Short Report

Intramedullary Tumours at King Khalid University Hospital

Abdulhakim B Jamjoom; FRCS Ed (SN)*, Zain Alabedeen B Jamjoom, MD*, Naim-Ur-Rahman; FRCS Ed (SN)*, Sherif El-Watidy; FRCS (Ed), Mohammed Al-Sohaibani, FRCP**

From the Division of Neurosurgery and the Department of Pathology,** King Khalid University Hospital, Riyadh, Saudi Arabia*

Abstract - *A retrospective analysis of 16 consecutive cases of intramedullary lesions treated at King Khalid University Hospital between 1985-1995, was conducted to provide data regarding the characteristics of patients with intramedullary tumours in Saudi Arabia in comparison with the existing literature. Ninety four percent of the patients were unable to walk independently at presentation. This series is unusual in that it included a case of intramedullary tuberculoma, in the low incidence of low grade astrocytoma (13%), the high incidence of malignant astrocytoma (25%) and the relatively high incidence of tumour located in the thoracic (31%) and cervicomedullary region (19%). Our 4 malignant astrocytoma patients were unique in that two showed growth of their neoplasm into the medulla, one had a multicentric tumour and the other developed neuraxial dissemination. At a median follow-up of 12 months, 6 of the 13 survivors had a "favourable" outcome in that they were able to function and ambulate independently. This study shows that there is a slight difference in the characteristics of intramedullary tumours in Saudi Arabia compared to what is reported in literature. Patients with intramedullary tumours should be diagnosed by MRI and referred early to specialised neurosurgical units capable of performing a tumour resection as completely as possible.*

Key Words: Astrocytoma, Ependymoma, Intramedullary tumours, Saudi Arabia, Spinal cord tumours.

Intramedullary tumours are infrequent challenging lesions that account for 6.8-19.3% of all spinal tumours[1]. They have attracted interest in recent years because more of these neoplasms can now be excised radically with good results. This has been facilitated by the ability to accurately localise these tumours by MRI and by the utilisation of microsurgical techniques, Cavitron Ultrasonic Surgical Aspirator (CUSA) and the CO_2 laser. The characteristics of the various tumours that occur within the spinal cord are well documented in the literature, however, data about their incidence, pattern and management results in Saudi Arabia or the Middle East remains lacking. This article is a survey of 16 cases of intramedullary lesions treated over a decade in a neurosurgical unit in Saudi Arabia. This tumour is rare and most of the intramedullary tumour series published in the international literature over the last 6 years ranged from 15-86 patients[2-6]. The aim of the article is to highlight the clinical and pathological features of patients with intramedullary tumours in Saudi Arabia compared to what is documented in the literature.

Subjects and Methods

This is a retrospective analysis of 16 consecutive cases of intramedullary lesions that were treated at King Khalid University Hospital (KKUH) between 1985-1995. One case with an intramedullary tuberculoma was also included in the study. Each case was analysed by using hospital records and the following data were collected: age, sex, clinical presentation, histological diagnosis, radiological findings, level of the lesion, extent of surgical treatment and other treatment modalities. The clinical and functional grades of the patients were assessed preoperatively, immediately postoperatively and at follow-up using the scheme described by McCormick, *et al*[7] which is:

Grade I: Neurologically normal; mild focal deficit not significantly affecting function of involved limb, or gait.

Grade II: Sensorimotor deficit affecting function of involved limb; mild to moderate gait difficulty; severe pain impairing patient's quality of life; nevertheless functions and ambulates independently.

Grade III: More severe neurological deficit; requires assistance to ambulate, or has significant impairment in the upper limb; may or may not function independently.

Grade IV: Severe deficit; requires wheelchair; usually not independent.

At follow-up, the surgical outcome was considered "favourable" for patients in grade I and II and "non-favourable" for patients who died or were in grades III and IV.

Results

The distribution of the cases according to age, sex, tumour histology, tumour location, extent of excision, treatment with radiotherapy, and the clinical grade before surgery and at follow-up are summarised in Table 1. The duration of symptoms was 1 month to 7 years (median 14 months). One patient (case 12) was being treated for tuberculous meningitis when the intramedullary tuberculoma was diagnosed. All patients had evidence of motor and sensory disturbances at the time of presentation. Seven (44%) patients had local or referred

Table 1 - Characteristics of Patients with Intramedullary Tumours Presenting at King Khalid University Hospital, Saudi Arabia.

Case	Age (Yrs) & Sex	Histology	Location	Excision*	Clinical & Functional Grade[1] Pre-Oper	At follow-up (months)
1	21 M	Ependymoma	T11-L4	T	III	I(60)
2	19 M	Ependymoma	T11-L2	ST(R)	III	III(12)
3	50 M	Ependymoma	T12-L1	T	III	I (36)
4	36 M	Ependymoma	T11-L2	T	IV	II (12)
5	56 F	Ependymoma	C5-7	T	IV	III(12)
6	11 F	Malig. astrocytoma	T8-9	ST(R)	IV	Died(4)
7	18 M	Malig. astrocytoma	Medulla-C7	ST	IV	Died(2)
8	16 M	Malig. astrocytoma	Medulla-C5	ST(R)	IV	Died(4)
9	55 F	Malig. astrocytoma	C3-T2	ST(R)	IV	IV(6)
10	20 M	Low grade astrocytoma	C1-7	T	III	II (12)
11	13 M	Low grade astrocytoma	C5-L2	B(R)	III	III(6)
12	30 M	Tuberculoma	T3-4	B	IV	IV(6)
13	¾ F	Lipoma	Medulla-T2	ST	IV	IV(36)
14	35 F	Haemangioblastoma	T2-5	B	IV	III(12)
15	43 F	Neurofibroma	T1-4	T	III	II(18)
16	19 F	Cavernoma	T6-8	ST	II	II(6)

*T – Total; ST = Subtotal; B = Biopsy; R – Radiotherapy

pain while 9 (56%) patients had urinary dysfunction. Four of the latter patients were in urinary retention at presentation. Six (37%) patients, who were treated early in the series, were investigated by CT myelography while ten (63%) patients were investigated by MRI. In 5 of the latter patients (2 ependymoma, 1 neurofibroma, 1 covernoma, 1 haemangioblastoma), the MRI demonstrated a syrinx associated with the intramedullary tumour.

Five patients in this series had intracranial involvement. In three cases*, there was direct spread of the cervical tumour to involve the medulla *(Fig. 1)*. Case 6, who was diagnosed to have malignant astrocytoma at T8-9, had been treated for frontoparietal malignant astrocytoma a year prior to the diagnosis of her spinal lesion. Case 9 presented initially with symptoms of raised intracranial pressure and found on the CT scan to have hydrocephalus which was thought to be due to aqueduct stenosis. Six months later, the patient became quadriparetic and the MRI revealed the cervical intramedullary tumour which had disseminated intracranially obstructing the aqueduct *(Fig. 2a,b)*.

Surgery was performed using standard microsurgical techniques. Early in the series, dorsal myelotomy

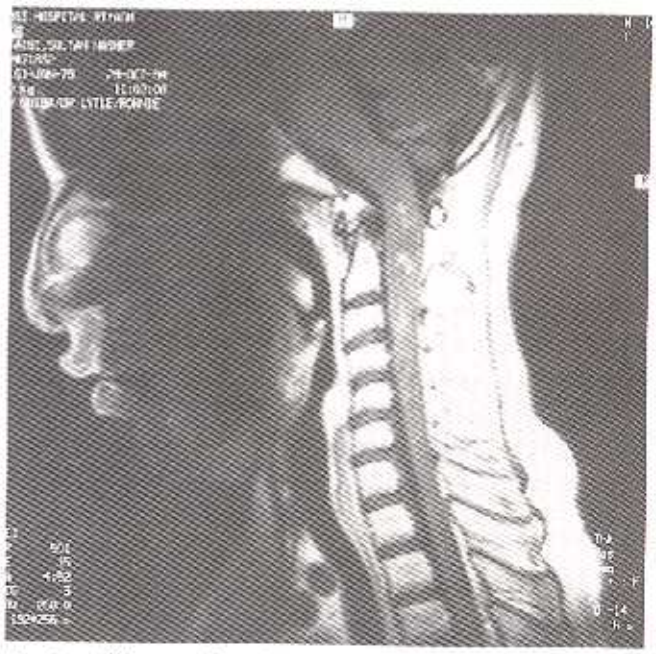

Fig. 1 MRI cervical cord showing an intramedullary malignant astrocytoma extending from C5 into medulla (Case 8)

was performed using the bipolar diathermy. In recent years, we have used the CO_2 laser mounted on the microscope to do the myelotomy in some cases. Tumour debulking is usually carried out using the CUSA. Total

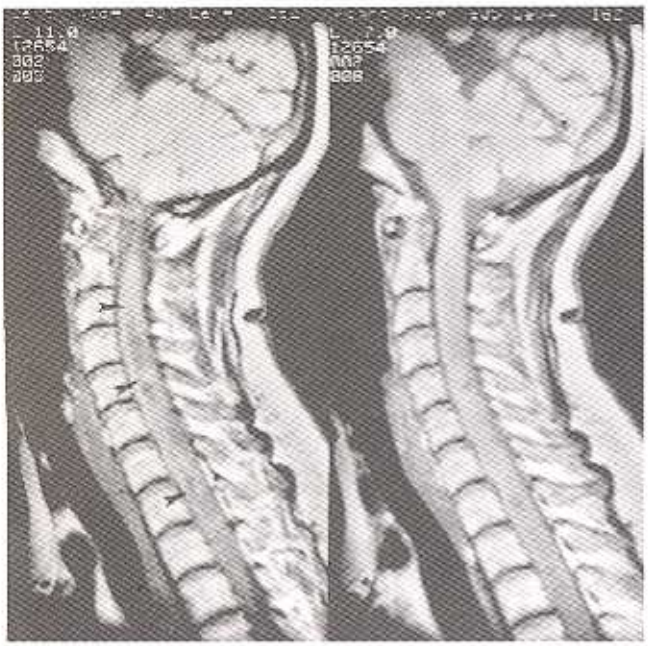

Fig. 2(a)

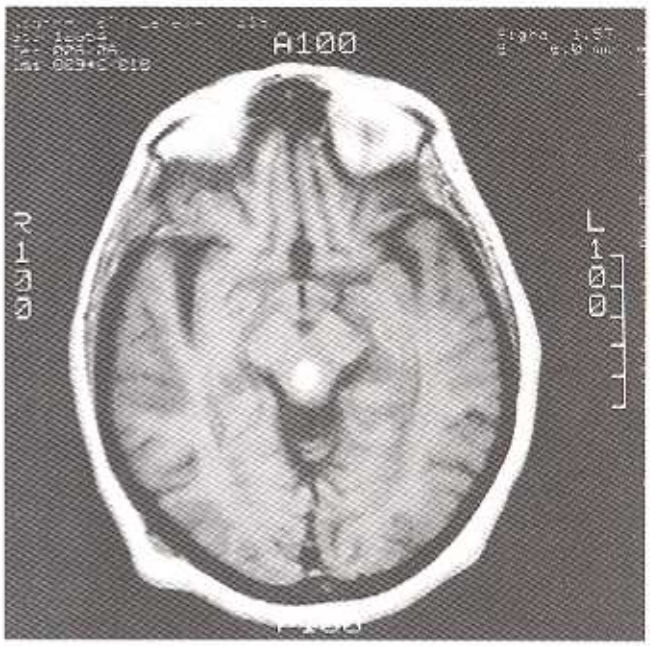

Fig. 2(b)

Fig. 2a,b: MRI showing a cervical intramedullary malignant astrocytoma (arrowheads 2a) with intracranial metastasis obstructing the aqueduct (2b) (case 9).

excision was achieved in 6 patients by dissecting the tumour along its cleavage plane with the spinal cord or conus. A more limited excision was performed because of obvious infiltration of the cord by tumour in another 6 patients and because the lesion (cavernoma) was heavily calcified in one case. A biopsy only was done for the patients with intramedullary tuberculoma and very vascular haemangioblastoma. These patients were treated with anti-tuberculous medication and embolisation respectively.

None of our cases deteriorated following surgery. At a follow-up ranging from 2-60 (median 12) months, the outcome was considered "favourable" in 6 patients.

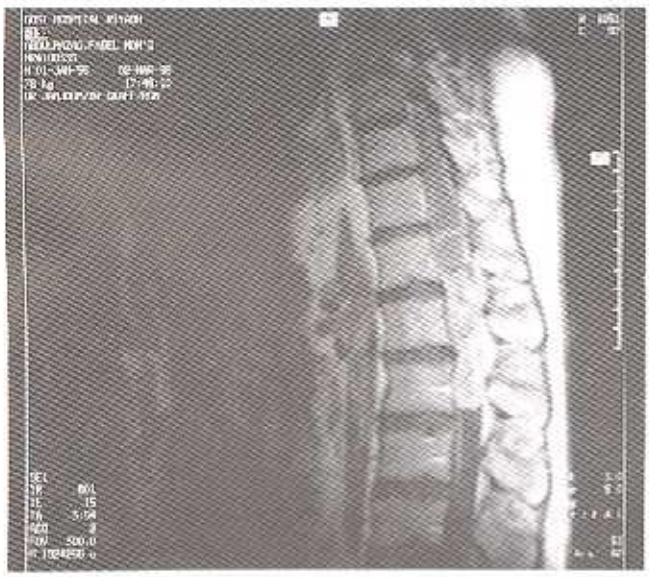

Fig. 3(a)

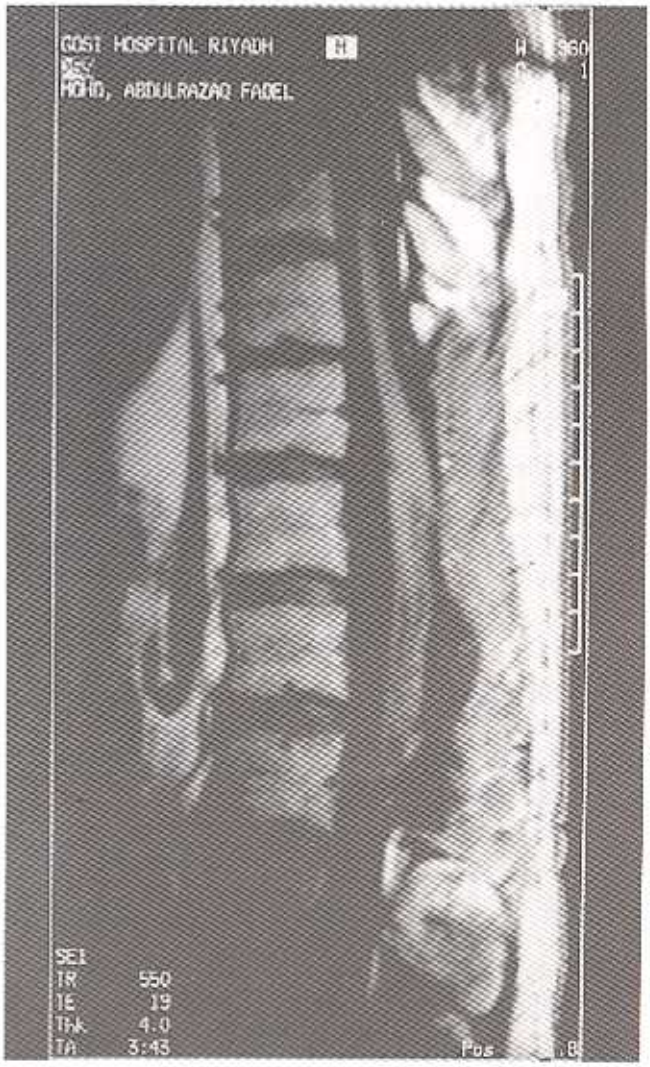

Fig. 3(b)

Fig. 3a,b: A preoperative MRI showing an ependymoma of the conus with syringomyelia proximal to the lesion (3a) and a postoperative MRI showing no residual tumour and a collapsed syrinx (3b).

Only 4 of our recent patients had a follow-up MRI to check the extent of the tumour excision *(Fig. 3a,b)*.

Discussion

Intramedullary tumours account for 17% of spinal tumours in our hospital[1]. The duration of symptoms in patients was relatively short (median 1.2 years vs 4.5 years[2,7]), nevertheless, 94% of our patients were unable to walk independently at presentation (McCormick *et al*[7] grades III and IV) compared to only 9-48% of patients in similar grades in other reports[2,4,7,9,11]. This may reflect delay in presentation of our patients who may have ignored the early mild symptoms, or delay in diagnosis.

There is no doubt that MRI is the method of choice for visualizing tumours within the spinal cord. However, the MRI diagnosis is correct in only 70% of cases[5], and allowing for the varied nature of tumours that can occur within the spinal cord, a histological diagnosis is mandatory. MRI is also sensitive in showing the associated syringomyelia which was evident in 5 of our 10 cases that were investigated by MRI. Samii and Klekamp[12] reported that a syrinx was present in 45% of their intramedullary tumours and that ependymomas and haemangioblastomas were the most common tumours to be associated with syringes.

On comparing the various tumour histology and location in our patients to what is reported in the literature[1-5], it appears that we have a higher incidence of malignant astrocytoma (25% vs 0-20%) and a lower incidence of low grade astrocytoma (13% vs 20-36%). In addition, the occurrence of tumour in the cervicomedullary region (19%) and the thoracic cord (31%) appears to be more frequent in our patients.

Spinal cord astrocytomas are rare lesions, usually low grade with a long natural history and a 5 year survival rate of 68%[10]. The extent of the safe surgical excision of low grade astrocytoma remains a controversial issue. Radical or quasiradical excision of low grade astrocytoma, which was performed on one of our two patients, was done in 38-67% of cases in different series[2-4]. Our policy is to explore the lesion. In the presence of a discrete cleavage plane and possibly an adjacent cystic component a complete excision is attempted. It is generally accepted that malignant astrocytoma is an incurable lesion and the tumour recurs within 1 year and the patient is dead within 2 years, irrespective of the amount of tumour excision[2,4,11]. It is therefore, agreed that a less radical intervention with minimal morbidity is usually adequate for this tumour.

The association of hydrocephalus with malignant spinal cord lesions has long been established and it is believed to be related to malignant seeding or due to obstruction of outlets of the fourth ventricle as a result of thickening of the leptomeninges overlying the cervicomedullary junction or due to excess protein production by an ependymoma[1,2]. Our case 9 is unusual in that the intracranial metastasis occurred prior to the tumour surgery and produced symptoms (hydrocephalus) before the primary cervical lesion became symptomatic. The other malignant astrocytoma cases in this series were also unusual in showing evidence of intracranial extension[14] and multicentricity. The latter is a unique feature that occurs in only 2.4% of glioma patients[13].

Ependymomas tend to have a discrete cleavage plane and their complete excision is more straightforward. We achieved total excision in 4 of our 5 ependymoma patients without an increase in morbidity, which is comparable to other reports[4,7,9]. It is well recognised that the recurrence of an ependymoma is exceptional when the excision is total, whereas an incomplete excision is associated with a higher recurrence rate[2,4,7,9].

All neurosurgeons agree that radiotherapy has a palliative role in the treatment of intramedullary malignant astrocytoma. The use of radiotherapy in the treatment of other intramedullary tumours is however, controversial. Some authors, like us, favour its use for residual low grade astrocytoma and ependymoma[4], while others found no advantage in its use for these lesions[3].

The isolated lesions which were encountered in this series included a lipoma, cavernoma, haemangioblastoma, neurofibroma and tuberculoma. The lipoma was the subject of a previous publication[8]. Only 65 cases of intramedullary cavernoma have been reported in the literature up to 1991 of which only 24 lesions were excised completely. Haemangioblastoma accounted for 4-14% of cases in other series[2-4], while an intramedullary neurofibroma is rare[17] and was encountered in only 2% of the tumours reported by Cristante and Herrmann[2]. Intramedullary tuberculoma is a very unusual occurrence that was reported in one of the 18 cases of central nervous system tuberculomas reported by Kioumehr *et al*[18] and it can develop as a consequence of tuberculous meningitis[19] as in our case.

In conclusion, the characteristics of patients with intramedullary tumours in Saudi Arabia appear to differ slightly from what is reported elsewhere. Patients should be investigated by MRI early before they develop severe neurological deficits. All intramedullary tumours

should be treated surgically at an early stage. It is advisable that these patients should be treated only in specialised neurosurgical units in Saudi Arabia.

Acknowledgement

The authors are grateful to Ms Hilda A Sumandar for her excellent secretarial assistance.

References

1 Jamjoom AB, Jamali A, Jamjoom ZAB, *et al*: Spinal tumours: experience at King Khalid University Hospital. *Ann Saudi Med.* 1993;**13**:47-51.

2 Cristante L and Herrmann HD: Surgical management of intramedullary spinal cord tumours: functional outcome and sources of morbidity. *Neurosurgery*. 1994;**35**:69-76.

3 Brotchi J, Dewitte O, Levivier M, *et al*: A survey of 65 tumours within the spinal cord: surgical results and the importance of preoperative magnetic resonance imaging. *Neurosurgery*. 1991; **29**:651-657.

4 Cooper P R: Outcome after operative treatment of intramedullary spinal cord tumours in adults: Intermediate and long term results in 51 patients. *Neurosurgery*. 1989;**25**:855-859.

5 Lunardi P, Lucastro G, Missori P, *et al*: Management of intramedullary tumours in children. *Acta Neurochir (Wein)*. 1993;**120**: 59-65.

6 Herrmann HD, Neuss M and Winkler D: Intramedullary spinal cord tumours resected with CO_2 laser microsurgical technique: recent experience in fifteen patients. *Neurosurgery*. 1988;**22**:518-522.

7 McCormick P C, Torres R, Post K D, *et al*: Intramedullary ependymoma of the spinal cord. *J Neurosurg*. 1990;**72**:523-532.

8 Rahman N, Salih M, Jamjoom AB and Jamjoom ZAB: Congenital intramedullary lipoma of the dorsocervical cord with intracranial extension: Case report. *Neurosurgery*. 1994;**34**:1081-1084

9 Ferrante L, Mastronardi L, Celli P, *et al*: Intramedullary spinal cord ependymomas: a study of 45 cases with long term follow up. *Acta Neurochir (Wein)*. 1992;**119**:74-79.

10 Sandler HM, Papadopoulos SM, Thorton AF, *et al*: Spinal cord astrocytomas: results of therapy. *Neurosurgery*. 1992;**30**: 490-493.

11 Epstein F J, Farmer JP and Freed D: Adult intramedullary astrocytomas of the spinal cord. *J Neurosurg*. 1992;**77**:355-359.

12 Samii M and Klekamp J: Surgical results of 100 intramedullary tumours in relation to accompanying syringomyelia. . *Neurosurgery*. 1994;**35**:865-873.

13 Epstein F J and Farmer JP: Pediatric spinal cord tumour surgery. *Neurosurg Clin N Am*. 1990;**1**:569-590.

14 Inagawa T, Kamiya K and Nagasako R: A case of spinal low grade astrocytoma with exophytic and intracranial extension. *Surg Neurol*. 1995;**43**:261-264.

15 Mishra H D, Haran R P, Singh J P, *et al*: Multicentric gliomas: two case reports and a review of literature. *Br J Neurosurg*. 1990;**4**:535-539.

16 Canavero S, Pagni CA, Duca S, *et al*: Spinal intramedullary cavernous angioma: a literature metaanalysis. *Surg Neurol*. 1994; **41**:381-388.

17 Rout D, Pillai SM and Radhakrishnan W: Cervical intramedullary schwannoma. Case report. *J Neurosurgery*.1983;**58**:962-964

18 Kioumehr F, Dadsetan MR, Rooholamini SA, *et al*: Central nervous system tuberculosis: MRI. *Neuroradiology*. 1994;**36**: 93-96.

19 Lin SK, Wu T and Wai YY: Intramedullary spinal tuberculomas during treatment of tuberculous meningitis.*Clin Neurol Neurosurg*. 1994;**96**:71-78.

Correspondence to

ABDULHAKIM JAMJOOM, Associate Professor and Consultant, Division of Neurosurgery, Security Forces Hospital, P O Box 3643, Riyadh 11481, Saudi Arabia.