Lipomatous Hemangiopericytoma of the Sellar Region: Case Report and Review of the Literature

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Abstract. Lipomatous hemangiopericytoma is a rare variant of hemangiopericytoma. Only a few cases of LHPC have been reported to date, with only two cases of this entity having been reported in the head and neck area, and none intracranially. We present the clinical, radiological, and histological features of the first reported case of a sellar lipomatous hemangiopericytoma, and review the literature. Material and Methods. We report a case of a 51-year-old man who presented with visual field compromise and was thought to have a pituitary adenoma based on clinical, radiographic and endocrinologic workup; a sellar mass with suprasellar extension was found. Results. Histological and immunohistochemical studies demonstrated evidence of a lipomatous hemangiopericytoma of the sellar region. Conclusion. A handful of sellar hemangiopericytomas mimicking pituitary adenomas have been previously described. Although one case of lipomatous hemangiopericytoma has been reported in the soft tissues of the occipital region, and one in the skull base and jugular foramen, to our knowledge this is the first reported case of an intracranial (sellar) lipomatous hemangiopericytoma. This tumor represents a distinctive pathologic entity that can only be diagnosed on histological examination.

Key Words: Lipomatous Hemangiopericytoma, Hemangiopericytoma, sella, transsphenoidal approach.

Introduction

Lipomatous hemangiopericytoma (LHPC) is a recently recognized rare hemangiopericytoma (HPC) variant that is histologically composed of an admixture of benign hemangiopericytomatous and mature lipomatous components. There have been a total of 50 pathologically-confirmed cases reported in the literature, none of which were located intracranially. Even though a handful of hemangiopericytomas have been described in the sellar region, there has never been a case of a lipomatous variant described in this region. Herein, we report such a case and present a review of the literature.

Case report

A 51-year-old male initially presented to an ophthalmology clinic with a two-month history of left eye visual disturbances associated with intermittent headaches, nausea, and vomiting. On physical examination there was evidence of optic disk edema. An MRI of the brain showed evidence of a sellar mass, and the patient was referred to our neurosurgical clinic. On physical examination his visual acuity was 20/30 OD, and 20/70 OS. His pupillary reflexes were normal without evidence of relative afferent defect. Formal visual testing showed evidence of a superior quadrantanopia in the right eye. His brain MRI showed a large lobulated lesion arising from the pituitary fossa, extending into the suprasellar cistern, and displacing the optic chiasm superiorly on the left. The mass measured 3.0x2.3x2.0 centimeters in transverse, craniocaudad, and AP dimensions, respectively. Focal areas of low signal intensity indicating calcification were found, and the mass invaded the cavernous sinuses bilaterally.
Preoperative endocrine workup showed a prolactin level of 28ng/mL, an ACTH of 15pmol/L, an FSH of 3.2IU/L, an LH of 0.32IU/L, a TSH of 1.10mIU/L, a total testosterone of 14nmol/L, a cortisol of 1.6μg/dL, and a free T4 of 0.6ng/dL.

A clinical diagnosis of a nonfunctional pituitary adenoma was made and the patient underwent elective surgical intervention. A trans-sphenoidal sublabial transeptal approach was performed. During surgery, a markedly vascular, soft, red tumor was found. A significant amount of bleeding from the tumor bed was controlled with bipolar cautery and topical hemostatic agents. Intra-operatively, there was evidence of a minimal amount of cerebrospinal fluid (CSF) leak from the region of the diaphragm sellae to which the tumor was quite adherent. A fat graft was harvested, and the dural defect was repaired. Once the procedure was completed, a lumbar drain was placed, and the patient was monitored in the ICU. The postoperative course was uneventful, the lumbar drain was removed on postoperative day 3, and there was no postoperative evidence of CSF leak or endocrine abnormalities.

Results - Pathology

Histological studies showed a neoplasm made of spindle to epithelioid cells with a rich vascular component (Figure 1 C). The vascular component consisted of thin, arcuate, and “stag-horn” type vessels, as well as vascular telangiectatic areas and thick-walled blood vessels. There was evidence of previous hemorrhage with focal collections of hemosiderophages. The tumor showed no necrosis or mitoses. A lipomatous component was scattered throughout the tumor and consisted of mature adipose tissue (Figure 1 A, B, and C). A rich reticulin network was present with reticulin fibers surrounding individual tumor cells (Figure 1 D and E). Immunohistochemical staining was performed with antibodies to CD34, CD31, CD99, BCL-2, S100 protein, glial fibrillary acidic protein (GFAP), synaptophysin, vimentin, smooth muscle actin (SMA), Factor VIII related antigen, epithelial membrane antigen (EMA), and pan-keratin. The neoplastic cells were strongly positive for vimentin and BCL-2. Focal areas of the tumor were positive for GFAP and S100 protein. CD99 showed cytoplasmic punctate staining. The neoplastic cells were negative for CD34 (highlighting the vascular component of the tumor; Figure 1 F), CD31, EMA, smooth muscle cell actin, synaptophysin, and pan-keratin. A diagnosis of lipomatous hemangiopericytoma was made.

Follow-up. The patient was seen in clinic for follow-up with 2 month post-op imaging. MRI revealed residual compression of the optic chiasm. Despite clinical improvement in visual symptoms it was decided to perform a craniotomy for resection of the residual tumor and decompression of the optic apparatus. Gross total extirpation was achieved.

Discussion

Hemangiopericytomas (HPC) are rare vascular neoplasms with unpredictable malignant potential. Accounting for less than 1% of primary intracranial neoplasms and roughly 1% of all blood vessel-related neoplasms [1], they arise from the capillary pericyte, which surrounds the endothelial cell and regulates the luminal diameter of the vessel. Bailey et al. first described HPC in 1928 as an angioblastic meningioma [2]. However, it was not until 1942 that Stout and Murray first coined the term hemangiopericytoma (3). Over the next half-century there were several case reports of HPC both inside and outside the CNS [2,4-12].

One particularly uncommon variant is the lipomatous hemangiopericytoma (LPHC), also known as fat-forming HPC, lipomatous solitary fibrous tumor (SFT), and fat-forming SFT, first described by Theunissen et al. in 1990 [12] and so named by Nielson et al. in 1995 [13]. Since its discovery, there have been about 50 reported cases. The majority are found in the deep soft tissue of either the lower extremities or retroperitoneum [1,14]. However, there have been reported cases involving the orbit, skull base, and spine, as well as the soft tissues of the head and neck region [1,15-17]. A handful of hemangiopericytomas of the sellar region have been reported in the literature, but to our knowledge there has never been a lipomatous variant described in the sellar region.
Common neoplasms arising from the sellar/suprasellar region include pituitary adenomas with suprasellar extension, meningiomas, craniopharyngiomas, gliomas, and germinomas. Sellar and parasellar hemangiopericytomas and the lipomatous variant can mimic the signs and symptoms of pituitary adenomas, and these entities should also be considered in the differential diagnosis of lesions in this area. Due to the vascular nature of hemangiopericytomas, excessive bleeding can complicate surgical resection of these lesions in the sellar region, and complete resection may not be feasible.

Figure 1. A. Lipomatous and vascular lesion H&E x 100 B. Mature adipose tissue and hemangiopericytoma H&E x 200. C. Mature adipose tissue and hemangiopericytoma H&E x 200. D. Lipomatous and vascular lesion Reticulin x 100. E. Lipomatous and vascular lesion Reticulin x 200. F. Lipomatous and vascular lesion CD34 x 200
In our case, bipolar cauterization and hemostatic agents controlled the excessive bleeding that occurred during resection, and a subtotal resection was achieved.

Hemangiopericytomas have a slight male predominance and typically present in the fourth to fifth decade [6,8,10], usually secondary to mass effect. Extracranial HPCs and deep soft-tissue lipomatous HPCs typically present as slow growing, minimally tender masses [1,2,4-14,18]. The only reported skull-base LHPC presented with unilateral facial paraesthesia, blurred vision, headache, and light-headedness [16]. Orbital LHPCs presented with acute eye pain, epiphora, and proptosis. Finally, the spinal LHPC presented with low back pain, erectile dysfunction, and L5/S1 paraesthesia [17]. If the HPC is intrasellar or intracranial, this manifests predominantly with headaches, visual deficits [2,5,6,8-12], and endocrinological changes (i.e., acromegaly) [4,7]. In our case, the patient presented due to evidence of visual compromise caused by the suprasellar extension of the tumor and optic nerve/chiasmal compression, as well as non-specific symptoms of worsening headaches. Many authors have described that HPCs mimic other tumor types on presentation and imaging. Intracranially they can appear clinically and radiologically identical to meningiomas or pituitary adenomas [5,6]. In our case, the imaging findings seen on MRI, the clinical presentation, and the hormonal workup led us to believe that the patient had a nonfunctional pituitary macroadenoma. However, in retrospect the MRI demonstrated unusual vascularity for pituitary adenoma.

While meningioma and pituitary adenoma are, perhaps and for our purposes, the most common neoplastic entities mimicked by HPC/LHPC, many authors suggest considering several other entities, depending on the anatomic location of the lesion. Liposarcoma [1,14], glomus jugulare for skull base lesions [16], myolipoma, angiomylipoma, hamartoma, sclerotic lipoma [16], neurofibromma, schwannoma, and ependymoma for spinal tumors [17] are reasonable to include in the initial differential diagnosis.

For the clinician, the take-home point is that a non-invasive definitive diagnosis is not possible from such a widely variable differential diagnostic list. Thus, surgical resection and pathologic evaluation of the tumor is required.

Grossly, lipomatous HPC have been repeatedly described as well-circumscribed, tan/white, rubbery lesions with a variable amount of admixed yellow, grossly fatty regions [1,14,15,18]. In the largest reported series, a pseudo-capsule was universally described, with rare descriptions of infiltrative growth [14]. Microscopically, these lesions demonstrate a lobular pattern with bimodal differentiation: regions of hypercellular nodules exhibiting the classic appearance of HPC (spindle cells arranged in short, whorling fascicles surrounding stag-horn type vessels), interspersed with regions of mature adipocytes [14,16-18]. While the percentage of fat varied from case to case, these lesions were almost universally described as having low or no atypia or mitotic activity. Importantly, necrosis, vascular invasion, and lipoblasts have never been described [1,14]. Some authors suggest that immunohistochemical staining aids in definitive diagnosis. The majority of cases stained positive for type IV collagen, factor XIIIa, smooth muscle actin, Bcl-2, vimentin, and CD34 [1,14,15,18], and several stained positive for S100 [1,14,18]. However, the gross, microscopic, and immunohistochemical characteristics have not been detailed in all the reported cases. In our case, the neoplastic cells were strongly positive for vimentin and BCL-2, with focal areas positive for GFAP and S100 protein. While the neoplastic cells were negative for CD34, up to 10% of solitary fibrous tumors are also negative with this antibody [17].

The only way to definitively diagnose LHPC is through surgical removal. Such an approach provides enough tissue to yield sufficient proportions of both the HPC and lipomatous components, solidifying the diagnosis of lipomatous hemangiopericytoma [14,17]. Unfortunately, such an approach may not always be feasible in the sellar region due to tumor invasion of nearby neurovascular structures.
The role of adjuvant radiotherapy in the treatment of HPC has been well established because of its accepted malignant potential [5,6,8-11]. However, because the malignant potential for LHPC is quite low, initial treatment should consist of local or gross resection. Stereotactic radiosurgery should be reserved for incomplete resection and recurrent or metastatic disease [16].

In conclusion, to the best of our knowledge, this is the first reported case of a lipomatous HPC tumor affecting the sellar region. Tumors in this area can mimic the signs and symptoms of pituitary adenomas, and this entity should be included in the differential diagnosis of sellar regions. If complete surgical resection is not feasible due to excessive hemorrhage or invasion of nearby neurovascular structures, close follow-up and expectant management is recommended.

References