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# Intracranial Hemangiopericytoma: Study of 12 Cases

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## Summary

Most hemangiopericytomas (HPCs) are located in the musculoskeletal system and the skin, while the intracranial location is rare. They represent 2 to 4% in large series of meningeal tumours, thus accounting for less than 1% of all intracranial tumours. Many authors have argued about the true origin of this tumour. The current World Health Organization classification of Central Nervous System tumours distinguishes HPC as an entity of its own, and classified it into the group of ''mesenchymal, non-meningothelial tumours''. Radical surgery is the treatment of choice, but must be completed with postoperative radiotherapy, which has proved to be the therapy most strongly related to the final prognosis. HPCs have a relentless tendency for local recurrence and metastases outside the central nervous system which can appear even many years after diagnosis and adequate treatment of the primary tumour.

Twelve patients with intracranial HPC were treated at our Unit between 1978 and 1999. There were 10 women and 2 men. Ten tumours were supratentorial and most located at frontoparietal parasagittal level. The most common manner of presentation was a focal motor deficit. All tumours were hyperdense in the basal Computed Tomography scans and most enhanced homogeneously following intravenous contrast injection. In 50% of cases, tumour margins were irregular or lobulated. Seven tumours were studied with Magnetic Resonance Imaging, being six of them iso-intense with the cortical gray matter on T1-weighted and T2-weighted images. Twenty operations were performed in the 12 patients. In 10 cases radical excision could be achieved with no operative mortality. Total recurrence rate was 33.3%. Eight patients were treated with external radiotherapy at some time through the course of their disease. Eight out of the 12 patients in this series are disease-free (Glasgow Outcome Scale categories 1 and 2) after a mean follow up of 52 months.

*Keywords:* Hemangiopericytoma; intracranial; meningeal; treatment; radiotherapy; prognosis.

## Introduction

Hemangiopericytoma (HPC) is a malignant tumour originating from Zimmermann's pericytes around capillaries and postcapillary venules which is most commonly located in the musculoskeletal system and the skin [24, 52].

Intracranial HPCs are rare as they represent 2 to 4% of meningeal tumours in large series, thus comprising less than 1% of all intracranial tumours [13, 20, 21, 26].

The histogenesis of this tumour has been a matter of controversy for a long time. In 1942, Stout and Murray identified a soft tissue tumour located primarily in the thigh, buttock and retroperitoneum which apparently consisted of proliferating pericytes and called it HPC [52]. In a later personal series these authors reported on 25 new cases of HPC, of which only one was intracranial and apparently invaded the meninges [53].

However, it was not until 1954 when Begg and Garret first reported a primary cranial meningeal hemangiopericytoma. They noted that it was histologically identical to both the soft tissue HPC previously described by Stout and Murray and the aggressive variant of angioblastic meningioma reported by Cushing and Eisenhardt in their classical monograph [5, 11].

Since then, many authors have argued about the true origin of this tumour and whether or not those found intracranially should be included in the group of meningiomas. Popoff *et al.* proposed not to classify this tumour as a meningioma, because it is identical to HPCs arising in soft tissues [43]. By contrast, Horten *et al.* after reviewing 79 cases of angioblastic meningiomas, which showed areas apparently transitional between HPC and fibrous meningiomas or hemangioblastomas, concluded that these tumours arise from multipotential presursor cells and should be classified with the group of meningiomas [24, 36].

The current World Health Organization (WHO) classification of Central Nervous System tumours distinguishes HPC as an entity of its own, and classified it with the group of ''mesenchymal, non-meningothelial tumours'' [29]. The International Classification of

Table 1. *Clinical Data, Treatment and Final Outcome in 12 Patients with Intracranial Hemangiopericytoma*

| *Case no.* | *Age sex* | *Tumor location* | *Clinical data* | *CT basal & postcontrast* | *Size (cm.)* | *Borders* | *Oedema* | *MR basal & postcontrask* |
|---|---|---|---|---|---|---|---|---|
| 1 | 58 F | F-P parasagittal left | 1 yr hemipareris tremor | hyperdense homogeneous enhancement | 8 × 4 × 4 | lobulated | ++ | not stated |
| 2 | 72 F | P parasagittal right | 2 mths hemiparesis | hyperdense cystic homogeneous enhancement | 4 × 3 × 3 | regular | ++ | not stated |
| 3 | 66 F | F-P parasagittal left | 2 mths monoparesis | hyperdens homogeneous enhancement | 4 × 3 × 2 | lobulated | − | not stated |
| 4 | 75 F | P right | 1 mth hemiparesis behaviour changes | hyperdense homogeneous enhancement | 6 × 6 × 3 | regular | ++ | not stated |
| 5 | 12 F | CPA right | 4 mths deaph facial paresis | hyperdense bone erosion calcium homogeneous enhancement | 4 × 3 × 2 | regular | − | not stated |
| 6 | 39 M | optic canal right | 2 mths vision loss | erosion homogeneous enhancement | 2 × 2 × 2 | regular | − | Iso T1 T2 homogeneous enhancement |
| 7 | 40 F | falx bilateral | 1 wk monoparesis | heterogeneous heterogeneous enhancement | 10 × 5 × 5 | lobulated | + | Iso T1 T2 heterogeneous enhancement |
| 8 | 23 M | P parasagittal bilateral | 2 mths seizure monoparesis | hyperdense obst SLS homogeneous enhancement | 5 × 4 × 4 | lobulated | ++ | Iso T1 T2 homogeneous enhancement |
| 9 | 38 F | optic canal left | 3 mths diplopia blurred vision | hyperdense bone erosion homogeneous enhancement | 2 × 1.5 × 2 | regular | − | Iso T1 T2 homogeneous enhancement |
| 10 | 62 F | F convexity left | 2 mths motor disphasia | hyperdense homogeneous enhancement | 2.5 × 3 × 2 | lobulated | − | Iso T1 T2 homogeneous enhancement |
| 11 | 25 F | occipital left | 2 mths headache seizure | heterogeneous cystic no enhancement | 3 × 2.5 × 3 | regular | ++ | heterogeneous T1 T2 hemosiderin mild enhancement |
| 12 | 29 F | tentorial right | 6 mths gait ataxia vertigo | hyperdense cystic homogeneous enhancement | 5 × 3 × 3 | lobulated | − | Iso T1 T2 homogeneous enhancement |

Diseases for Oncology (ICD-O) and the Systematized Nomenclature of Medicine (SNOMED) consider 9150/1 as the morphology code for HPC (behaviour is coded "/1" for low or uncertain malignant potential or borderline malignancy tumours) [30].

In spite of the above mentioned controversies regarding the histogenesis of HPC, there is a general agreement that it is more aggressive than meningioma, showing a propensity for both local recurrence and extraneural metastases, which may appear even many years after initial diagnosis.

The purpose of this paper is to describe the clinico-radiological features and the results of treatment in 12 patients with intracranial HPC at our unit between 1978 and 1999 and review the literature.

## Patients and Methods

Tissue registry of the Neuropathology Unit of our Hospital was reviewed for the diagnosis of malignant, atypical, angioblastic or vascular meningioma, as well as HPC involving the central nervous system. The records of 15 patients with the unequivocal diagnosis of HPC who were treated at the Department of Neurosurgery were reviewed. Three pure orbital HPC were excluded, thus leaving 12 intracranial HPC for the analysis.

In addition to the clinical and imaging studies (Computed Tomography Scans (CT), Magnetic Resonance Imaging (MRI) and angiograms), we reviewed the original operative records to ascertain tumour macroscopic features and the extent of tumour removal, which was classified according to Simpson's scale for the removal of meningiomas [49].

Relevant data throughout the postoperative course such as the ocurrence of tumour recurrence or extracranial metastasis and the need for radiation therapy or reoperation were consigned.

Six patients had plain skull films. Basal and postcontrast CT scans

Table 1. *(continued)*

| *Case n°* | *Operative findings* | *Simpson Grade* | *Outcome (6 mths)* | *Radiotherapy (dose)* | *Follow-up (mths)* | *Time (mths) and treatment at recurrence* | *Outcome at last date* |
|---|---|---|---|---|---|---|---|
| 1 | moderate bleeding | I | GR | recurrence (60 Gy) | 144 | 120 (surgery + RT) | death disease free (1994: SD) |
| 2 | reddish nodul no dural attachment | I | GR | no | 81 | – | death |
| 3 | moderate bleeding | IV | GR | no | 6 | – | lost |
| 4 | dural & bone infiltration bleeding | I | GR | recurrence (60 Gy) | 78 | 60 (holocraneal RT) | GR |
| 5 | bleeding | 1 : IV<br>2 : IV<br>3 : IV<br>4 : IV<br>5 : IV | GR | initial (64 Gy) tumor & margins | 76 | 24–15–14–14 (embolization + surgery) | death |
| 6 | bleeding | I | GR | no | 48 | – | GR |
| 7 | reddish bleeding | I | GR | no | 17 | – | GR |
| 8 | bleeding | II | GR | initial (60 Gy) | 8 | – | GR |
| 9 | bleeding | 1 : I<br>2 : I<br>3 : I<br>4 : II | GR | last recurrence (56 Gy) | 168 | 67–48–31 (surgery (last with surgery + RT)) | vision defect ophthalmoplegia GR |
| 10 | bone infiltration bleeding | I | GR | initial (60 Gy) | 6 | – | mild dysphasia GR |
| 11 | hemosiderin | I | GR | initial (60 Gy) | 78 | – | GR |
| 12 | bleeding | II | GR | initial (60 Gy) | 6 | – | GR |

*F* Frontal; *P* Parietal; *CPA* cerebellopontine angle; *Obst SLS* obstruction of superior longitudinal sinus; *Iso T1 T2* isointense in T1 & T2; *GOS* Glasgow Outcome Scale; *GR* good recovery; *SD* severe disability; *RT* radiotherapy. Oedema: −: No, +: mild, ++: moderate.

were available in all patients and seven of them also had MR examinations (T1-weighted, T2-weighted and contrast-enhanced T1-weighted studies). Only three patients had preoperative cerebral angiography.

A total of 20 operations were performed for resection of either a primary tumour or recurrences. Histological examinations included both conventional (HE, reticulin) and immunohistochemical techniques such as antibodies against vimentin, epitelial membrane antigen (EMA) and endothelial markers (CD 34).

Eight patients received conventional external radiation therapy at different moments through their courses, and up to date stereotactic radiotherapy has been not administered.

Recurrence in patients previously operated upon, was defined as the presence of tumour in contrast imaging studies (CT or MRI) producing or not symptoms or signs as well as regrowth of tumours not totally resected.

One patient was lost to follow-up. The median follow up for the remaining 11 patients is 64.5 months (range 6–168). The final outcome was classified according to the Glasgow Outcome Scale [27].

## Results

Table 1 reflects clinical and radiological data as well as the operative findings, additional treatment and follow-up.

### *Clinical Data*

There were 10 women (83.3%) and 2 men (16.7%). The age at diagnosis ranged from 12 to 72 years (mean of 44.9 yrs). Ages were uniformly distributed through decades.

### *Tumour Location*

Ten tumours were supratentorial and most located at the frontoparietal parasagittal region (Fig. 1). The falx was frequently involved and the superior longitudinal sinus was invaded in two cases. One tumour located in the tentorium extended to both supra and infratentorial compartments (Fig. 4). Two tumours had orbital extension (Fig. 2). Only one tumour was entirely located in the posterior fossa at the level of the cerebellopontine angle. In one case the lesion was purely intraparenchymal. None of the HPCs grew along the base of the skull.

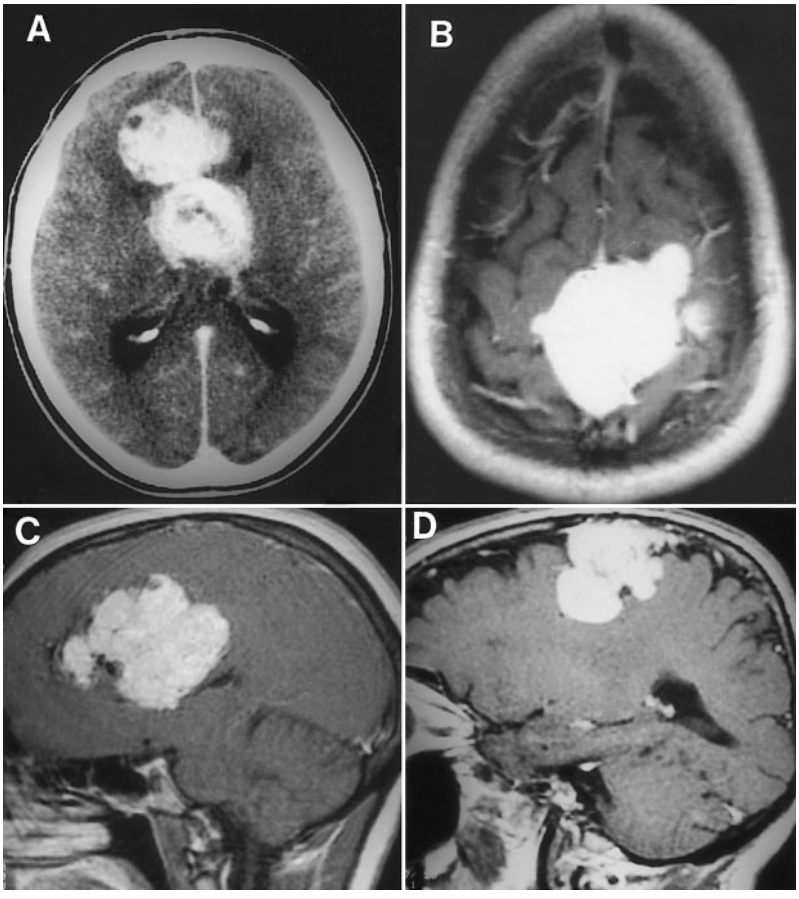


Fig. 1. Left side (Case 7). Preoperative axial contrast-enhanced CT scan (A) and sagittal contrast-enhanced MR T1 weighted (C) images showing a midline mass with irregular borders and heterogeneous enhancement. The prominent peripheral signal voids seen in C reveal the hypervascular nature of the lesion. Right side (Case 8). Preoperative axial (B) and sagittal (D) contrast-enhanced MR T1 weighted images showing a large polylobulated bilateral falcine mass with broad based dural attachment and homogeneous enhancement. A dural tail sign is seen along the falx

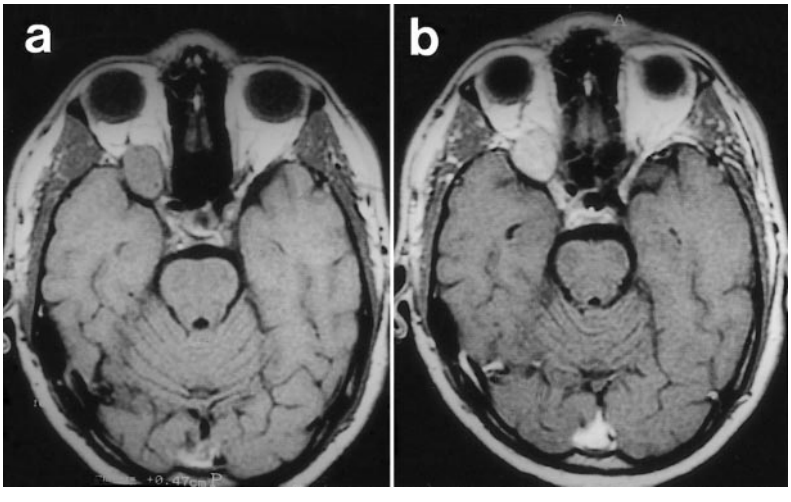


Fig. 2. (Case 9). Axial plain (a) and postcontrast (b) enhanced MR T1 weighted images reveal a densely enhancing, well demarcated mass producing destruction of the lateral bone margin of the orbit. The lesion extends both into the middle cranial fossa and into the right orbit

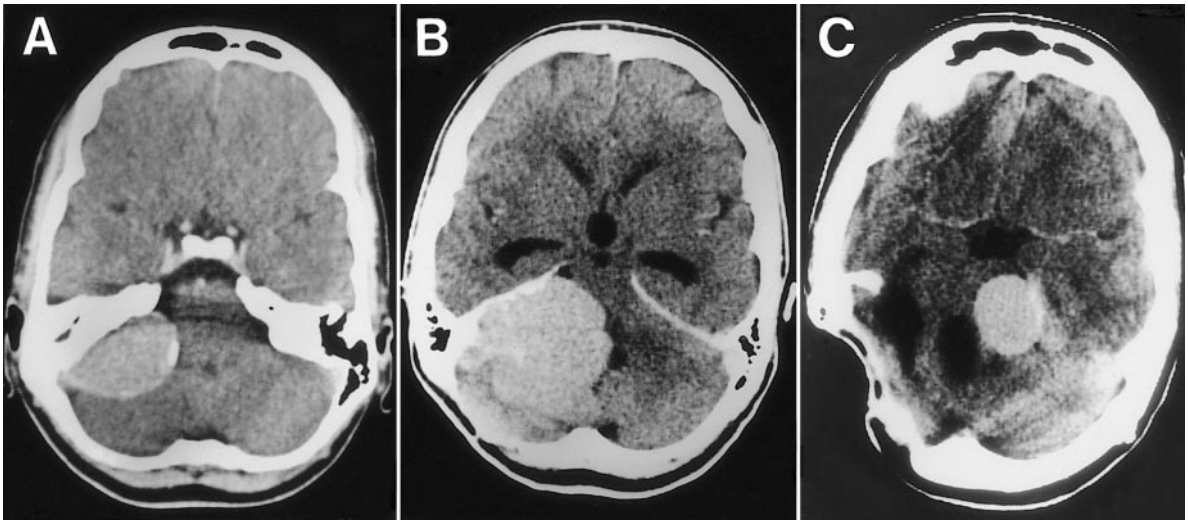


Fig. 3. (Case 5). Contrast enhanced CT scans performed before initial surgery (A), and following the third (B) and the fourth (C) recurrences. The tumour initially located at the right cerebellopontine angle, showed dense homogeneous enhancement and linear, peripheral calcification. At the third recurrence it appears as a huge extra-axial posterior fossa mass with a broad based dural attachment causing secondary hydrocephalus. Notice the contralateral location at the tentorial marging with the fourth recurrence

*Clinical Presentation*

Symptomatology was related to tumour location. In contrat to other series, intracranial hypertension and seizures were not common, as only one patient had headache and another two seizures at the time of diagnosis. The most common manner of presentation was a focal motor deficit in parasagittal or falcine tumours and a visual deficit in patients with an orbital component (Fig. 2). The patient with the posterior fossa HPC presented with hearing lost as well as facial and hypoglossal paralysis. The average duration of symptoms and signs from the onset to the time of first surgery was 3.1 months (range 1 week–1 year)

*Imaging Studies*

All tumours were hyperdense in the basal CT scans and 10 enhanced homogeneously following intravenous contrast injection (Fig. 1 and 3). In six cases (50%) tumour margins were irregular or lobulated (Fig. 1 and 4), but in the remaining cases the margins were regular (Fig. 2). In only one tumour a small peripheral lined calcification was observed (Fig. 3). Three of the HPCs showed bone erosion but none of the tumours in this series produced hyperostosis.

In the MR studies 6 tumours were mainly isointense with the cortical gray matter on T1-weighted and T2-weighted images (Fig. 4), and one (case 11) was heterogeneous, suggesting tumoural bleeding, with mixed signal on both sequences. Seven tumours were imaged with contrast enhanced T1 weighted imaging and all tumours enhanced. The enhancement was heterogeneous in 2 and homogeneous in 5. The "dural tail" sign was observed in two cases. In 3 cases a cystic component was observed (Fig. 4). Two of the 7 tumours studied with MR had prominent internal or peripheral serpentine signal voids suggesting vessels (Fig. 1).

The three tumours studied by cerebral angiography showed dense tumour stain with slow circulation and retarded venous drainage.

Preoperative imaging diagnosis was meningioma except in all but one of the cases (case 2) in whom the absence of dural attachment suggested a glioma or metastatic tumour.

*Surgery*

Twenty operations were performed in the 12 patients (8 re-operations for recurrence). Eleven patients were initially operated on at our Hospital. Tumours were generally reddish and rubbery in consistency. In 10 patients (83.3%) tumour resection caused brisk bleeding, and prominent dilated vessels were seen, both on the outer aspect and on cutting the tumour. In one patient (case 2) the lesion had no apparent dural

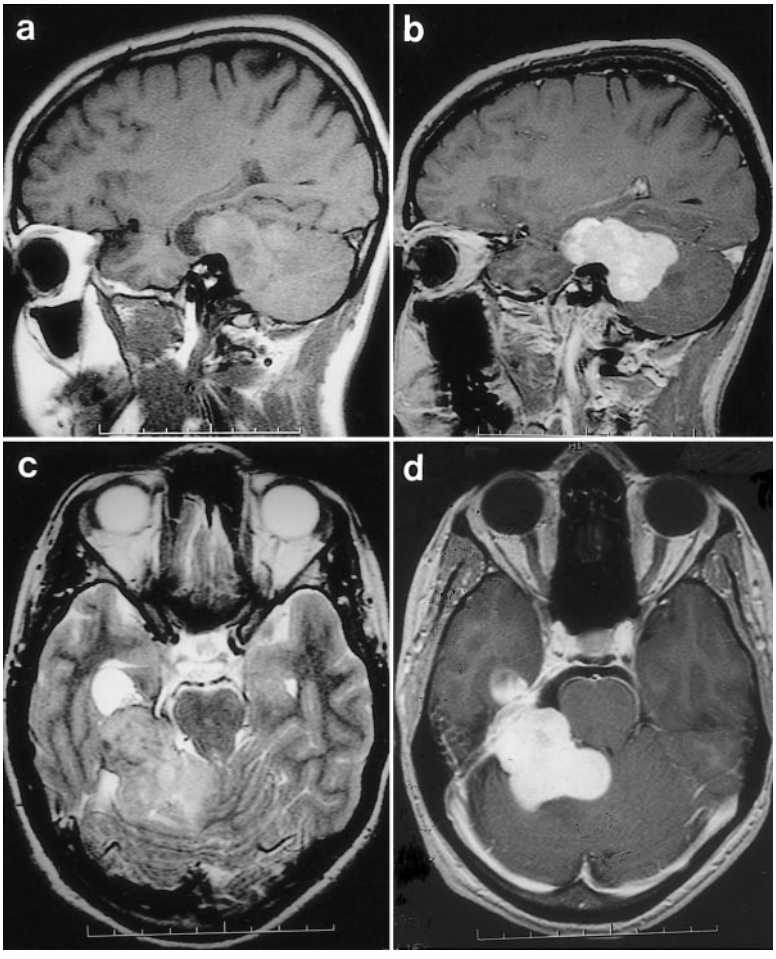


Fig. 4. (Case 12). Sagittal MR T1 weighted (a) and axial T2 weighted (c) images showing a poly-lobulated mass involving the middle and posterior fossae. The lesion is iso-intense with the cortex on both T1 and T2 sequences excepting for a small anteriorly located cystic component which is iso-intense with the cerebrospinal fluid. (b and d) Postcontrast sagittal and axial T1 images showing the dense contrast enhancement of the tumour

attachment and was considered as purely intracerebral.

In ten cases radical excision could be achieved (Simpson grades I and II) but two patients had Simpson IV resections.

There was not operative mortality and all patients improved clinically after the first operation, with a good recovery on the Glasgow Outcome Scale (GOS) at 6 months in every case.

### *Histological Features*

Microscopically, all tumours were very cellular. Nuclei were generally round to fusiform in shape, showing neither coarse chromatin nor nucleolar prominence. Cytoplasms were scarce and poorly defined. Vascularity was abundant, with thin-walled vascular networks showing a staghorn-like arrangement and lining with flat endothelial cells (Fig. 5a). Reticulin stain typically showed a pattern of fine fibers surrounding individual or small groups of tumour cells (Fig. 5b).

Occasionally, multinuclear cells, giant cells, and monstrous nuclei were observed (case 7). The mitotic index was variable, with more than 10 per 10 high power fields in case 8, and less than 5 per 10 high power fields in the remaining cases.

Immunohistochemical findings were similar in all cases. All tumours were positive for vimentin and negative for epithelial membrane antigen. Only in three cases tumour cells were focally positive for CD34. Endothelial cells were always positive for CD34 (Fig. 5c).

### *Recurrences and Metastases*

Four patients developed at least one local recurrence after initial surgery at an average interval of 68 months

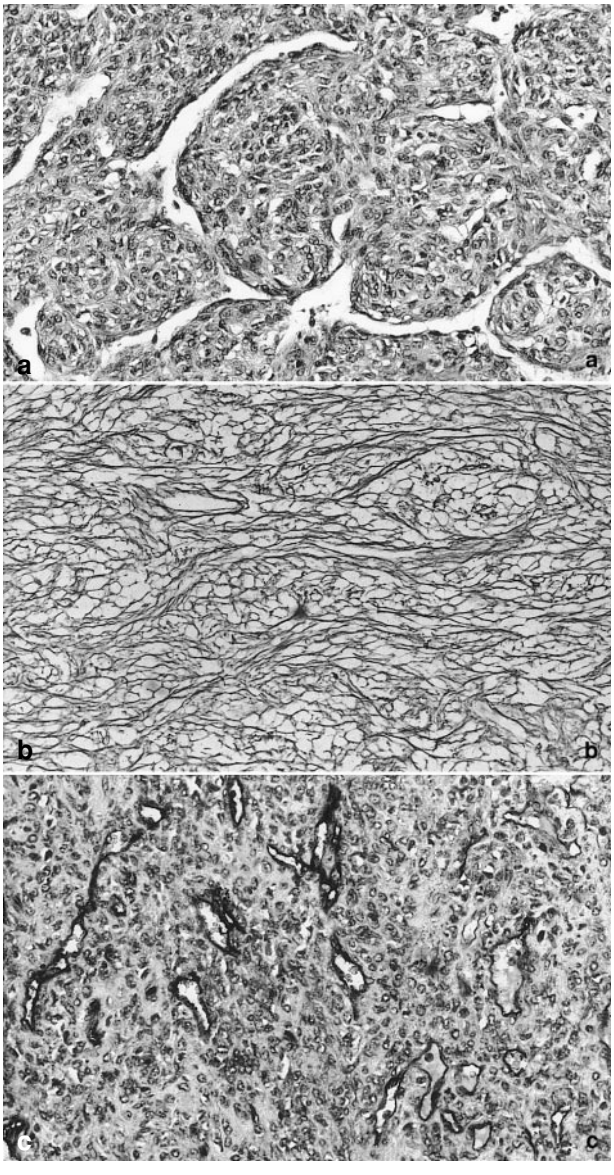


Fig. 5. (a) Photomicrograph showing staghorn-type vessels lined by thin endothelia and surrounded by hypercellular tumour (H & E × 200). (b) Photomicrograph showing individual tumour cells surrounded by a dense reticulin network (Wilder's reticulin stain × 200). (c) Photomicrograph showing immunostaining of endothelial cells for CD34 (CD34 × 200)

(range 24–120 mths), with a total recurrence rate of 33.3%. Two patients had one tumour recurrence; one had three, and another had four recurrences (Fig. 3). In cases with more than one recurrence, tumours tended to recur at shorter intervals.

It is noteworthy that we did not observed shift in the histological pattern or tendency toward increasing anaplasia when comparing recurrent to original tumour. Moreover, the mitotic index remained the same, or even diminished (case 5)

None of our patients have developed extracranial metastases up to now.

*Radiotherapy*

Irradiation following the initial operation was either not considered (three cases) or rejected by the patient (two cases). However, during the last years radiotherapy has been strongly recommended independently of the completeness of resection.

Eight patients were treated with external radiotherapy at some time during the course of their disease. Five patients (cases 5, 8, 10, 11 and 12) received doses ranging from 60 to 64 Gy after initial tumour removal. Of these patients four have lived 6, 15, 16 and 85 months without recurrence and the other one (case 5), who had only subtotal resection (Simpson grade IV), died 76 months after surgery.

Three patients (cases 1, 4 and 9) were irradiated at recurrence; two of them (cases 1 and 9) after total resection of recurrent tumour and the other (case 4) received only radiotherapy for recurrence. Two patients (cases 4 and 9) are still alive (GOS 1) and free of recurrence after 78 and 168 months at follow up and the other one (case 1) died 24 months after the treatment.

*Survival*

Eight out of the 12 patients in this series are disease-free after a mean follow up of 52 months (range 6 to 168). One patient was lost during the follow up and three died, two because of tumour relapse (cases 1 and 5) and one due to an unrelated disease (case 2).

## Discussion

The histological origin of central nervous system HPC has been a matter of controversy for a long time. It is now widely accepted that this tumour arises from meningeal capillary pericytes and the current WHO classification includes HPC in the group of meningeal, mesenchymal non-meningothelial tumours with uncertain malignant potential or borderline malignancy [29, 30].

In any case, it seems clear that HPCs behave in a different way than meningiomas. They have a relentless tendency to local recurrence and even when local control can be achieved, the risk of distant metastases

remains a threat as long as the patient lives. Thus, it is very important to identify the true nature of the tumour from the beginning, in order to make an appropriate prognosis and give correct treatment.

### *Incidence and Clinical Presentation*

Meningeal HPCs represent 1.8% of all meningiomas operated on over the last 20 years in our Unit.

Though in other series 50 to 70% of the patients with HPC were male [6, 20, 21, 26, 61], in contrast to the sex distribution seen with meningiomas, 83.3% of our patients were female.

The average age of presentation in patients with HPCs ranges from 38 to 42 years in different series, which is lower than with meningiomas (peak at the early fifties) [6, 20, 21, 26, 38, 61]. In our patients the mean age of presentation was 44.9 years.

Likewise, the interval between initial symptoms and diagnosis is shorter with HPCs as compared to meningiomas, which are symptomatic for 1–2 years on average. The median interval between initial symptoms and diagnosis in our 12 patients was 3.1 months, which is shorter than in other series [6, 20, 21, 26].

Unlike other series, in which intracranial hypertension was the most common form of presentation, most of our patients firstly developed motor or sensory deficits and only one presented with seizures. Intracerebral haemorrhage and other less common ways of presentation have been documented in the literature [17, 33, 39, 50].

Intracranial location of HPCs is similar to that of meningiomas and about 15% occur in the posterior fossa [41, 61]. Most HPCs have dural attachments, but there are reports of tumours in the pineal, sellar and suprasellar regions and the third ventricle [1, 2, 44, 51]. Exceptionally they are purely intraparenchymal [35, 42], as ocurred in one of our cases.

### *Imaging*

In general terms HPCs are indistinguishable from meningiomas in imaging studies [8, 10, 45, 56, 60].

On the CT scan HPCs usually show a broad-based dural attachment although some authors have suggested that a narrow-based attachment favours the diagnosis of HPC rather than classical meningioma [8]. HPCs tend to show with a higher frequency than meningiomas several features which have been associated with an aggressive behaviour such as apparent parenchymal invasion (mushrooming), irregular or polylobulated borders, bone erosion and heterogeneous contrast enhancement [8, 9, 22, 40, 48]. It has been noted that unlike meningiomas HPCs do not have tumour calcifications [21, 22, 26, 40]. However, one of our patients had intratumoural calcifications. Brain oedema, if present, is usually mild to moderate as ocurred in our patients [48], the incidence of severe brain oedema in our series of intracranial meningiomas being of the order of 21.5% [32].

Findings on MR are also similar to that of meningiomas [8, 10, 60]. HPCs are iso-intense with cortical gray matter on T1 and T2-weighted images. However they show heterogeneous enhancement more often than meningiomas. The "dural tail" sign can be present and the presence of associated brain oedema is also mild to moderate [8, 10].

After analyzing 18 cases of HPC studied by cerebral angiography, Marc *et al.* considered characteristic the following findings: dural arterial supply from branches of both the internal and external carotid arteries; few main feeders (1–3) from which a myriad of small corkscrew vessels arise; dense tumour stain, slow circulation and retarded venous drainage [34]. In contrast to this opinion, Guthrie *et al.* considered that only one of 20 angiographic studies in their patients was diagnostic of meningeal HPC [21, 22].

In our series three patients were studied by angiography and in all of them the tumour typically showed a dense blush and delayed venous drainage.

Di Chiro *et al.* (1987) found that metabolically active meningiomas, including angioblastic ones may be differentiated by hypermetabolic activity on positron emission tomography (PET) and that such "hot spots" on the PET study has an adverse prognostic significance [12]. However, these findings have not been replicated up to date.

A recent study has suggested that it is possible to distinguish between meningiomas and HPCs by using in vivo magnetic resonance spectroscopy (MRS) because of the higher levels of myo-inositol in the latter [3].

### *Histological Features*

Light microscopy, ultrastructural, and immunohistochemical data indicate that meningeal HPCs represent the intracranial counterpart of soft tissue HPCs [59]. The histological findings in the present series were similar to those previously reported. In keeping with

other authors we did not find a tendency to increasing anaplasia when recurrent lesions were compared with the original tumour and only a mild elevation in cell density was seen at recurrence [21].

The analysis of the correlation between histology and the final result yields contradictory results. Some authors have noted a relationship between some histological characteristics (including proliferationan indices) and the final prognosis, but others have found little or no correlation [16, 21, 31, 36, 57].

*Surgery*

Surgery is the treatment of choice for HPCs and because of their propensity to recur, resection must be as radical as possible [14, 55]. Unfortunately complete excision was possible in only 50–67% of the cases in different series [47]. In the present study radical resection was achieved in 83.3% of the cases.

The predominant feature of these tumours is their vascularity, which may result in substantial intraoperative blood loss. For this reason, some authors have recommended pre-operative embolization when HPC is suspected in order to reduce intra-operative bleeding [7, 19]. However, it should be noted that HPCs can parasitize leptomeningeal vasculature explaining that embolization of meningeal feeders may not be as effective in reducing surgical bleeding as it is in patients with meningiomas. Casasco *et al.* have reported devascularization of craniofacial HPCs by percutaneus puncture and intratumoural injections of N-butyl cyano-acrylate (NBCA) 24 to 48 hours before surgery [7].

Operative mortality for meningeal HPC ranged from 0 to 27% in different series and some deaths were attributable to exanguination [47]. Guthrie *et al.* reported no surgical deaths since 1974, and proposed the use of microsurgical techniques in order to decrease surgical complications [21]. In our series there was no operative mortality and morbidity was minimal.

*Survival and Recurrence*

It is difficult to compare the results between series of patients with HPC reported in the literature, because of the variability in the definition of recurrence and the extent of follow-up. In their series of 44 patients, Guthrie *et al.* found that median survival after the first operation was 60 months, with actuarial survival rates of 67%, 40%, and 23% at 5, 10, and 15 years respectively [21], which are comparable to the 65%, 45%, and 15% observed by Schroder *et al.* after reviewing 118 cases reported in the literature up to 1985 [47]. For comparison, it should be noted that in patients with meningiomas, Mirimannoff *et al.* reported a 5 and 10-year absolute survival rate of 83% and 77% respectively [38].

Eight of our 12 patients are still alive and "disease free" after a mean follow-up of 65 months.

As stated above, HPCs have a relentless tendency to recur. After defining recurrence as a progression of symptoms with radiographic or operative confirmation, Guthrie *et al.* found a mean recurrence free-interval of 47 months and calculated that the recurrence rate at 5, 10, and 15 years was 65%, 76%, and 87%, respectively [21]. Jääskeläinen *et al.* reported a mean interval to recurrence of 78 months in their 18 patients [26]. Four of our 12 patients had at least one local recurrence, with a median recurrence free-interval of 65 months. Furthermore, meningeal HPC tend to recur at shorter intervals after the first recurrence as occurred in our patients with multiple recurrences.

The small number of cases in our series does not allow one to stablish correlations between the final result and factors such as tumour location, degree of resection, histological appearance or the giving of radiotherapy. However, we think that radical resection followed by radiotherapy initially are related to a better prognosis.

To conclude, it seems clear that meningeal HPCs are more aggressive than typical meningiomas. In fact, recurrence rate is higher and when the patient lives for a long time it is extremely likely that the tumour will recur. In addition, HPCs may metastatize extracranially.

*Metastases*

Unlike other primary intracranial tumours, meningeal HPCs frequently metastasize outside the CNS, an event which significantly shortens the survival time. The most common sites in descending order of frequency are bone, lungs and liver [22]. Some authors have suggested that surgical manipulation may be associated with distant metastases by entering cells in the circulation through torn veins [28, 46].

Though early spread is definitely rare, the probability of developing metastases increases steadily with time, reaching 64% at 15 years [21]. The median interval to metastatic spread was 84 to 99 months in different studies with a range of 1–20 years. Awareness that

distant metastases can develop after many years of apparent tumour-free intervals is important for appropriate long-term management of these patients.

The absence of metastases in our series is probably related to the small number of patients and the time of observation.

### *Radiotherapy*

The impressive recurrence rate of HPC coupled with its potential for developing extracranial metastases make clear that surgery, even if radical, cannot be considered as the definitive treatment [14, 15, 18, 23].

When still classified as "highly vascular" or "hemangiopericytic" meningiomas some authors documented that pre-operative radiotherapy reduces vascularity and facilitate resectability [58].

The regression of peripheral HPC with irradiation is well documented [37] but postoperative radiotherapy has been more commonly used for meningeal HPC. Jääskeläinen *et al.* reported two patients with meningeal HPC treated postoperatively with 40 and 60 Gy reaching 167 and 263 months of disease-free survival [26]. Guthrie *et al.* convincingly demostrated the role of postoperative radiotherapy. Among the 44 cases included in their series, those receiving postoperative radiotherapy had significantly increased disease-free survival time (mean of 74 vs 29 months, $p < 0.05$) as well as a longer overall survival (92 vs 62 months). According to these authors there is a radiation dose-response relationship, without local recurrences among patients receiving $> 51$ Gy [21]. A similarly favourable response to radiation has been observed by others [18, 23, 37, 54].

Of the 10 patients who had radical surgery in the present series, none of the 4 receiving initial radiotherapy developed recurrences (during a mean follow up of 26 months). By contrast 3 (50%) of the 6 patients who did not receive radiotherapy developed at least one recurrence, and 2 patients died (89 months follow up).

Stereotactic radiosurgery has been used for recurrences in patients with high surgical risk [4]. Though most authors achieved initial tumour control in 99–100% of the cases, regrowth ocurred in 11 to 33% of the patients [41].

Despite the clear beneficial effects of radiation therapy, it should be noted that all recurrences after radiation develop within the treatment field, indicating that there is little to be gained from whole brain or spinal axis irradiation [15].

## Conclusions

Meningeal HPC is a rare tumour with biological behaviour totally different from meningioma. Though some clinicoradiological features may favour the diagnosis of HPC, pre-operative differentiation from meningioma is difficult because of the similarities in their radiological presentation. Radical surgery is the treatment of choice, but must be complemented by postoperative radiotherapy, which has proved to be the therapy most strongly related to the final prognosis. HPCs have a relentless tendency for local recurrence and radiotherapy can delay them. Metastases outside the CNS can appear even many years after diagnosis and adequate treatment of the primary tumour. The treatment of choice for recurrences is not clear, but it seems that recurrent tumour must be treated like the primary. Radiosurgery has proved to be effective in the management of recurrences when conventional radiotherapy was previously given. Survival rates after appropriate treatment range from 65 to 70% at 5 years and from 40 to 45% at 10 years.

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