POSTERIOR CERVICAL HAEMANGIOPERICYTOMA WITH INTRACRANIAL AND SKULL BASE EXTENSION

Diagnostic and therapeutic challenge of a rare hypervascular neoplasm

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SUMMARY

Haemangiopericytomas are rare hypervascular tumors arising from pericytes. They may occur anywhere in the body, but posterior cervical location is rather uncommon. A case of posterior cervical haemangiopericytoma with posterior fossa and temporal bone extension is reported. Although the patient had undergone preoperative endovascular embolization and surgical resection on three separate occasions, control of the skull base extension was not successful. Following endovascular embolization combined with radiotherapy, the patient has been asymptomatic for 48 months. Angiographic features may help in differentiating haemangiopericytomas from other hypervascular lesions. Preoperative endovascular embolization is recommended due to the pronounced tendency for haemorrhage throughout biopsy and surgical procedures.

Reported case

A 56 year-old woman presented with a long history of asymptomatic right suboccipital tumor. Physical examination revealed a well-delimited...
painless subcutaneous mass. Neck lipoma was initially diagnosed. The lesion had remained clinically unchanged for 18 years, but had progressively increased in volume in the last few years. Several weeks prior to presentation, the tumor showed abrupt enlargement associated with throbbing local pain radiating to the head and neck with stabbing paroxysms. Examination at that time disclosed a hypervascular pulsatile tumor with a systolic thrill located in the right suboccipital area, partially extending to the upper neck.

Non-enhanced CT scan showed a well-defined mass, 7 cm in its greatest diameter and slightly hyperdense compared with muscle, within the right upper posterior cervical space and in contact with the occipital bone, the right mastoid and the upper cervical spine (figures 1a and 1b). Images also showed two well-circumscribed, rounded cellular compartments of lesser density attenuation. The right portion of the occipital nuchal plane, the right atlantooccipital joint and the right transverse process of the atlas presented signs of bone erosion. The tumor enhanced intensely after contrast injection. The areas of lesser attenuation identified at initial scans also enhanced but mildly. Moreover, contrast injection revealed a broad-based mass in the right side of the posterior fossa close to the foramen magnum that was not visible on the non-enhanced CT images (figures 1c and 1d).

MR imaging demonstrated a well-circumscribed tumor with important peritumoral vessel flow voids, exhibiting three well-delimited areas of low and intermediate signal intensity on T1-weighted images, and high signal intensity on T2-weighted images. The tumor disrupted the hypointensity of the occipital cortical bone. After gadolinium injection, intense but slightly heterogeneous enhancement of the mass was seen, with extension to the meninges of the right side of the posterior fossa and tentorium, right jugular foramen and right mastoid cells. 3D-TOF sequences disclosed a high signal blush defining the neovascular bed and the enlarged feeding vessels from the right external carotid artery (figure 2).

Digital angiography disclosed a large, highly hypervascular tumor with a persistent, intense neovascular blush, staining heterogeneously. The major feeding arterial vessels were muscular branches from an abnormally enlarged right occipital artery. In addition, minor feeders of the right auricular artery were also observed and the posterior branch of the right middle meningeal artery supplied the intracranial portion of the tumor. Right vertebral angiogram demonstrated further arterial supply from muscular branches of the V3 segment (figures 3a and 2b).

Preoperative endovascular embolization was attempted. The right occipital artery was selectively catheterized with a 5 French catheter and a suspen-

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**Fig. 1.** – CT scans. a) Right, well-defined, slightly hyperdense extracranial mass attached to the occipital bone, with two cellular compartments of lesser density attenuation. b) Erosion of the occipital bone and mastoid. c) Areas of lesser attenuation identified at initial scans also enhanced but mildly.

**Fig. 1.** – TDM. a) Masse extracrânienne de limites nettes, attachée à l’os occipital, hyperdense par rapport aux muscles, montrant des aires bien définies de densité moindre. b) Érosion de l’os occipital et de la mastoïde à droite. c) Les zones de densité moindre décrites au sein de la tumeur prenaient le contraste mais de façon moins intense.
sion of 300-600 micron polyvinyl-alcohol (PVA) particles was injected. Postembolization angiography demonstrated satisfactory tumor devascularisation with parent vessel conservation (figure 3c). Thus, no other arterial pedicles were treated.

The extracranial mass was surgically removed. The nuchal tumor enclosed an enlarged, right occipital artery, which was carefully dissected and preserved. The lesion was also slightly adherent to muscular and aponeurotic planes but was easily dissected without significant intraoperative haemorrhage. Biopsy results initially suggested meningioma. Therefore, the intracranial portion of the tumor was not immediately removed. Patient progress was carefully monitored though rigorous clinical follow-up.

Eight months later, the patient complained of increasing deep pain in the right auricular region with right pulsatile tinnitus. Neurological examination was normal. MR images revealed a significant increase of the intracranial mass dimensions that extended to the right lateral cerebellar and right cerebellopontine angle cisterns, and slightly infiltrated the dura and aponeurosis in the craniotomy area. The tumor had mild mass effect over adjacent cerebellar structures and invaded the right petrous bone and the right sigmoid sinus. It was hyperintense with respect to the cerebellar cortex and showed a slightly heterogeneous pattern on T2 weighted sequences. Veins of the right lateral recess of the fourth ventricle showed important signs of stasis. The mass showed intense homogeneous enhancement with a dural tail sign. Antero-inferior mastoid cells yielded higher signal intensity than CSF on T2-weighted sequences, probably related to the retention of high-protein concentration reactive or inflammatory fluid (figure 4). CT scan confirmed the extension to the posterior mastoid cells, the right external ear and the middle ear, and demonstrated partial destruction of the inferior portion of the ossicular chain (figure 5).

Pre-operative endovascular embolization was attempted a second time. An angiogram disclosed three main arterial feeders: right auricular, right ascending pharyngeal and right occipital arteries. Early venous drainage to the right lateral sinus was
observed. Each of the arterial pedicles was selectively catheterized with a 5 French catheter and embolized with calibrated microspheres 150-300 microns in diameter. Post-embolization selective injections demonstrated an important reduction of the neovascular blush.

One day later, the patient was taken to surgery. The intracranial mass and the compromised segment of the right sigmoid sinus were resected without complications. However, surgical dissection of the petrous bone component was difficult and it could not be removed completely.

A new histopathological analysis of the extracranial portion disclosed that the nuchal tumor was composed of short spindle to oval cells forming a staghorn pattern around normal dilated vascular channels. Neither mitotic activity nor nuclear atypia were seen. Immunohistochemical analysis indicated that the tumor reacted with antibodies against vimentin and reticulin. Fusiform cells did not express cytokeratin, actine or s-100 protein. The intracranial component of the tumor presented the same histological and immunohistochemical characteristics as the extracranial mass, but this time some mitotic figures were seen. A diagnosis of “haemangiopericytoma-like tumor” was then proposed.

The patient was asymptomatic for 2 years, after which she gradually developed a cerebellar syndrome associated with intracranial hypertension. MR imaging demonstrated a local recurrence in the right petrous bone and mastoid (figure 6). Endovascular embolization followed by external radiotherapy was performed. The patient has remained asymptomatic with no evidence of local progression at clinical and radiological follow-up during the last 48 months.
Haemangiopericytoma is an uncommon hypervascular mesenchymal neoplasm originating from modified contractile smooth muscular cells that surround capillaries and postcapillary venules, called pericytes of Zimmerman. It represents less than 2-5% of soft tissue sarcomas and about 1% of all vascular tumors [11, 13, 15]. It occurs at all ages with a predilection to middle age [13, 20, 29] and sex distribution is almost equal [15, 20].

Haemangiopericytomas are ubiquitous tumors. The most frequent areas of occurrence are inferior extremities (particularly in the soft tissue of the thigh), retroperitoneum, pelvic region, and the head and neck [13, 16, 20, 29].

Approximately 13% to 25% of haemangiopericytomas originate in the head and neck [4]. In order of descending frequency, the principal extracranial locations include orbit, tongue, nasopharynx, nose, paranasal sinuses, mandible, and pharynx [8]. The posterior cervical region is an extremely uncommon area for the development of haemangiopericytomas [8, 22] and only two cases have been previously reported [22, 31]. Although age and gender prevalences of extracranial haemangiopericytomas have not been stated, they seem to be basically adult tumors that affect both sexes with no clear gender predominance [34, 38, 39]. Intracranial haemangiopericytomas only represent 0.29% of all intracranial neoplasms [33]. However, they constitute 2.4% of meningial tumors [21] and their anatomic distribution is approximately similar to that of meningiomas [9, 31].

Although the radiologic presentation of haemangiopericytomas may be rather variable, they show typical but non-pathognomonic characteristics [9, 29-31]. Most of the studies available in the literature do not describe precisely the radiological features of haemangiopericytomas of the head and neck [1, 3, 4, 6, 10, 31, 33, 38, 39].

As we could see in the present case, haemangiopericytomas generally progress slowly and are usually revealed clinically by sudden enlargement or non-specific mass effect on adjacent structures [13, 22, 38]. The slow, silent growth and the existence of degenerative phenomena may account for the large lesions frequently observed at diagnosis and the presence of necrosis, haemorrhage, pseudocysts, or calcifications occasionally detected at imaging studies. This particular course of development may explain the heterogeneous pattern usually observed on both CT and MR images [9, 20, 30, 31, 33, 38].

Extracranial haemangiopericytomas may undergo skull base and/or intracranial growing [1, 3, 4, 6, 8, 10, 12, 24, 34]. Most of head and neck tumors with endocranial extension were located close or adjacent to a skull base foramen, or experienced important growth during subclinical evolution [1, 3, 4, 6, 8, 10]. Conversely, intracranial lesions can experience extracranial extension, in particular at the time of local recurrence when surgical changes facilitate regional spread [33]. Only one of the reported cases of posterior neck haemangiopericytomas showed intracranial extension [31]. In our case, the nuchal occipital plane, which was in contact with the extracranial portion of the tumor, showed bone erosion of the inner and external tables close to the foramen magnum, and enlargement of multiple trans-osseus vascular channels by neoplastic infiltration was seen during surgery. Thus perivascular infiltration might explain intracranial extension of the neoplasm to the posterior fossa. In head and neck tumors, intracranial development might be considered as a predictive sign of potential aggressive behaviour, because it was frequently associated to a higher rate of local recurrence [1, 2, 10, 34].

Osseous changes associated with haemangiopericytoma may be primary or secondary to tumor development [12]. Osteolytic and/or pressure-induced erosion or destruction of adjacent bones are usually seen [1, 3, 4, 6, 9, 10, 12, 15, 31]. Bone destruction appears to be associated with a more aggressive local behaviour in haemangiopericytomas, as in our patient. One case of posterior neck haemangiopericytoma [31] associated destruction of the adjacent nuchal occipital plane and extension to the posterior fossa. This tumor finally recurred and metastasised years later. Murphey et al. [29] described osteolytic lesions with honeycomb and hole-within-whole appearances, osseous expansion, and cortical permeation in more undifferentiated musculoskeletal haemangiopericytomas. Chiechi et al. [9] observed bone erosion in most anaplastic intracranial haemangiopericytomas, but in only about 50% of well differentiated tumors. Conversely, no bone hypertrophic response is described in meningeval haemangiopericytomas [9, 31, 33, 35]. Thus, the absence of hyperostosis might allow for the differentiation of haemangiopericytomas from meningiomas intracranially. However, peripheral, lower-grade haemangiopericytomas may present bone sclerosis, particularly at the osseous margin [29].

Calcifications are infrequent in haemangiopericytomas [30, 31, 33, 35, 38]. Unlike meningiomas, meningeval haemangiopericytomas rarely present intratumoral calcifications [31, 32, 35]. They are more frequently observed in peripheral tumors as mild calcifications with marginal or sparkled configuration [9, 31, 33]. Nevertheless, no calcifications were detected in head and neck haemangiopericytomas [1, 3, 4, 6, 10, 36, 38, 39]. As in the previously reported cases of posterior cervical haemangiopericytomas [22, 31], no calcifications were found in our case.

Intracranial tumors are isodense or slightly hyperdense compared with brain tissue at non-enhanced CT [9, 31, 35]. They are typically heterogeneous [6, 9, 18, 24-27]. However, some tumors may have a homogeneous pattern or be less dense than the brain [11]. Head and neck [34, 39] and peripheral haemangiopericytomas [29, 30] have similar attenuation to that of muscular tissue. However in one case of posterior neck haemangiopericytoma CT scans disclosed a slightly hyperdense tumor [31], as in the present case (figure 1).

Intracranial, extracranial and peripheral haemangiopericytomas are similar, based on MR imaging [9, 14, 19, 25, 29, 30, 33] and posterior cervical tumors present the same signal characteristics [22] (figure 2). They are isointense or hypointense on T1-weighted images. On T2-weighted sequences, they usually give high signal intensity. However, Chiechi et al. [9] ob-
served that almost 60% of the intracranial haemangiopericytomas were isointense on T2-weighted images and hyperintense in only 26% of cases. Furthermore, intracranial haemangiopericytomas can show a slightly brighter signal compared to brain tissue on proton-density-weighted images [12].

Intense diffuse enhancement with well-delineated margins can be observed after iodine-contrast or Gd-DTPA injection in most haemangiopericytomas [9, 12, 25, 31], including posterior cervical tumors [22, 31]. These neoplasms produce a critical network of penetrating capillaries that may allow accumulation of contrast products in the expanded extracellular space [12]. Although enhancement can be homogeneous [35], a heterogeneous pattern is more often observed after contrast administration [9, 14, 28] and some haemangiopericytomas only enhance in the periphery, revealing a thick and regular rim-like pattern [33, 35]. Heterogeneity is due to the presence of necrotic or pseudocystic areas, which are frequently observed in large tumors [9, 14, 19, 28-30, 33].

We observed well-defined zones with slightly varying degrees of density attenuation, signal intensity, and enhancement after contrast administration within the mass (figure 1 and figure 2). This observation may not necessarily be associated with necrosis, haemorrhage, calcifications, or pseudocystic degeneration, and might be explained by the presence of compact cellular or loose cellular components within the same tumor that can be seen on pathological analysis [12].

Haemangiopericytomas showed prominent intratumoral and peritumoral vessel flow voids [9, 12, 25, 29, 30]. In most cases, vascular channels demonstrate low signal intensity on all MR sequences, indicating high blood flow [9, 12, 25, 29, 30]. The presence of these structures may allow the differentiation of meningeal haemangiopericytomas from meningiomas [9, 12, 25, 33, 38], but not from their angioblastic form [33]. Nevertheless, these vascular channels can also be observed at MR imaging in arteriovenous haemangiomas, haemangoendotheliomas and angiosarcomas and sometimes in other extracranial hypervascular tumors, such as alveolar soft-part sarcomas, rhabdomyosarcomas, and extraoesophageal Ewing sarcomas [29, 30]. However, flow voids occur most often in the periphery of the mass and are particularly prominent in haemangiopericytomas [29] (figure 2b).

MR angiograms may merely disclose a faint high signal blush due to the neovascular bed and dilated feeding vessels [22]. In our experience, the three-dimensional time of flight angiographic sequences only reveal the hypervascular nature of the neoplasm. The origin and number of principal feeders could not be determined precisely and no other specific signs were observed (figure 2c).

MR spectroscopy has been introduced recently in the diagnostic analysis of haemangiopericytomas. Intracranial haemangiopericytomas show a larger peak at 3.5 ppm owing to a high concentration of myoinositol that may allow differentiation from meningioma [5].

Sonographic evaluation has not proven useful for differential diagnosis. Peripheral haemangiopericytomas may be hypoechoic or hyperechoic with a complex structure showing pseudocystic zones owing to necrosis or haemorrhage [29, 38].

Characterization of tumor vascularity with precise identification of arterial feeders is extremely important for planning the therapeutic approach. Doppler sonography may be useful in vascular evaluation of haemangiopericytomas. Color and duplex Doppler ultrasound findings can demonstrate arteriovenous shunt in peripheral haemangiopericytomas, showing good correlation with angiographic features [23, 29]. This information may be used for guiding tumor biopsy in order to avoid profuse bleeding [23].

Angiography has an important role in patient management. Haemangiopericytomas may have a more typical angiographic appearance, compared with other hypervascular tumors [26, 27, 29]. Very few arterial pedicles are revealed by angiography [20, 29]. Major branches are often displaced by the tumor, and dilated serpentine feeding branches encircle the mass [29]. They converge upon a well-delimited microvascular bed, which accumulates contrast and provides a strong, well-delimited tumor blush [15, 20, 22, 27, 29, 31]. Manifest arteriovenous shunting and/or early venous drainage may be detected [8, 27]. Some have considered simultaneous cortical and meningeal arterial supply a sign of aggressive behaviour and an attribute distinguishing intracranial haemangiopericytomas from meningioma [31, 35, 37]. Marc et al. [27] have proposed 4 diagnostic criteria for intracranial haemangiopericytomas based on their most common angiographic characteristics, allowing differentiation from other hypervascular tumors, especially from meningiomas [27, 35]:

1. dual meningeal/cortical arterial supply,
2. one to three main feeders giving rise to many irregular corkscrew-like vessels,
3. a dense, well-defined and long-lasting tumor stain,
4. and early venous drainage rarely.

Nevertheless, these distinguishing angiographic features are more often found in large haemangiopericytomas, whereas the small ones are more similar to meningiomas [35]. Thus, angiography may improve the diagnostic value of other imaging modalities in some cases [19, 22, 33, 35].

Others have observed the same angiographic characteristics that meningeal haemangiopericytomas in non-meningeal tumors [20, 25, 31, 35]. In the present case, angiography disclosed similar findings. No pial blush was disclosed in carotid angiogram, but two small pial vessels that fed the intracranial mass were coagulated during surgical dissection. Abnormal, dilated, serpentine arterial feeders were observed in angiograms of both extracranial and intracranial portions of the tumor. The neovascular stain was intense, heterogeneous and well demarcated, and persisted during the venous phase in all angiographic acquisitions. Early venous drainage to the dural sinus was detected during the angiography of the residual intracranial tumor. Similar angiographic features were described by Horky et al. [22] in another case of posterior neck haemangiopericytoma.

The propensity to recur and to metastasise characterizes the natural evolution of most haemangio-
pericytomas [13-16, 18, 21, 28, 34, 36]. Primary or secondary extensive development along dural surfaces [2, 35] as well as hypercellularity and high mitotic activity [16] have been suggested as signs of aggressive biological behaviour. However, haemangiopericytomas have a variable malignant potential [13, 38], and nor radiological nor histological specific parameters have been identified to predict recurrences or metastatic dissemination [13, 20].

Haemangiopericytomas typically have a high rate of local recurrence [13, 32], in particular tumors of retroperitoneal or meningeal origin [13, 18, 21]. The disease-free interval may last for few months to decades [13, 32, 38]. Haemangiopericytomas of the head and neck also show a potential aggressive course [34]. They frequently present one or more recurrences despite initial wide surgical excision, multiple surgical procedures, and even after adjuvant radiotherapy [1, 2, 10, 34].

Local recurrence precedes metastases in more than 50% of cases [13]. Large or incompletely resected lesions, and histologically malignant tumors are associated with higher mortality rates [13, 18, 34]. The 10-years survival rate for patients with haemangiopericytomas smaller than 6.5cm has been reported to be 90% [4, 16]. Nevertheless, the overall mortality rate in the literature is about 50% [13]. Therefore, all haemangiopericytomas have to be treated as potentially malignant, and all patients should be considered for curative purpose, and undergo life-long follow-up.

Adequate surgical resection with negative margins appears to be the most appropriate therapy for haemangiopericytoma [18, 21, 24, 34, 38]. However, surgical resection may be impossible secondary to severe bleeding due to the hypervascular nature of the neoplasm [21, 32]. Many cases of extensive or severe bleeding during biopsy and surgery have been reported in the literature [8, 11, 32, 34] including two deaths as a result of massive uncontrolled intraoperative haemorrhage [16]. Consequently, embolization is mandatory to reduce intraoperative bleeding and avoid haemorrhagic complications as well as to facilitate dissection and complete resection of the mass.

Many embolic agents have been used for this purpose. PVA, gelatine sponge particles, N-butyl-2-cyanoacrylate (NBCA) and electrolytically detachable platinum micro-coils may be used separately or in combination to embolize the arterial feeders [8, 11, 12, 22, 33, 39]. Nonetheless, effective devascularisation may be dangerous or impossible in some cases of head and neck haemangiopericytoma as a result of further vascular supply from vertebral or internal carotid arteries, or previous surgical or endovascular treatment. In such situations, direct puncture of the tumor with percutaneous injection of NBCA may be safely and successfully used [7, 39].

Friedman and Egan [13] were the first reporting the use of irradiation in the treatment of haemangiopericytomas.

Adjuvant radiotherapy is considered a valuable option for local management of haemangiopericytoma [9, 13, 14, 16, 18, 28], regardless of the histologic subtype [28]. Moreover, radiotherapy may allow local control with good symptomatic relief in patients with bone metastasis and external beam radiation therapy may be useful in the management of primary inoperable tumors [18]. Dufour et al. [14] observed that the use of postoperative external radiotherapy in primary intracranial haemangiopericytomas reduced the risk of local recurrence and central nervous system metastasis, but did not protect against later metastatic lesions.

In addition, radiosurgery appears to be useful in the control of recurrent lesions smaller than 25mm [14, 18], but does not avoid further recurrence in these cases or after treatment of CNS metastasis in intracranial haemangiopericytomas [14]. The mean time of local disease control in those cases is longer if a total response is achieved after irradiation [18].

Chemotherapy has not proven to be useful for the treatment of resectable haemangiopericytomas [13, 18]. However, alone or in combination with radiation therapy, it has been used in the management of metastatic disease with variable results [13, 18, 34]. Although further studies and new drug combinations are needed [34], this approach represents the last chance of treatment in such cases.

CONCLUSION

Haemangiopericytomas of the posterior cervical space are uncommon hypervascular tumors. However, a diagnosis of haemangiopericytoma should be considered for all hypervascular lesions of the head and neck region due to the high risk of haemorrhage following invasive procedures, endocranial extension, and local recurrence or metastasis following inadequate treatment. Imaging features are not pathognomonic in haemangiopericytomas, but are necessary for presumptive diagnosis and pretherapeutic planning. Angiography may suggest the diagnosis of haemangiopericytoma, disclosing a typical pattern of neovascularity, and evidences of bone erosion or meningeal extension must be considered as signs of more aggressive biological behaviour. Therefore, haemangiopericytoma is a diagnosis of exclusion, and immunohistochemical analysis is necessary for diagnostic confirmation.

Wide surgical excision is the treatment of choice for all resectable haemangiopericytomas. Total resection must be achieved at the first surgical attempt in all lesions, even when histological diagnosis is not confirmed. Otherwise, the risks of recurrence and extension to neighbouring areas are high. Endovascular embolization has proven to be critical in avoiding hemorrhagic complications and facilitates safer, more aggressive surgical resection with satisfactory disease-free margins. Radiotherapy is a necessary complement to surgery to prevent or control local recurrence, and may be helpful in therapeutic management of tumors involving vital or delicate anatomic structures, in which dissection becomes risky or impossible.

Finally, we emphasize the importance of long-term clinical and radiological follow-up especially in cases with dural extension, because of the propensity for recurrence and metastasis, even years after adequate surgical resection or combined treatment.