Sporadic spinal hemangioblastomas can be effectively treated by microsurgery alone.

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Introduction

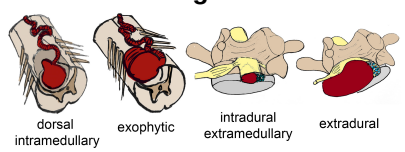
Hemangioblastomas are benign, slow-growing vascular tumors that can occur throughout the neuraxis. They can occur sporadically or as a manifestation of the autosomal dominantly inherited von Hippel-Lindau (vHL) syndrome, in which case the lesions tend to be multiple and widely distributed throughout the central nervous system. Sporadic cases commonly present as solitary lesions that are potentially curable by complete surgical excision, and due to the absence of associated systemic disease, a long and disease-free survival can be expected.

Herein, we present our institutional experience in the treatment of sporadic spinal hemangioblastomas (SSHB). We also performed a meta-analysis of all sporadic SSHB cases reported in the literature(1).

Methods

This is a retrospective analysis of 14 SSHB patients (8 males and 6 females) operated on over 23 years. The median age was 41.5 years (24–70). vHL was excluded by imaging in all. The median follow-up was 4 years (1–23). We also conducted a meta-analysis of all 271 SSHB cases reported in the English-language literature from 1967 to 2011. All patients underwent preoperative and early postoperative contrast-enhanced MRI examinations. The diagnosis of hemangioblastoma was confirmed by histopathology in all cases.

Fig-1

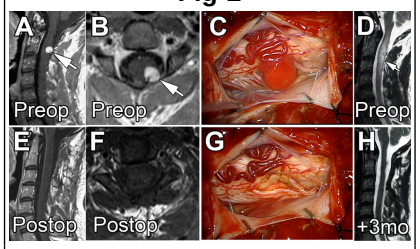


Common types of sporadic spinal hemangioblastomas

Review of Literature

In our literature search, we came across 47 studies between the years 1967 and 2011. Including our cohort, a total of 271 SSHB cases have been reported. The mean age was 40.6 years and 130 (62%) were male, and 79 (38%) were female. Information on localization along the spinal axis was present for 239 cases: 50.4% were cervical, 36.4% were thoracic and 13.4% were lumbosacral.

Fig-2



SSHB may present with marked “diffuse segmental spinal cord enlargement” and this resolves after complete resection.

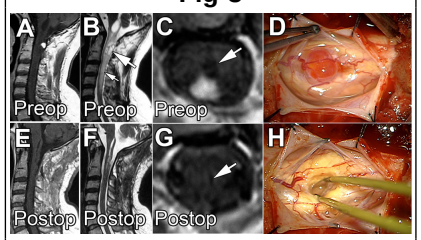
Learning Objectives

Preoperative embolisation for the treatment of spinal hemangioblastoma can be used in the clinics. We aimed to show our cases to do this surgery without embolisation.

Results

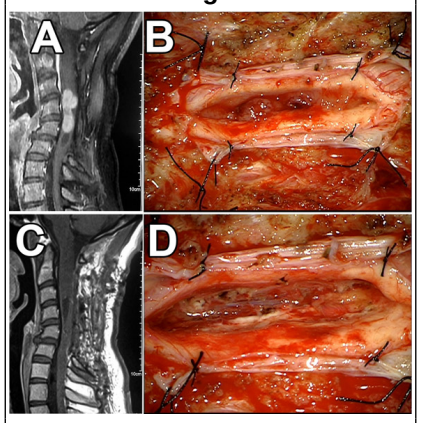
All lesions were solitary. Nine (64.3%) lesions were cervical, 3 (28.5%) were thoracic, and 1 (7.1%) was lumbar. Eight (57.1%) tumors were dorsal intramedullary, 4 (28.6%) were exophytic, 1 (7.1%) was intradural extramedullary and 1 (7.1%) was completely extradural. Diffuse segmental cord enlargement was present in 7 (50%) and a cyst/syrinx was present in 7 (50%) cases. The 14 patients underwent 15 operations, and gross-total resection was achieved in all operations. There was no mortality. Symptoms improved after 8 (53.3%) of 15 operations, remained the same after 5 (33.3%) and worsened after 2 (13.3%). There was one recurrence 15 years after MRI had confirmed total resection, and this patient underwent a repeat total resection with improvement in neurological status. The recurrence rate in other series ranged from 6.25 to 7.7%.

Fig-3



Syrinx and “diffuse segmental spinal cord enlargement” can be observed simultaneously.

Fig-4

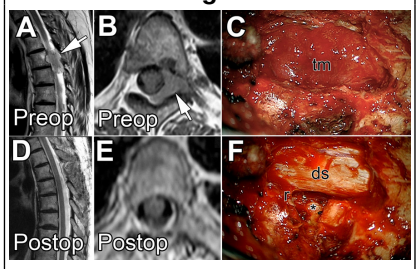


Upon recurrence SSHB may be lobulated rather than globular.

Discussion

Hemangioblastomas are rare, benign tumors of the neuraxis. In surgical case series, spinal hemangioblastomas make up 2–15% of all spinal intramedullary tumors(3). Our analysis revealed that sporadic tumors make up slightly more than half of the cases that are affected in the spinal cord.There are differences in epidemiological and clinical variables and outcomes between sporadic and vHL-associated hemangioblastomas.Recurrence in completely resected SHB cases is rare. However, we observed that the recurrent tumor was multilobular, which made the resection more complex. Common types of SHB are shown in figures.

Fig-5



Even a large syrinx resolves without specific treatment after tumor resection.

Conclusions

SHB occur slightly more common than those associated with von Hipppel Lindau disease. SHB cases are most commonly encountered as solitary lesions and are most frequently located in the upper spinal cord. Excellent surgical results can be achieved with microsurgery without the use of preoperative or postoperative adjuvant therapies. The long-term outcome is good, with only rare recurrences.

References

1-) . 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 42(6):436-440, 2007

2-) . Mandigo CE, Ogden AT, Angevine PD, McCormick PC: Operative management of spinal hemangioblastoma. Neurosurgery 65(6):1166-1177, 2009