Surgery of a Solid Hemangioblastoma at the Cervicomedullary Junction

Tae-Won Kim, M.D., Shin Jung, M.D., Tae-Young Jung, M.D., Sam-Suk Kang, M.D.
Department of Neurosurgery, Chonnam National University Hwasun Hospital, Medical School, Hwasan, Korea

The surgical removal of solid deep-seated hemangioblastomas remains challenging, because treatment of these lesions is often complicated by severe bleeding associated with the rich vascularity of this tumor, and by severe neural tissue injury associated with the difficulty of en bloc resection, especially when the tumor is located at the cervicomedullary junction. Therefore, preoperative embolization of deep-seated solid hemangioblastomas may play an important role in successful surgical removal by reducing major bleeding and neural tissue damage. A 24-year-old woman, 28-weeks pregnant, was admitted to our hospital for the evaluation of quadripareisis, and brain magnetic resonance imaging(MRI) revealed intra-axial mass lesion in the cervicomedullary junction. After delivery, her neurologic symptoms became aggravated, and we decided to operate. Preoperative angiography revealed a hypervascular tumor in the posterior fossa, and embolization of the main feeding artery using gelfoam and microcoils, resulted in marked reduction of tumor vascularity. She underwent a midline suboccipital craniotomy involving the removal of the arch of C-1. The tumor was totally removed through a midline myelotomy, and at her 6-month follow-up she walked independently. We report on the combined use of the preoperative embolization of feeding vessels and subsequent operative resection in a patient with a solid hemangioblastoma at the cervicomedullary junction immediately after delivery.

KEY WORDS: Preoperative embolization · Solid hemangioblastoma · Cervicomedullary junction · Midline myelotomy · Pregnancy.

Introduction

Hemangioblastomas are solid or cystic benign vascular tumors that may arise anywhere in the body. The incidence of these tumors in the CNS is relatively low, and they account for only 1–2.5% of all intracranial neoplasms. Surgical removal of brain stem and cervicomedullary junction hemangioblastomas is often complicated and difficult, because of tumor hypervascularity and location. In addition, though surgical mortality rates have been reduced from 40% to 50% before the 1970s to 24%, they remain unacceptable even given the advantages of modern microsurgical techniques. Moreover, the presurgical embolization of solid hemangioblastomas has been known to be effective. Pregnancy appears to hasten the symptomatic presentation of certain intracranial neoplasms, including hemangioblastomas, possibly due to endocrinological changes and increases in maternal blood volume. Here, we report on a pregnant patient with a solid hemangioblastoma in the cervicomedullary junction whose symptoms became aggravated after Cesarean-section delivery. The patient was successfully treated by the preoperative embolization of feeding vessels and subsequent en bloc resection.

Case Report

A 24-year-old woman who was 28-weeks pregnant was admitted to our hospital for the evaluation of a one year history of headache, vomiting, and progressive aggravation of quadripareisis. Brain magnetic resonance imaging(MRI) revealed intra-axial mass lesions in the cervicomedullary junction (Fig. 1). The predominantly solid lesion contained a cystic component. On neurological examination she was oriented, and here quadripareisis was of Grade IV. Because she was 28 weeks...
vascularity (Fig. 2B). She underwent a midline suboccipital craniotomy and removal of the posterior arch on C1. We found both cervical rostral entry zones, and determined that medulla oblongata and cervical cord midlines turned to the right (Fig. 3A). A midline medullotomy was performed under an operating microscope (Fig. 3B), and the hemangioblastoma was dissected from the brain stem. The tumor was gently retracted laterally, and its small feeding vessels were identified, coagulated with a bipolar forceps, and dissected sharply. The tumor was then gently rolled back, and its blood supply was progressively interrupted. Finally, we were able to excise the mass lesion totally (Fig. 3C). Histology confirmed the lesion to be a hemangioblastoma. The tumor was composed of many capillaries with intermingled stromal cells, and tumor cells immunostained positively for vimentin and negatively for CD34 (Fig. 4). Postoperative computed tomography (CT) confirmed that tumor was totally resected. Postoperatively, quadriplegia improved to Grade II, transient respiratory dysfunction developed and intubation and assisted ventilation were required for 5 days. However, she gradually recovered, and was able to walk using a walker 4 weeks postoperatively. At the 6-month follow-up she walked independently.

Discussion

In 1926, Lindau described a cystic tumor of the cerebellum associated with retinal angiomaticus and tumors of the kidney. Cushing and Bailey named this CNS tumor "hemangioblastoma," thus emphasizing its neoplastic nature. The tumor has a benign natural history with a low mitotic rate and little cellular pleomorphism, and microscopically, the mural nodules and solid tumor are composed of thin-walled vessels and capillaries. Hemangioblastomas of the CNS usually occur in the posterior fossa, including the cerebellum, brain stem and spinal
certain intracranial tumors, including hemangioblastomas. Possible reasons for this include endocrinological changes during pregnancy and an increase in maternal blood volume. In our case, the patient’s neurological symptoms were aggravated after C-section delivery. Perhaps, blood and fluid volumes in our patient rapidly changed during the abdominal surgery. Blood and fluid volume may be changed and this edematous and ischemic situation may have damaged neuronal cells of the peritumoral lesion and aggravated her neurological symptoms. Thus, we administrated steroid and her neurological symptoms gradually recovered. Hemangioblastoma in this patient was located at the cervicomedullary junction, and it was 3 cm in size. We decided to operate but were concerned with its size and location, and thus, we opted for preoperative embolization, and followed this within 48-hours with resection. We found

Pregnancy appears to hasten the symptomatic presentation of
cord, but also may occur in supratentorial structures. Approximately 25% of hemangioblastomas are associated with von-Hippel-Lindau (VHL) disease.

In general, cerebellar and supratentorial hemangioblastomas are composed of cyst and intramural nodule. The surgical outcome of most cystic hemangioblastomas is favorable after total removal of the mural nodule. However, brain stem, cervicomedullary junction, and spinal cord hemangioblastomas are almost always solid and tend to easily lead to massive bleeding during operation. This is the first reason for the postoperatively high morbidity and mortality of these locations, given their eloquent locations. Although surgical intervention of brain stem and cervicomedullary junction hemangioblastomas is associated with control rates ranging from 75-90%, intervention has historically been the treatment of choice for solitary hemangioblastomas. Radiation therapy and stereotactic radiosurgery have also been utilized in cases of recurved and multiple lesions, but most neurosurgeons advocate surgical removal as a treatment of first choice.

The safe excision of solid hemangioblastomas, with minimal blood loss, requires surgical techniques similar to those used to treat arteriovenous malformations. In particular, piecemeal tumor resection may result in marked surgical difficulties due to intraoperative hemorrhage. To solve these intraoperative hemorrhages, neurointerventional techniques are used, which enable to facilitate the devascularization of hemangioblastomas, thus simplifying surgical removal. Reports of successful presurgical embolization and surgical resection of hemangioblastomas emphasize the efficacy of the technique. Because brain stem and cervicomedullary junction hemangioblastomas are predominantly solid tumors, the preoperative embolization of feeding vessels is being important factor of successful tumor removal. In cases of cervicomedullary junction hemangioblastoma, a midline incision on the intercolumnar space of the medulla oblongata is too important. In many cases, the midline deviates to right or left, surgeons should find both cervical rootlet entry zones and decide on the myelotomy area (Fig. 5).

Fig. 5. This figure shows that the midline in the cervicomedullary junction is displaced laterally. Surgeons should find both cervical rootlet entry zones and decide on the myelotomy area.
both cervical rootlet entry zones and performed a myelotomy, which resulted in en-block tumor removal. After 6-months follow-up she walked independently.

Conclusion

Hemangioblastomas at the cervico-medullary junction are rare lesions, and most of them are solid. Moreover, because of its eloquent location, neurological symptoms may show rapid aggravation during pregnancy or major operations due to the changes in blood and plasma volume. Therefore, body fluid balance should be kept in an euolemic state as possible. We believe that total surgical resection is the treatment of choice for solitary cervico-medullary hemangioblastomas and recommend preoperative embolization of the feeders for the ease of tumor resection. Surgeons should find both cervical rootlet entry zones and decide on the myelotomy area. It is our opinion that exact midline incision will decrease surgical morbidity.

References


Commentary

The authors presented a case in which surgical removal of hemangioblastoma in the cervicomedullary junction had been achieved successfully with the aid of preoperative embolization of the tumor feeders. The surgery of the lesion in the brain stem and high cervical area has been among the most challenging neurosurgical procedures, due to the proximity to the vital neural components and complex microsurgical characteristics, although modern neurosurgical techniques, including employment of the surgical microscope and microsurgical instruments, has contributed to improved surgical outcomes during the last decades. The authors were able to accomplish a complete resection of the hypervascular lesion in the cervicomedullary area without grossly damaging surrounding structures, by approaching...
via midline medullotomy, the least invasive incision, with meticulous microsurgical dissection. Also, effective preoperative embolization was assumed to be one of the pivotal elements for this successful resection. The authors described peripar-tum neurological changes of the patient, suggesting the plausible mechanism, and timely surgical treatment that they had provided.

In the present case, the authors had attained favorable functional outcome by preserving the anatomical integrity of surrounding normal neural structures. Recent technical advances enable practical use of real-time neurophysiological monitoring during surgery, which are currently available at most of major health institutes in Korea. It has been strongly recommended that a neurosurgeon should employ those monitoring feedback systems available in his or her clinical environment, especially when the surgical target involves the eloquent area. Anatomical maintenance of the vital neural structures alone would not always guarantee satisfactory functional preservation after surgery in those eloquent areas, because a minute injury without definite anatomical alteration can result in catastrophically disabled neurological deficit. One cannot deny that the intraoperative neurophysiological monitoring of neural integrity, such as continuous monitoring of sensory or motor evoked potential recordings, has become the essential part of the modern neurosurgical practice to avoid unwanted neurological deterioration from the surgical procedures, which is truly so when the surgery for the lesion located in the brain stem or cervicomedullary junction is planned.

The authors presented a valuable case in which a complete resection of the cervicomedullary hemangioblastoma was performed successfully along with microsurgical preservation of the surrounding neural structures. I would stress, however, that careful microsurgical dissection could not dispense with reasonable intraoperative neurophysiological monitoring in the cases like the present one.

Jeong Hoon Kim, M.D.
Department of Neurosurgery,
Ulsan University, Asan Medical Center