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CASE REPORT

Extra- and intracranial dumbbell-shaped hemangiopericytoma

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Abstract Hemangiopericytomas are malignant tumors arising from pericytic cells and account for less than 1% of all vascular neoplasms. We report a rare case of an extra- and intracranial dumbbell-shaped hemangiopericytoma originating from the soft tissue of the neck and penetrating the skull base with invasion into the posterior cranial fossa. The 59-year-old female patient presented with a large pulsating neck mass and reported weakness, abnormal fatigue and headache. MRI revealed an inhomogeneously enhancing tumor and cerebral angiography showed intensive vascularization. Preoperative embolization was performed in order to decrease the operative blood loss. The tumor was operated via a far lateral approach through an osteoclastic suboccipital craniotomy. Total resection of both the intra- and extracranial part of the neoplasm (grade I by Simpson) could be achieved. The histopathological analysis revealed a mesenchymal, hypervascular tumor with the classic staghorn vascular pattern. In this article, we discuss the clinical presentation and multidisciplinary management of hemangiopericytoma and describe the radiological and pathological features of this tumor entity.

Keywords Hemangiopericytoma · Skull base · Vascular neoplasms

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Case report

A 59-year-old female patient was referred for evaluation of a left suboccipital mass. This slow-growing tumor had been recognized for the first time 5 years before by the patient and was under the control of her general practitioner who regarded this lesion as a lipoma. The patient reported weakness and abnormal fatigue for about one year and increasing headache for several months.

On physical examination a large pulsating tumor could be easily palpated at the left suboccipital area under the skin. Auscultation of the tumor revealed a systolic thrill. There were neither cranial nerve deficits nor other neurological symptoms and the neck was free of lymphadenopathy.

Magnetic resonance imaging (MRI) demonstrated a lobulated and irregular tumor with its main part in the soft tissue of the neck. It penetrated through the occipital bone and dura mater (CT not shown) and extended into the posterior cranial fossa. No dural attachment was observed (Fig. 1). The intracranial part of the tumor spread to the temporal bone, the jugular foramen and the carotid canal. Both the intra- and the extracranial part of the tumor showed a larger maximum diameter (4 and 7 cm, respectively) than the osseus defect of the skull base (3 cm). Thus, the tumor represented the remarkable shape of a dumbbell (Fig. 2a, b).

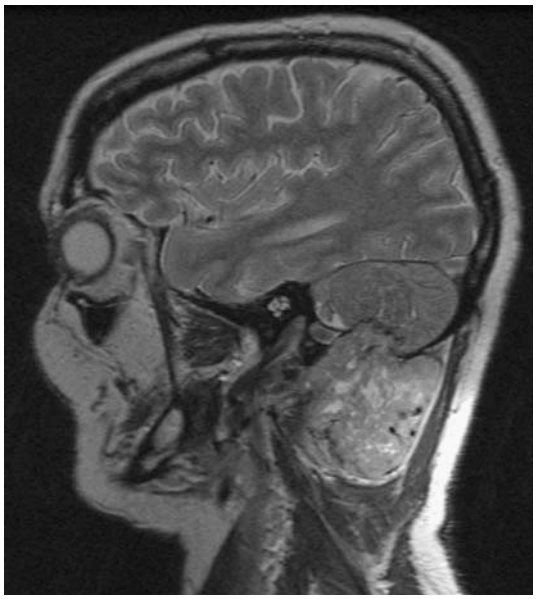

Fig. 1 T2-weighted sagittal MR depicts an inhomogeneous 4 × 7 cm neck-mass with prominent internal signal voids. The mass has eroded the skull and invades the posterior cranial fossa

Cerebral angiography revealed intensive vascularization of the tumor by the left occipital artery and muscular branches of the left vertebral artery. Additional feeders from the ascending pharyngeal and middle meningeal arteries were detected. A dense but heterogeneous tumor stain with a neovascular blush and early venous drainage was observed (Fig. 3a).

Test occlusion of the left vertebral artery was tolerated. Preoperative endovascular embolization of the tumor with particles (300–500 μm) and vortex coils was carried out via the occipital artery (Fig. 3b).

The patient was operated via a far lateral approach. Following the dissection of the extracranial part of the tumor an osteoclastic suboccipital craniotomy was performed to reach the intracranial part of the lesion. The intradural tumor mass was dissected carefully along its capsule from the arachnoidea. No infiltration of the caudal cranial nerves was observed and those parts of the tumor, which were near the jugular foramen could be removed without endangering the neighboring nerves. After total resection of both the intra- and extracranial part of the neoplasm (grade I according to the scale proposed by Simpson) plastic closure of the dura defect was performed with a fascia lata transplant.

The histopathological examination of the surgical specimen revealed a mesenchymal tumor demonstrating robust coexpression of CD 34 and bcl-2 and a proliferative activity of less than 5% (Ki-67-index). The tumor showed a rich vascular pattern, constituted by vessels lined with flat endothelium surrounded by abundant oval to spindle-shaped pericytic cells of perithelial appearance without mitotic figures.

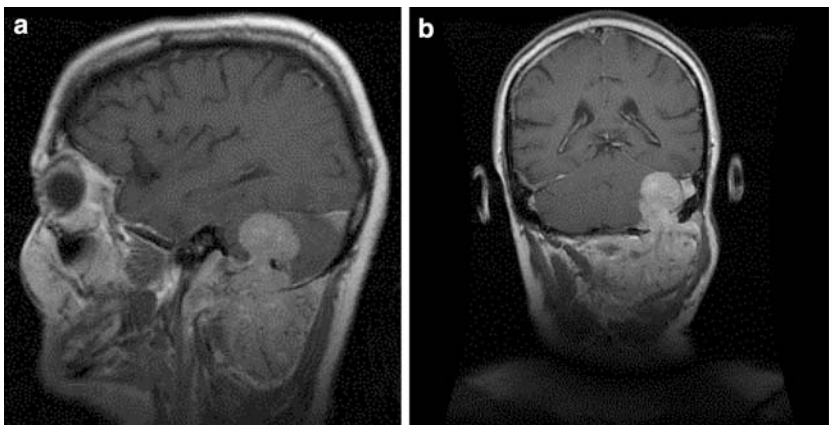

Fig. 2 **a** Parasagittal T1-weighted image shows a strongly but inhomogeneously enhancing neck-mass. The neck and posterior fossa mass imitates the form of a dumbbell. **b** Coronal T1-weighted image demonstrates a strongly enhancing lobulated neck-mass with a dumbbell appearance. The tumor invades the posterior fossa via an osteolytic defect in the skull (CT not shown)

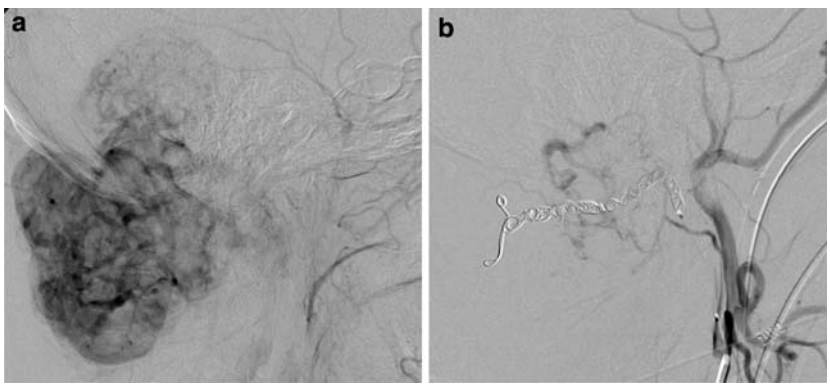

Fig. 3 **a** Lateral view of left external carotid angiogram. In the capillary phase dense enhancement of both, the extra- and intracranial part of the tumor is visible. Note the enlarged intratumoral vessels corresponding to flow-voids of MR-images. **b** Lateral view of left external carotid angiogram after embolisation of the tumor with particles (300–500 μm) and coil occlusion of the left occipital artery

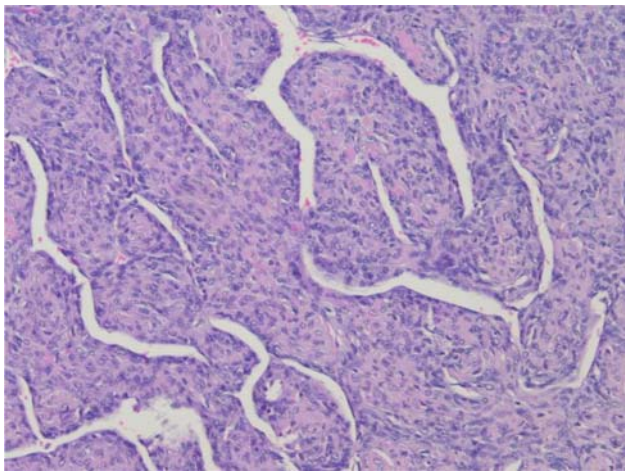

Fig. 4 Branching so called staghorn or hemangiopericytoma-like vessels are especially prominent in this field. Here, the tumor displays a rather homogenous cellularity (H&E, ×200)

Based on these morphological and immunohistochemical findings the tumor was classified as a hemangiopericytoma (HPC) (Figs. 4, 5).

Postoperative radiation therapy with an overall dose of 54 Gy and single doses of 1.8 Gy was performed. One and a half year later, follow-up examinations with MRT/CT showed no evidence of local recurrence or distant metastasis.

Discussion

Hemangiopericytomas are unusual aggressive, hypervascular neoplasms with sarcomatous characteristics arising from pericytic cells. The understanding of the pericytes and their pathology dates back to Rouget, Vimtrup and Krogh [1] who described amoeboid cells in the perivascular space. Zimmermann was the first to introduce the term "pericyte" in 1923 [2]. In 1942, Stout and Murray distinguished the HPC from other vascular tumors such as glomus tumors or hemangioblastomas [3]. The clinical and pathological heterogeneity of HPC makes a definitive diagnosis challenging. A number of neoplasms may mimic the appearance of HPC, these include the following: glomus tumor, solitary fibrous tumor, leiomyoma, leiomyosarcoma, infantile myofibroma, infantile fibrosarcoma, monophasic synovial sarcoma, mesenchymal chondrosarcoma and myopericytoma.

Intracranial HPC represent less than 1% of intracranial neoplasms [4–6]. They are to be differentiated from meningiomas preoperatively because profuse bleeding might reduce the chance for complete resection. HPC in the neck is rare and complete resection was reported to be very challenging [7, 8].

Macroscopically, HPC is described as an irregular lobulated grayish-pink mass with a delicate pseudocapsule. Lobulation of the tumor is due to retraction of the surface at the entrance point of vessels. The histopathology of HPC is characterized by tightly packed cells that grow around ramifying, thin-walled, endothelium-lined vascular channels ranging from larger sinusoidal spaces to small capillaries. The classic vascular pattern of HPC has been described as "staghorn" or "antler-like" in configuration. There may also occur perivascular or interstitial hyalinization, myxoid or cystic changes and, occasionally, lipomatous differentiation [9]. The role of immunohistochemistry in the diagnosis of HPC is considered as controversial. Lacking a specific marker for HPC, the value of immunohistochemical techniques consists in their capability to exclude other diagnoses.

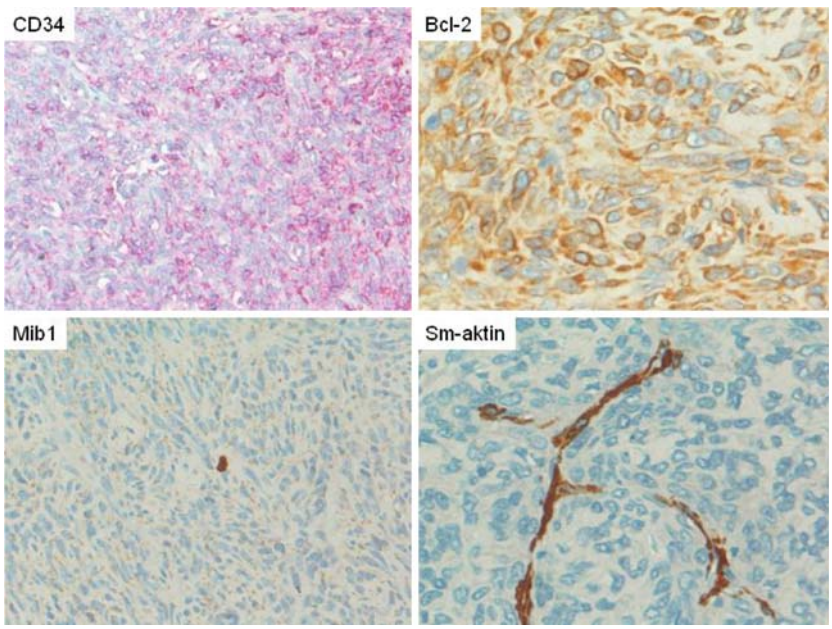

Fig. 5 This panel depicts the characteristic expression of CD34 and bcl-2, the proliferative activity as shown by the expression of Mib1 is below 3%. There is no expression of sm-actin, with tumor vessels acting as an internal positive control. Immunohistochemical stains with hematoxylin counterstain

In most cases expression of CD34 and bcl-2 is detected. In addition, focal sm-actin and desmin immunoreactivity may occasionally be found in HPC tissue.

On CT scans HPC presents as a hyperdense, inhomogeneous mass with intensive enhancement after contrast administration. Although mimicking intracranial meningeomas, the dural tail sign is infrequently observed in HPC. HPC tends to erode bone but reactive hyperostosis or intratumoral calcification has never been reported [4, 5, 10]. MRI depicts an inhomogeneous tumor matrix with low to intermediate T1 and intermediate to high T2 signal intensities. Prominent internal signal voids and marked gadolinium enhancement are typically found [4, 5, 10]. On angiography, dual vascularization of the external and internal carotid and vertebral artery, respectively, is often encountered [5]. Intensive tumor enhancement by multiple "corkscrew" feeders was described. In our case hypervascularity, inhomogeneous pooling of contrast agent and early venous drainage were most prominent.

HPC seems to be an insidious neoplasm with years of clinical stability and subsequent abrupt enlargement as in our case and that of others [8]. Although no parameters are known to predict the biological behavior and clinical course, a poor prognosis seems to be associated with a large tumor size and histological characteristics of malignancy like high cellularity, increased mitotic and proliferative activity, necrosis and haemorrhage [5, 10, 11].

The options in the management of intracranial HPCs comprise preoperative embolization, microsurgery, radiotherapy and gamma-knife surgery. Preoperative embolization of the feeding vessels has been reported as a powerful technique to limit blood loss during open surgery [5, 12]. Careful surgical excision of HPC is the treatment of choice. Intraoperative control and surgical resection of the tumor is difficult due to the high vascularity of HPC or, in addition, the extra- and intracranial extension as in our case [8]. Despite lacking therapeutic guidelines [5], complete en bloc resection of the tumor combined with excision of the contiguous dura mater is recommended. Guthrie et al. [6] pointed out the role of microsurgical techniques in reducing surgical complications. In order to decrease the risk of early recurrence of the tumor, surgery has to be carried out as radical as possible. Patients with an extended tumor, positive surgical margins, and histological high-grade lesions may benefit from adjuvant external-beam radiation therapy. A minimum of 50 Gy is recommended to prevent local recurrence [5, 6, 8, 13]. The value of palliative chemotherapy for the treatment of metastatic lesions has to be defined [5, 14].

Conclusion

HPC of the skull base represents a scarce pathological entity. The best treatment results for this tumor are achieved by preoperative embolization, complete en bloc resection and postoperative radiation therapy with a minimum dose of 50 Gy. Prolonged follow-up of the patients is strongly recommended because recurrence of the disease and distant metastases can occur even after years.

References

1. Berdjis CC (1957) Transplantable and metastasizing hemangiopericytoma in the rat' comments on the morphological aspects of the pericytic cells. Oncologia 10:336–347
2. Zimmermann KW (1923) Der feinere Bau der Blutkapillaren. Z Anat Entwcklngsgesh. 68:29
3. Stout AP, Murray MR (1942) Haemangiopericytoma: a vascular tumor featuring Zimmermann's pericytes. Ann Surg 116:26–33
4. Chiechi MV, Smirniotopoulos JG, Mena H (1996). Intracranial hemangiopericytomas: MR and CT features. AJNR 17:1365–1371
5. Fountas KN, Kapsalaki E, Kassam M, Feltes CH, Dimopoulos VG, Robinson JS, Smith JR (2006) Management of intracranial meningeal hemangiopericytomas: outcome and experience. Neurosurg Rev 29:145–153
6. Guthrie BL, Ebersold MJ, Scheithauer BW, Shaw EG (1989) Meningeal hemangiopericytoma: histopathological features, treatment, and long-term follow-up of 44 cases. Neurosurgery 25:514–22
7. Horky JK, Chaloupka JC, Putman CM, Roth TC, Sasaki CT (1997). Large haemangiopericytoma of the posterior cervical space. The role of imaging and therapeutic embolisation in the management of an unsuspected hypervascular head and neck neoplasm. Intervent Neuroradiol 3:155–160
8. Musacchio M, Mont'Alverne F, Belzile F, Lenz V, Riquelme C, Tournade A (2003) Posterior cervical haemangiopericytoma with intracranial and skull base extension. Diagnostic and therapeutic challenge of a rare hypervascular neoplasm. J Neuroradiol 30:180–187
9. Weiss SW, Goldblum JR (2001) Enzinger and Weiss's soft tissue tumours, 4th edn. C. V. Mosby, St Louis
10. Akiyama M, Sakai H, Onoue H, Miyazaki Y, Abe T (2004) Imaging intracranial haemangiopericytomas: study of seven cases. Neuroradiology 46:194–197
11. Constantinides H, Toh C, Warfield AT, Johnson AP (2002) Haemangiopericytoma of the submental region. J Laryngol Otol 116:969–972
12. Jaaskelainen J, Servo A, Haltia M, Wahlstrom T, Valtonen S (1985) Intracranial hemangiopericytoma: radiology, surgery, radiotherapy, and outcome in 21 patients. Surg Neurol 23:227–236
13. Carella RJ, Ransohoff J, Newall J (1982) Role of radiation therapy in the management of meningioma. Neurosurgery 10:332–339
14. Galanis E, Buckner JC, Scheithauer BW, Kimmel DW, Schomberg PJ, Piepgras DG (1998) Management of recurrent meningeal hemangiopericytoma. Cancer 82:1915–1920