, Rad	Not mentioned
Otsubo <i>et al.</i>	1992	6 children	Cervical (<i>n</i> = 1)	Motor weakness (<i>n</i> = 5)	Op	None
			Cervicothoracic (<i>n</i> = 3)	Gait disturbance (<i>n</i> = 4)	Op	None
			Thoracolumbar (<i>n</i> = 2)	Sensory disturbance (<i>n</i> = 2)	Op	None
			Cervicomedullary (<i>n</i> = 8)	Pain	Op	14/30
Lang <i>et al.</i>	1993	56 Cases 7 month to 25 years old/ M/F 1.7/1	Cervical <i>n</i> = 7	Paraparesis	Op, Rad	
			Cervicothoracic <i>n</i> = 21		Op, Chemo	

Table 1 Continued

Series/ref	Year	Age (year)/sex	Location	Symptoms/signs	Treatment	Recurrence
Park <i>et al.</i>	1993	6.5/M 3/F	Holocord Medulla-T3	Motor weakness Scoliosis Head tilt, Right paresis	Op Op	9 Years/None None
Hamburger <i>et al.</i> ¹	1997	4/F	C2-T2	Weakness, gait disturbance	Op	1 Year/None
		54/F	T12-L2	Weakness, gait disturbance	Op	2 Years/None
Kurtsoy <i>et al.</i>	1997		T12-L2		Op	
Sawin <i>et al.</i>	1999	2/F	Medulla-C4	Quadriparesis	Op	1 Year/None
Park <i>et al.</i>	2000	4/M	C5-T1	Hemiparesis	Op	1 Year/None
		17/M	T2-T3	Paraparesis	Op	None
		26/F	T8-T9	Scoliosis, gait disturbance	Op	Not mentioned
Jallo <i>et al.</i>	2004		Thoracic <i>n</i> = 16 Conus <i>n</i> = 4			30% recurred
Satyarthee	2004	40/M 7/M	C1-T2 C1-T3	Quadriparesis Quadriparesis	Op Op	Not mentioned Not mentioned
Costa <i>et al.</i>	2006	13/M	Conus medullaris	Paraparesis	Op	years/None
Jeffs <i>et al.</i>	2006	17/M	T8-Conus medullaris	Paraparesis	Op	Not mentioned
Karabekir <i>et al.</i>	2006	2/M	T9-L3	Gait disturbance	Op, Rad	2 Years/None
Amini <i>et al.</i>	2007		Conus medullaris	Paraparesis	Op	Not mentioned
Present case	2007	22/F	C2-C7	Motor weakness, Paresthesia	Op	1 Year/None

Abbreviations: Chemo, Chemotherapy; Op, operated; Rad, Radiotherapy.

Routine radiotherapy is not suggested;¹ however, no clinical trial exists. The reasons include the risk of differentiation of the astrocytic component into glioblastoma (Table 1), the probability of irradiation necrosis and neurological compromise. Radiotherapy is considered for cases in which the radical resection of tumour is not feasible, when progression is evident and for malignant histopathology.

The role of chemotherapy remains indefinite. In recurrence, surgery should be considered; however, radiotherapy might be another option. After the second surgery, adjuvant therapy should be considered.⁵

Conclusion

Intramedullary spinal ganglioglioma is a rarely discussed entity. MRI findings might be non-specific. Some characteristics may help in differentiation and are useful in planning

the surgery. Total resection is the standard treatment. The role of adjuvant therapy remains controversial.

References

- Hamburger C, Büttner A, Weis S. Ganglioglioma of the spinal cord: Report of Two Rare Cases and Review of the Literature. *J Neurosurgery* 1997; **41**: 1410-1416.
- Patel U, Pinto R, Miller D, Handler M, Rorke L, Epstein F *et al.* MRI of spinal cord ganglioglioma. *AJNR Am J Neuroradiol* 1998; **19**: 879-887.
- Rodewald L, Miller D, Sciorra L, Barabas G, Lee ML. Central nervous system neoplasm in a young man with martin-bell syndrome: fra(X)-XLMR. *Am J Med Genet* 1987; **26**: 7-12.
- Lang FF, Epstein FJ, Ransohoff J, Allen JC, Wisoff J, Abott IR *et al.* Central nervous system ganglioglioma part 2: clinical outcome. *J Neurosurg* 1993; **79**: 867-873.
- Jallo G, Freed D, Epstein F. Spinal cord gangliogliomas: a review of 56 patients. *J of Neuro-Oncology* 2004; **68**: 71-78.