Case Report

Spinal glioblastoma with brain relapse in a child: clinical considerations

E Caroli^{*1}, M Salvati² and L Ferrante¹

¹Department of Neurological Sciences, Neurosurgery-Ospedale S. Andrea-University of Rome 'La Sapienza', Rome, Italy; ²Department of Neurosurgery, INM Neuromed IRCCS, Pozzilli (Is), Italy

Study design: Case report.

Objectives: To describe a child with intramedullary glioblastoma at T₉–T₁₀–T₁₁, and to discuss the clinical features of this rare pathology.

Setting: Department of Neurological Sciences, Italy.

Case report: Spinal cord glioblastoma in children has only rarely been reported. It most frequently involves the thoracic region with a predilection for the second and third decades of life. This report describes one case of thoracic glioblastoma multiforme in a 6-year-old child and reviews other cases reported in the literature.

Results: Laminectomy and excision of the tumour were performed. Postoperative radiotherapy and chemotherapy were given, but 4 months later the patient presented with a brain relapse of the tumour. At 9 months after diagnosis the patient died from cerebral tumour regrowth.

Conclusions: Full neuraxis MRI is always recommended in order to detect possible metastases. The prognosis after multimodality therapy (surgery, radiotherapy, chemotherapy) remains poor. From the literature, only four cases of paediatric patients with long-term survival have been reported.

Spinal Cord (2005) 43, 565–567. doi:10.1038/sj.sc.3101749; published online 19 April 2005

Keywords: children; glioblastoma; intramedullary; radiotherapy; spinal cord tumour; surgery

Introduction

Primary spinal cord tumours in children are rare. In patients under 15 years of age, the ratio of brain to spinal cord tumours ranges from 8:1 to 22:1.^{1,2} Most intramedullary tumours in children are astrocytomas (47–88%) followed by ependymomas (24–37%),^{1–5} other tumours are uncommon. In particular, intramedullary glioblastomas are rarely described. These tumours occur mainly in the cervico-thoracic segments and have a short clinical history before diagnosis.⁶ The aim of this paper is to report one case of a 6-year-old boy with intramedullary glioblastoma that spread to brain and to discuss the clinical, radiologic and therapeutic findings of these tumours with a review of the pertinent literature.

Case report

A 6-year-old boy presented with a 4-month history of increasing weakness of the lower limbs. Neurological examination disclosed severe spastic paraparesis and hypoaesthesia to touch, temperature and pain with an upper border at T₁₀. A magnetic resonance imaging scan of the spinal cord revealed an intramedullary expanding

lesion (Figure 1a) with contrast enhancement (Figure 1b) from T₉ to T₁₁. Full neuraxis magnetic resonance imaging (MRI) revealed no other lesions. Surgery disclosed an intramedullary invasive necrotic tumour extending from T₉ to T₁₁; subtotal removal of the lesion and dural decompression were performed. The histological examination of the excised tumour revealed a glioblastoma multiforme. The patient's post-operative course was uneventful. The patient received radiotherapy (40 Gy to the entire spinal axis with LINAC) and chemotherapy (PCV). After 4 months, he was admitted to our institution for headache and vomiting. A brain MRI showed a ring enhancing mass in the left frontal lobe (Figure 2a, b). An MRI showed no other intramedullary lesions. The patient underwent a craniotomy. The tumor was subtotally removed (Figure 2c). The histological examination was consistent with a glioblastoma multiforme. The patient died 9 months later from cerebral tumor regrowth.

Discussion

Spinal cord tumours in children are quite rare. They represent approximately 4% of childhood central

*Correspondence: E Caroli, Via Meropia 85, 00147 Rome, Italy

nervous system tumours.^{1,4} The majority are astrocytomas (47–88%), followed by ependymomas (27–37%). Intramedullary glioblastoma is uncommon in all ages of life, but in children are extremely rare.^{6–8}

Intramedullary glioblastoma shows a slight predilection for the second and third decades of life, the mean age ranging from 28.5 years in the 20-case series of the Mayo Clinic⁹ to 18.1 years according to the review of the literature undertaken by Fortuna in 1971.¹⁰ Our patient was a 6-year-old. The clinical presentation of our case was very rapid, with a duration of symptoms of only 4 months. This is consistent with previous studies of malignant spinal tumours.^{6,11} Spinal cord glioblastomas occur mainly in the cervical or thoracic segments.^{9,10,12–19} In our case glioblastoma was localised at T₉–T₁₁. Spinal cord glioblastoma shows a tendency to spread via the subarachnoid space at the spinal and, more rarely, at the brain level.^{6,13,20,21} This can be

explained by the proximity of the neoplastic cells to the cerebrospinal fluid pathways.^{6,13,14} However, the possibility of surgical dissemination of glioblastoma through increased exposure of the subarachnoid spaces to the malignant glial cells cannot be excluded.^{6,13} Also, the presence of multiple foci should be considered.^{6,10,13,14} Seeding of a spinal cord glioblastoma to the brain is extremely rare.^{6,13,20} In our case, a cerebral relapse occurred 4 months after the diagnosis of a spinal tumour. In the similar cases reported in the literature, the interval between the onset of spinal cord symptoms and the onset of intracranial symptoms varied from 1 week to 1 year.^{6,18,21–23}

MRI is the diagnostic procedure of choice for spinal tumours.²⁴ In the presented case, MRI was able to accurately define the tumour extending from T₉ to T₁₁. Full neuraxis MRI is always recommended in order to detect possible metastases.

As these tumors are rare, reliable information on therapy and prognosis is limited. The role of surgery in the management of intramedullary tumours of the spinal cord has been expanded in recent years. Until recently, surgical management of an intramedullary mass consisted of biopsy, or cyst aspiration with decompressive laminectomy.^{25,26} Recent advances in imaging and microsurgical technique have established microsurgical removal as the single most effective treatment for benign intramedullary tumours.^{2,4,26} Surgical removal in patients with intramedullary glioblastoma cannot be radical because there is not a clear cleavage plane between the glioblastoma and the medullary parenchyma. However, surgery should be 'reasonably aggressive' so that the tumour bulk is reduced for radiation therapy. Our patient had no adverse sequelae from the subtotal resection of its tumour. Because of the high incidence of postoperative spinal deformities following laminectomy in paediatric patients, laminoplasty should be performed in spinal surgery.^{2,26–28} However, the onset of deformity ranges from 8 months to 7.5 years. Intramedullary glioblastoma is a fatal disease with so short survival time that iatrogenic spinal deformity does not occur. Dural decompression may be an advisable procedure.

Figure 1 Sagittal T₁-weighted MRI without gadolinium (a) and with gadolinium (b) showing the enhancing spinal cord tumour (arrow)

Figure 2 Axial T₁-weighted MRI without gadolinium (a) and with gadolinium (b) showing an enhancing left frontal lesion. Immediate postoperative MRI shows a small tumour residue (c, arrow)

The value of radiotherapy in spinal cord malignant tumours is a matter of debate.^{4,3,6,12-16,18,19} Ciappetta et al⁶ in their series of six adult patients and one paediatric patient reported a beneficial effect of radiotherapy. Moreover, one should take into account that survival time is so short that the late effects of radiotherapy will never be observed. Only four series of paediatric patients with spinal cord glioblastoma have reported long-term survivors.^{12,15,18,24,29} These cases represent an exception. Prognosis in patients with spinal cord glioblastomas remains poor. The value of chemotherapy is unknown. In our patient, we have performed a multimodality therapy (surgery, radiotherapy and chemotherapy) as for the intracranial glioblastomas, observing a survival time of 9 months.

References

- Farwell JR, Dohrmann GJ, Flamery JT. Central nervous system tumors in children. *Cancer* 1977; **40**: 3123-3132.
- Reimer R, Onofrio BM. Astrocytomas of the spinal cord in children and adolescents. *J Neurosurg* 1985; **63**: 669-675.
- Cooper PR, Epstein F. Radical resection of intramedullary spinal cord tumor in adults. *J Neurosurg* 1985; **63**: 491-492.
- Epstein F, Epstein N. Intramedullary tumors of the spinal cord. In: Section of Pediatric Neurosurgery of the American Association of Neurological Surgeons (eds). *Pediatric Neurosurgery. Surgery of the Developing Nervous System*. Grim & Stratton: New York 1982, pp 529-533.
- Garret PG, Simpson WJK. Ependymomas: results of radiation treatment. *Int J Radiat Oncol Biol Phys* 1983; **9**: 1121-1124.
- Ciappetta P, Maurizio S, Capoccia G, Artico M, Raco A, Fortuna A. Spinal glioblastoma: report of seven cases and review of the literature. *Neurosurgery* 1981; **28**: 302-306.
- O'Sullivan C, Jenkin RD, Doherty MA, Hoffman HJ, Greenberg ML. Spinal cord tumors in children: long-term results of combined surgical and radiation treatment. *J Neurosurg* 1994; **81**: 507-512.
- Nadkarni TD, Rekate HL. Paediatric intramedullary spinal cord tumors. Critical review of the literature. *Child's Nerv Syst* 1999; **15**: 17-28.
- Russel DS, Rubinstein LJ. *Pathology of Tumors of the Nervous System*. Edward Arnold: London 1989, pp 219-247.
- Fortuna A, Giuffrè R. Intramedullary glioblastoma. *Neurochirurgia (Stuttg)* 1971; **14**: 14-23.
- Bouffet E, Pierre-Kahn A. Prognostic factors in pediatric spinal cord astrocytoma. *Cancer* 1998; **83**: 2391-2399.
- Alvisi C, Cerisoli M, Giuliani M. Intramedullary spinal gliomas: long term results of surgical treatment. *Acta Neurochir (Wien)* 1984; **70**: 169-179.
- Cohen AR, Wisoff JH, Allen JC, Epstein F. Malignant astrocytomas of the spinal cord. *J Neurosurg* 1989; **70**: 50-54.
- Grisold W, Pernetzky G, Jellinger K. Giant cell glioblastoma of the thoracic cord. *Acta Neurochir (Wien)* 1981; **58**: 121-126.
- Guidetti B, Mercuri S, Vagnozzi R. Long-term results of the surgical treatment of 129 intramedullary spinal gliomas. *J Neurosurg* 1981; **54**: 323-330.
- Guidetti B, Fortuna A, Moscatelli G, Riccio A. I tumori intramedullari. *Lav Neuropsichiatri* 1964; **35**: 409.
- Helseth A, Mork SJ. Primary intraspinal neoplasms in Norway, 1955 to 1986. A population-based survey of 467 patients. *J Neurosurg* 1989; **71**: 84-85.
- Kopelson G, Lingood RM. Intramedullary spinal cord astrocytoma versus glioblastomas. The prognostic importance of histologic grade. *Cancer* 1982; **50**: 732-735.
- Malis LI. Intramedullary spinal cord tumors. *Clin Neurosurg* 1978; **25**: 512-539.
- Bell WO et al. Leptomeningeal spread of intramedullary spinal cord tumors. Report of three cases. *J Neurosurg* 1988; **69**: 295-300.
- Andrews AA, Enriques L, Renaudin J, Tomiyasu V. Spinal intramedullary glioblastoma with intracranial seeding. *Arch Neurol* 1978; **35**: 244-245.
- Johnson DL, Schwarz S. Intracranial metastasis from malignant spinal cord astrocytoma. *J Neurosurg* 1978; **66**: 621-625.
- Eade OE, Urich H. Metastasizing gliomas in young subjects. *J Pathol* 1971; **103**: 245-256.
- Kulkarni A, Armstrong DC, Drake JM. MR characteristics of malignant spinal cord astrocytomas in children. *Can J Neurol Sci* 1999; **26**: 290-293.
- Woltman HW et al. Intramedullary tumors of the spinal cord and gliomas of intradural portion of filum terminale: fate of patients who have these tumors. *Arch Neurol Psychiatry* 1951; **65**: 378-393.
- Kaufman BA, Park TS. Congenital spinal cord astrocytomas. *Child's Nerv Syst* 1992; **8**: 389-393.
- Gutierrez FA, Oi S, McLone DG. Intraspinal tumors in children: clinical review, surgical results and follow-up in 51 cases. *Concepts Pediatr Neurosurg* 1983; **4**: 291-305.
- Vles JSH, Grubben CPM, van Ooy A, Weil EHJ. Holocord astrocytomas in childhood. *Clin Neurol Neurosurg* 1990; **9**: 361-364.
- Przybylski GJ, Albright AL, Martinez AJ. Spinal cord astrocytoma: long-term results comparing treatments in children. *Child's Nerv Syst* 1997; **13**: 375-382.