Case Report

Spinal Haemangioblastomas in Nigerians: A Report of Two Cases

O.A Badejo[1,2], T.A Oyemolade[2], T.A Malomo[2].

[1]Division of Neurological Surgery, Department of Surgery, College of Medicine, University of Ibadan.
[2]Department of Neurological Surgery, University College Hospital, Ibadan

ABSTRACT

Background: Spinal haemangioblastomas are rare benign but potentially devastating vascular tumours. They may occur sporadically or as a component of Von Hippel-Lindau (VHL) disease. There is a dearth of information on these tumours among Nigerians. This study aims to evaluate the clinical profile and outcome of surgery as well as the challenges of the management of these tumours in a resource-limited country.

Methods: We retrospectively analysed data from the records of patients who had surgery for spinal haemangioblastoma at our centre between January 2004 and December 2018.

Results: We operated 2 cases of spinal haemangioblastomas during the study period (1 male, 1 female). The patients were aged 25 and 50 years. While both patients had thoracic lesions and presented with sensorimotor deficits, only one had sphincteric dysfunction. The onset of symptoms was 2 weeks in one patient and 18 months in the other. The tumour was located in the thoracic region in the 2 patients. One patient had extramedullary tumour patient had an intramedullary tumour location. Both individuals were paraplegic at the time of surgery, which was 3 days in one patient and more than 4 months in the other. Whereas gross total tumour resection was achieved both instances, the patient with shorter duration of symptoms and extramedullary tumour made a rapid-post operative neurological improvement, while the one with the longer duration of symptoms and intramedullary tumour remained paraplegic until his death nine months after surgery.

Conclusion: Spinal haemangioblastomas are rare tumours. Long duration of symptoms and intramedullary tumour location may be predictors of unfavorable surgical outcome.

INTRODUCTION

Haemangioblastomas are benign vascular tumours of the central nervous system, CNS (1, 2, 3, 4). They may occur sporadically (75%) or as a component of Von Hippel-Lindau (VHL) disease (25%) (1). Common locations in the CNS include the cerebellum (44%–72%), spinal cord (13%–50%), brainstem (10%–20%), supratentorial region (<1%), and lumbosacral nerve roots (<1%) (5). Spinal cord haemangioblastomas are relatively rare, accounting for only 2% to 6% of all spinal cord tumors (1, 6). In spite of their benign nature, they can result in clinically devastating morbidities to the affected patients due to severe neurological deficits resulting from tumour mass, perilesional oedema, cyst or syrinx (1, 3, 4, 7). There is a paucity of information on spinal tumours, particularly spinal haemangioblastomas, in Sub-Saharan Africa. We present a report of two cases of sporadic spinal haemangioblastomas in Nigerian patients. To the best of our knowledge, this is the first report on these tumours from our country.

Corresponding author:
Dr. Oluwakemi. A. Badejo
Division of Neurological Surgery,
Department of Surgery,
College of Medicine, University of Ibadan.
Email: kemibadejo@yahoo.com
Phone: +2348097317732

Key words: *Haemangioblastoma, Spinal cord, Nigerian, Surgery, Outcome*

Case reports

Case 1:

25-year-old right-handed female who presented with right flank pain, numbness and paraesthesia of the lower extremities of two weeks' duration. She developed progressive paraparesis about ten days later. Her symptoms were not associated with sphincteric dysfunction. Examination revealed full power in the upper extremities. She however had a spastic paraparesis which was significantly worse on the right (grade 1-3), hyperreflexia (worse in the lower limbs) and sustained ankle clonus bilaterally. Her plantar response was extensor on the right and flexor on the left. She had a sensory level of T9. There were no features suggestive of VHL. Magnetic resonance imaging of the thoracic spine showed a T10 intradural-extramedullary lesion (Figure 1 a-c). She had further neurologic deterioration (from Frankel C to B) a week after presentation while sourcing for funds for her care. She subsequently had (partial) T9/T10 laminectomies and gross total tumour excision. Her post-surgery recovery was satisfactory with progressive improvement in motor functions of the lower limbs. She was discharged home twenty two days after surgical intervention with lower limb power of grade 4 to 4^{+}. On follow-up evaluation five weeks post-operatively, she had full muscle power in the lower limb muscle groups, except for the hip and knee extensors which were grade 4 to 4^{-} bilaterally. At six months follow-up, she had regained full power in all the muscle groups of the lower extremities. Histology was in keeping with a capillary haemangioblastoma.

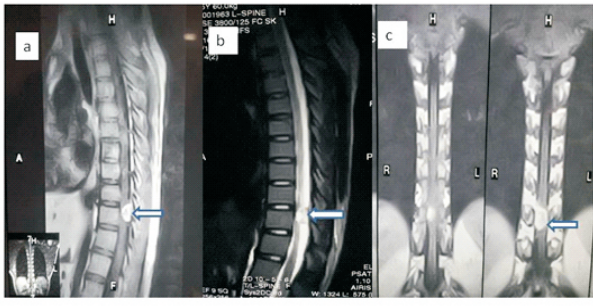

Fig. 1: Sagittal T1+C (a), T2+C (b) and coronal T1+C thoracic spine magnetic resonance imaging of patient 1 showing a contrast-enhancing T10 intradural-extramedullary tumour (arrows)

Case 2:

50-year-old right-handed man who presented with low back pain and progressive paraparesis of eighteen months and three months duration respectively. He had no co-morbidities. Examination revealed a middle-aged man with normal general examination findings. His long tracts were normal in the upper limbs. He had a spastic paraparesis with power being grade 4^{-} to 4^{+} in the right lower limb and 4 to 5 in the left lower limb. Sensory level was T7 bilaterally with residual to S4 and S5. Magnetic resonance imaging of the thoracic spine revealed a T9/T10 contrast-enhancing intramedullary tumour (Fig. 2).

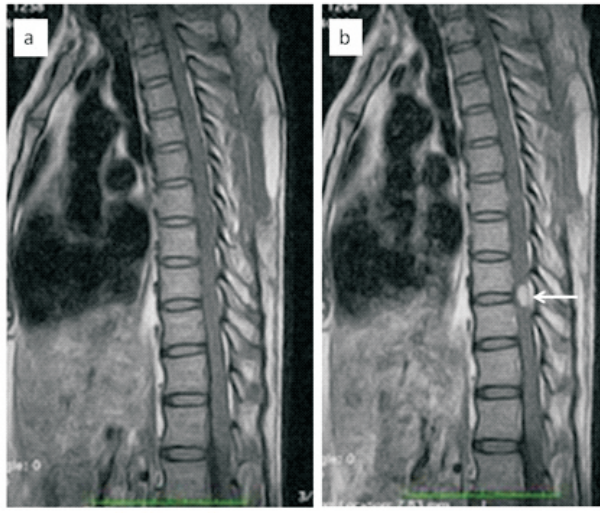

Fig. 2: Sagittal T1 (a) and T1+C (b) thoracic spine magnetic resonance imaging of patient 2 showing a T9/T10 contrast enhancing intramedullary tumour (arrow)

The patient initially declined surgical treatment and presented two months later with paraplegia, complete loss of sensation in the lower limbs, bisphincteric incontinence, infected grade IV mid-sacral decubitus ulcer and systemic evidence of sepsis. He had wound debridement and broad-spectrum intravenous antibiotic therapy. Following resolution of sepsis a month later, he had a T8-T10 laminectomies and gross total tumour excision (Fig. 3). Histology revealed a haemangioblastoma (Fig. 4). He made no neurologic gains post-operatively and was discharged home after five months of hospitalisation following healing of his pressure

sore. He was however readmitted three months later due to multiple bed sores (including a grade IV recurrent infected mid-sacral pressure ulcer) and septic shock. There was repetitive faecal soilage of the ischial component of the bedsores with waxing and waning systemic sepsis throughout his hospitalisation. The patient refused a colostomy. He developed multiple-organ dysfunction terminally and died nine months post-operatively.

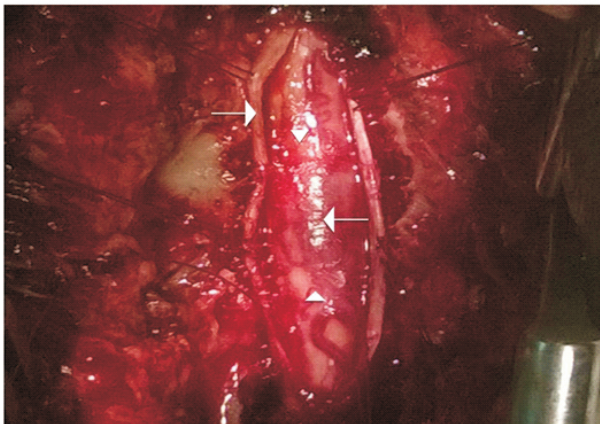

Fig. 3: Intraoperative photograph of the second patient after T8-T10 laminectomies and a durotomy. Note the purplish discoloration of the spinal cord at the site of the tumour (lower arrow). The upper arrow is on the dural edge and the arrow heads are on the cord proximal and distal to the lesion.

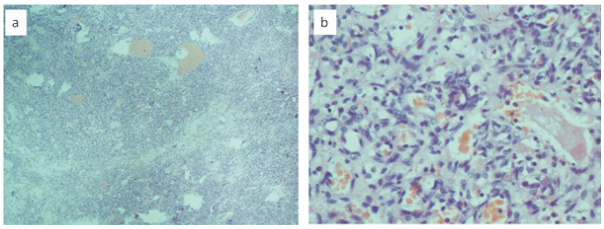

Fig. 4: Photomicrograph of the second patient showing proliferating stroma cells, tumour cells with oval to spindle shaped nuclei, occasional tumour giant cells and multiple thin-walled vascular channels (a-×25, b- ×40).

DISCUSSION

Spinal haemangioblastomas are relatively rare tumours accounting for 2-6% of spinal cord tumours (6, 8). They may occur sporadically or as part of VHL syndrome (1). Although histologically benign, these tumours can cause significant neurological deficits because of their location (1, 3, 4, 7). Between January 2004 and December 2018, we operated 2 patients with spinal haemangioblastoma, accounting for 2.1% of the operated cases of spinal tumours in our centre during the period. This is in keeping with the reported incidence of these tumours (6).

Male predominance of spinal haemangioblastomas has been documented, with a reported male to female ratio ranging between 1.6:1 to 5.5:1 (3, 8, 9, 10, 11, 12). The male gender accounted for 72.7% of the cases in the series by Yasuda et al (8) and 75% of the case in the series by Park et al (12). Conversely, there were more females in the study by Das et al with the male gender accounting for 42.9% of the patients' population (1). Our patients were equally distributed between the two genders. Spinal haemangioblastomas commonly occur in the third and fourth decades of life although occurrences in patients on either sides of this age range have been documented (1, 8, 12). The average age was 33.5 years in the series by Das et al (1) and 53.9 years in the series by Yasuda et al (8). The younger of our patients was in the third decade of life while the older one was in the sixth decade.

These tumours may present clinically with motor, sensory or autonomic deficits. In the series by Das et al (1) motor weakness was present in 86% of the patients at presentation, sensory deficits in 79% and bladder disturbance in 21%. Pain was the presenting symptom in 29% of their patients. Motor weakness and sensory deficits were the most common presenting symptoms in the study by Park et al (12) each occurring in 50% of the cases. Our patients had sensorimotor deficits at presentation but sphincteric dysfunction was present in only one of them.

Spinal haemangioblastomas are primarily intradural tumours and occur mostly in the cervical and thoracic regions (1, 3, 8, 13, 14, 15). In the series by Imagama et al (14) 84.6% of the tumours were intramedullary, 11.5% were intramedullary/extramedullary, and 3.8% were intradural-extramedullary. Deng et al (13) reported

41%, 37%, and 22%, respectively for the same groups of tumours respectively. In our patients the tumours were located in the thoracic region. The tumour was intramedullary in one patient and intradural-extramedullary in the other. Patients with intradural-extramedullary haemangioblastomas tend to be older than the patients with intramedullary tumours (8, 11, 16, 17, 18, 19, 20). Contrariwise, the younger of our patients had an intradural-extramedullary tumour location, whereas in the older one, the tumour location was intramedullary.

Microsurgical resection remains the treatment of choice for spinal haemangioblastomas but radiosurgery has been used in some patients (15, 21, 22, 23, 24, 25). The outcome of surgical management of these tumours depends on the pre-operative neurological status of the patients, location / volume of the tumour, duration of symptoms and the extent of surgical resection (1, 2, 4, 12, 26, 27, 28, 29). Good pre-operative functional status, complete tumour excision, small size and dorsal tumour location are predictors of good post-operative outcome. Our 2 patients were paraplegic at the time of surgery, the first for about 3 days and the second for more than 4 months. They both had gross total resection of their tumours. The duration of symptoms was significantly shorter in the patient with post-operative neurological improvement who also happened to be the one with an intradural-extramedullary tumour.

Our cases also highlighted the role of poverty, ignorance, and lack of social support in the outcome of neurological diseases in resource-poor countries. The first of our patients became paraplegic while sourcing for funds for surgery while the second patient (although better educated than the first patient) initially declined surgery. He returned for further evaluation after the onset of paraplegia and sepsis from a mid-sacral decubitus ulcer which further delayed his surgery. Following discharge from the hospital, his lack of social support contributed to his development of multiple decubitus ulcers and sepsis which eventually led to his death. Of note is the intramedullary location of his tumour, a poor prognostic indicator.

CONCLUSION

Spinal haemangioblastomas are relatively rare tumours. They are mostly located in the cervical and thoracic regions. Although benign, they can cause significant morbidity and even mortality. Poverty, ignorance and lack of social support continue to pose a serious challenge to the management of these tumours and other neurological diseases in the developing countries.

Acknowledgements

Not applicable

Conflict of interest

The *authors report no conflicts of interest*

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1. Das JM, Kesavapisharady K, Sadasivam S, Nair SN. Microsurgical treatment of sporadic and von Hippel-Lindau disease associated spinal Hemangioblastomas: a single-institution experience. Asian Spine J. 2017;11(4):548–55. https://doi.org/10.4184/asj.2017.11.4.548
2. Lee DK, Choe WJ, Chung CK, Kim HJ. Spinal cord hemangioblastoma: surgical strategy and clinical outcome. J Neurooncol. 2003;61:27–34.
3. Mandigo CE, Ogden AT, Angevine PD, McCormick PC. Operative management of spinal hemangioblastoma. Neurosurgery. 2009;65:1166–1177.
4. Na JH, Kim HS, Eoh W, Kim JH, Kim JS, Kim ES. Spinal cord hemangioblastoma: diagnosis and clinical outcome after surgical treatment. J Korean Neurosurg Soc. 2007;42:436–440.
5. Pluta RM, Wait SD, Butman JA, et al. Sacral hemangioblastoma in a patient with von Hippel-

Lindau disease: case report and review of the literature. Neurosurg Focus. 2003;15:E11.

6. Mehta GU, Asthagiri AR, Bakhtian KD, Auh S, Oldfield EH, Lonser RR. Functional outcome after resection of spinal cord hemangioblastomas associated with von Hippel-Lindau disease. J Neurosurg Spine. 2010;12:233–242.
7. Richard S, Campello C, Taillandier L, Parker F, Resche F. Haemangioblastoma of the central nervous system in von Hippel-Lindau disease. J Intern Med. 1998;243:547–553
8. Yasuda T, Hasegawa T, Yamato Y, et al. Relationship between spinal hemangioblastoma location and age. Asian Spine J 2016;10:309–313. doi: 10.4184/asj.2016.10.2.309
9. Chu BC, Terae S, Hida K, Furukawa M, Abe S, Miyasaka K. MR findings in spinal hemangioblastoma: correlation with symptoms and with angiographic and surgical findings. AJNR Am J Neuroradiol. 2001;22:206–217.
10. Conway JE, Chou D, Clatterbuck RE, Brem H, Long DM, Rigamonti D. Hemangioblastomas of the central nervous system in von Hippel-Lindau syndrome and sporadic disease. Neurosurgery. 2001;48:55–62.
11. Barbosa-Silva E, Carvalho GT, Frota Mde O, Sousa AA, Souza CB. Intradural extramedullary hemangioblastoma. Arq Neuropsiquiatr. 2009;67:530–533.
12. Park CH, Lee CH, Hyun SJ, Jahng TA, Kim HJ, Kim KJ. Surgical outcome of spinal cord hemangioblastomas. J Korean Neurosurg Soc. 2012;52(3):221–227. https://doi.org/10.3340/jkns.2012.52.3.221.
13. Deng X, Wang K, Wu L, et al. Intraspinal hemangioblastomas: analysis of 92 cases in a single institution: clinical articles. J Neurosurg Spine. 2014; 21:260-269.
14. Imagama S, Ito Z, Wakao N, et al. Differentiation of localization of spinal hemangioblastomas based on imaging and pathological findings. Eur Spine J. 2011;20:1377–1384
15. Lonser RR, Oldfield EH. Spinal cord hemangioblastomas. Neurosurg Clin N Am. 2006;17:37–44.
16. Wisoff HS, Suzuki Y, Llena JF, Fine DI. Extramedullary hemangioblastoma of the spinal cord: case report. J Neurosurg. 1978;48:461–464.
17. Brisman JL, Borges LF, Ogilvy CS. Extramedullary hemangioblastoma of the conus medullaris. Acta Neurochir (Wien) 2000;142:1059–1062
18. Taniguchi S, Ogikubo O, Nakamura T, et al. A rare case of extramedullary-intradural hemangioblastoma in the thoracic spine. Spine (Phila Pa 1976) 2009;34:E969–E972.
19. Kitanaka C, Kuwahara M, Teraoka A. Intradural, purely extramedullary hemangioblastoma of the spinal cord: case report. Neurol Med Chir (Tokyo) 1993;33:377–380.
20. Toyoda H, Seki M, Nakamura H, Inoue Y, Yamano Y, Takaoka K. Intradural extramedullary hemangioblastoma differentiated by MR images in the cervical spine: a case report and review of the literature. J Spinal Disord Tech. 2004;17:343–347
21. Roonprapunt C, Silvera VM, Setton A, Freed D, Epstein FJ, Jallo GI. Surgical management of isolated hemangioblastomas of the spinal cord. Neurosurgery 2001;49(2):321–327; discussion 327–328.
22. Ammerman JM, Lonser RR, Dambrosia J, Butman JA, Oldfield EH. Long-term natural history of hemangioblastomas in patients with von Hippel-Lindau disease: implications for treatment. J Neurosurg 2006;105(2):248–55.
23. Cohen-Gadol AA, Spencer DD, Krauss WE. The development of techniques for resection of spinal cord tumors by Harvey W. Cushing. J Neurosurg Spine 2005;2(1):92–97.
24. Parsa AT, Chi JH, Acosta FL, Ames CP, McCormick PC. Intramedullary spinal cord tumors: molecular insights and surgical innovation. Clin Neurosurg 2005;52:76–84.

25. Moss JM, Choi CY, Adler JR, Soltys SG, Gibbs IC, Chang SD. Stereotactic radiosurgical treatment of cranial and spinal hemangioblastomas. Neurosurgery. 2009. 65: 79-85
26. Shin DA, Kim SH, Kim KN, Shin HC, Yoon DH. Surgical management of spinal cord haemangioblastoma. Acta Neurochir (Wien) 2008;150:215–220. discussion 220.
27. Takai K, Taniguchi M, Takahashi H, Usui M, Saito N. Comparative analysis of spinal hemangioblastomas in sporadic disease and Von Hippel-Lindau syndrome. Neurol Med Chir (Tokyo) 2010;50:560–567.
28. Boström A, Hans FJ, Reinacher PC, et al. Intramedullary hemangioblastomas : timing of surgery, microsurgical technique and follow-up in 23 patients. Eur Spine J. 2008;17:882–886
29. Lonser RR, Weil RJ, Wanebo JE, DeVroom HL, Oldfield EH. Surgical management of spinal cord hemangioblastomas in patients with von Hippel-Lindau disease. J Neurosurg. 2003;98:106–116.