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Case Report
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# Intracranial Masson Tumor: A Rare Tumor. Case Report and Literature Review

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## Abstract

The Masson tumor is a rare intracranial lesion and only 21 cases have been reported to date in the international literature. This pathological entity, called also intravascular papillary hyperplasia, must be considered when evaluating an intracranial mass, especially in dealing with young patients. We report the first case in a very elderly male patient where this lesion, presented with the appearance of a single intraparenchymal mass. The operation was curative. We present the clinical features of this entity and performed a literature review.

**Keywords: Masson tumor; IPEH; Brain tumor**

## Introduction

Masson Tumor or Intravascular Papillary Endothelial Hyperplasia (IPEH) is a rare, benign, vascular entity which can occur as an intracerebral lesion as well. It involves, most commonly, the skin and subcutaneous soft tissues but, it has also been reported in other locations like the nasal cavity, pharynx, larynx, internal auditory canal, labyrinth, heart valve, breast, digestive tract, liver, kidney, cervix, uterus, female urethra, and pelvic veins [1-3]. Its location in the Central Nervous System (CNS) is extremely rare and all the reported cases were in young patients. We report the first case of IPEH in an elderly male patient and review all the CNS cases published in the international literature.

## Case Presentation

A 84 year-old right-handed man, without any important past medical history, presented with a 1-month history of difficulty in walking and two episodes of partial motor seizures involving the left arm. Neurological examination revealed a left side mild brachio-crural hemiparesis (4/5 MRC). A CT brain scan showed a round, slightly hyper dense parietal intracerebral lesion measuring approximately 2.5 cm in diameter with surrounding edema (Figure 1). MR imaging confirmed the presence of a heterogeneous, lobulated, partially hemorrhagic lesion with a slight enhancement (Figure 2). Our working diagnosis was consistent for secondary lesion although the enhancing pattern and the amount of edema were not typical of such type of lesion. Furthermore, a PET and all the tumor markers were negative. After an extensive discussion with the patient and his family it was decided to remove the lesion surgically. *Via* a right parietal craniotomy the tumor was entirely resected "en bloc". Intra-operatively it was a well circumscribed, compact, red-brown lesion. Histopathological examination revealed vascular spaces of various sizes containing organized thrombus surrounded by numerous papillary structures (Figure 3A). The vascular spaces and papillary structures were lined with endothelial cells with immunoreactivity for CD 31 (Figure 3B) and CD 34. The endothelial cells showed no evidence of cytological atypia and no mitosis or necrosis

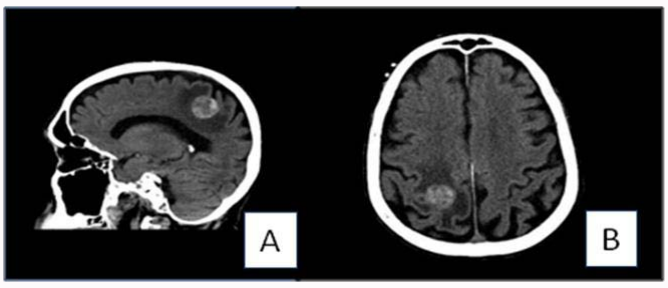


**Figure 1:** Sagittal (A) and axial (B) CT scan images showing a round subcortical parietal lesion.

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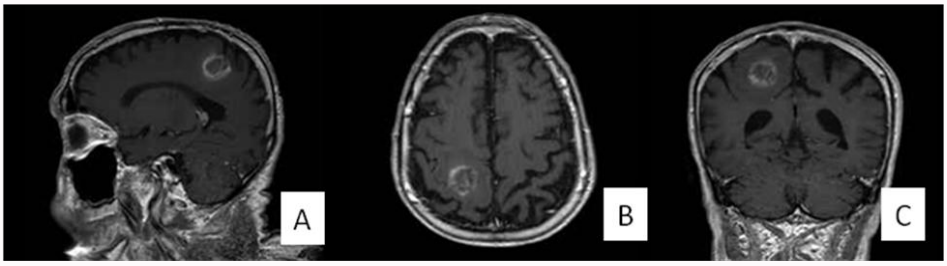


**Figure 2:** Sagittal (A), axial (B) and coronal (C) T1-weighted MR scan images confirming an heterogenous, partially hemorrhagic lesion with faint contrast enhancement.

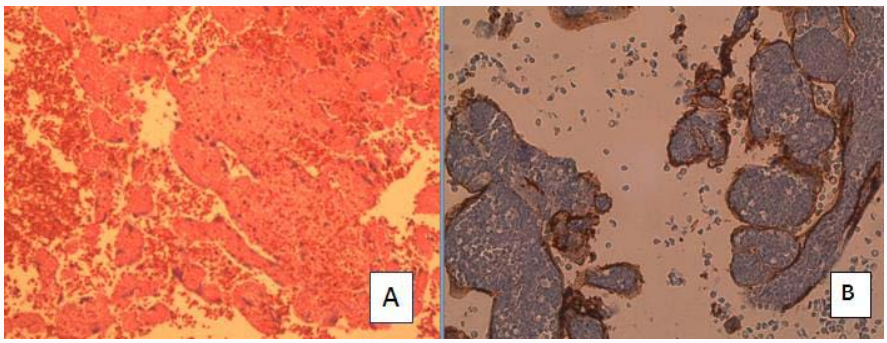


**Figure 3:** Photomicrograph of the lesion showing: (A) papillary projections and thrombotic center (HE stain, × 10) and (B) CD 31 immunoreactivity in the endothelial cell lining the papillary projections (peroxidase-antiperoxidase stain, × 20).

were seen (Figure 3C). The final diagnosis was IPEH. The patient's postoperative course was unremarkable and was discharged home a week later. At a 1 month follow up his hemiparesis had disappeared and at six months there was no evidence of recurrence tumor on a MRI.

## Discussion

The IPEH is a benign tumor and its pathogenesis is still debated. It is considered to be a proliferation of endothelial cells with secondary thrombosis and fibrin deposition or an excessive reaction following the normal reorganization process in a thrombus [1-11]. Only 21 intracranial cases have been previously reported in the literature (Table 1). From a review of these lesions we found heterogeneity with regard to patient age, gender, and location, prognosis, and treatment options. The age at presentation ranges from 2 days to 70 years and appears to be a female predominance (female/male ratio 18:4). Their location varied: 7 cases were intracerebral, 6 in the sellar or parasellar region, 7 in the subdural compartment and 1 within a venous sinus. In addition, in two cases, the lesions were multiple. The clinical symptoms usually reflect the local mass effect or cortical irritation. In some cases they may present as intracranial hemorrhage as well. The radiographic findings are non specific and in most of the cases there is a heterogeneous, minimal enhancement on CT or MRI. Often there is also evidence of hemorrhage. These features may mimic malignant tumors. Although these rare tumors may occur infrequently, perhaps, they should be kept in mind, in the differential diagnosis with other intracranial masses, especially if the radiological features are not so typical for a secondary lesion. The main treatment remains the complete surgical excision when possible or at least a debunking with a tissue diagnoses. Histological the lesion consists of a microscopically aggregated of endothelial cells (papillary projections) entangled by dense collage nous strands. The projections are associated with thrombotic material and are covered by a single layer of plump endothelial cells that lack anaplasia, pleomorphism or significant elevated mitotic activity. This benign appearance of the endothelial cells differentiates IPEH lesions from angiosarcomas [1,12-15]. Immunohistochemistry (positivity of CD31 and CD34) plays an essential role in establishing the vascular nature of IPEH. The natural history of intracranial IPEH remains unknown because of its rarity although it seems to have a tendency for a slow grow even in old patients. As matter of fact recurrences of IPEH after incomplete resection have been reported. In a review of the literature 57% (12 cases) had subtotal resection or biopsy and only 43% (9 cases) achieved a complete resection. Among this last group of patients no recurrences were reported. While all the cases with subtotal resection showed evidence of progression which required further treatment between 2 months and 9 years from the first operation or resulted in the patient's death (2 cases). Our case is the oldest one ever reported. The patient's age might have been a contraindication to surgery but his wishes, the good medical condition as well as the impossibility to find a primary tumor has pushed us to a surgical treatment. The prognosis for adults appears to be better than for young and neonates. The reported cases of incompletely resection of IPEH in infants exhibited a rapid progression. For symptomatic recurrent disease, further resection, stereotactic radiotherapy or chemotherapy, especially in young children, appear to be warranted although the effect of this last modality of treatment remains to be proven. Extended long-term follow up should be advocated in all patients, especially if young of age or with residual tumor.

## Conclusion

The intracranial location of IPEH is very rare and its presence in a very old patient can be considered extraordinary. If the lesion is completely resected, the prognosis can be very good.

**Table 1:** Summary of Clinical features of intracranial intravascular papillary endothelial hyperplasia.

| Author (Year) | Age, Sex | Clinical presentation | Neuroimaging findings | Location | Surgery | Radiotherapy and chemotherapy | Follow-up |
|---|---|---|---|---|---|---|---|
| Nagib et al., [9] | 16 yrs, F | Neurocutaneos disseminated form: epileptic seizures | CT: multiple intracranial enhancing supratentorial lesions. | Bilateral temporal and bilateral parietal regions. | Subtotal | None | Reoperation19 mos later |
| Chen and Kuo [4] | 3,5 mos, F | Increased ICP, seizures | CT: frontal, large enhanced lesion. | Left frontoparietal region. | Biopsy | None | Died 6 mos later |
| Izukawa et al., [7] | 55 yrs, F | Hemianopsia, sensory dysphasia, hemiparesis, seizures | CT: parietooccipital mass of mixed density, no enhancement. | Left parieto-occipital lesion. | Complete | None | No follow-up available |
| Sickler and Langford [12] | 12 days, F | Increased ICP | CT: mass with nonenhanced rim; MR: hemorrhagic mass. | Right middle cranial fossa extending into fronto-parietal region. | Subtotal | Doxorubicin hydrochloride, dacarbazine | Local recurrence at 2 mos treated by chemotherapy |
| Wen et al., [15] | 15 days, F | Increased ICP | MR: small enhanced process within the confluens sinuum. | Torcular herophili. | Subtotal | None | Neuroradiologically no progression for 6 mos |
| Patt et al., [10] | 27 yrs, F | Unilateral deficit of CNs III, V, and VI; headache | CT and MR: small enhanced lesion of the orbital fissure. | Left fissura orbitalis superior. | Complete | None | No evidence of recurrence for 6 mos |
| Tsuji et al., [13] | 18 yrs, F | Seizures, hemiparesis | CT and MR: intracerebral haemorrhage. | Left frontal lobe. | Complete | None | No evidence of recurrence for 2 yrs |
| Kristof et al., [6] | 70 yrs, F | Transient diplopia | MR: small enhanced sellar lesion. | Left cavernous sinus. | Subtotal | 46 Gy radiotheraphy | Enlargement of residual mass 3 mos treated by radiotheraphy |
| | 51 yrs, F | Diplopia | MR: small enhanced sellar mass. | Right cavernous sinus. | Subtotal | None | Small residual mass 3 mos later |
| | 24 yrs, F | Intermittent diplopia | CT and MR: small enhancing sellar mass. | Left sellar region. | Complete | None | Inconspicuous for 4 mos |
| Duong et al., [5] | 51 yrs, F | Headache, left visual field deficit | CT: multiple slightly hyperdense, enhanced masses with edema; MR: T1 hypointense, enhanced, T2 hyperintense masses surronded by heamosiderin rim and edema. | Bilateral occipital, right frontal and left parietal lobes. | Complete | None | No evidence of recurrence for 6 mos |
| Baylor et al., [2] | 27 yrs, F | Right facial nerve paresis | CT: no mass lesion; MR: enhanced, T2 hypo-to isointense mass. | Right internal auditory canal and falloppian segment of facial nerve. | Complete | None | No follow-up available |
| Avellino et al., [1] | 62 yrs, F | Ear pain, facial nerve paresis, dysphagia | CT: enhanced mass; MR: enhanced mass. | Left CPA and middle cranial fossa region. | Subtotal | After 1° operation 45 Gy radiotheraphy, after 2° operation radiosurgery | 2° operation for local recurrence at 9 yrs and radiosurgery |
| Lesley et al., [8] | 46 yrs, F | Right ear pain extending into right side of the face and neck, disphagia | MR imaging: T1 hypointense, enhanced, T2 hyperintense mass. | Right PICA aneurysm. | Complete | none | No evidence of recurrence for 1 yr |
| Stoffman and Kim [13] | 54 yrs, F | Headache, speech difficulty | MR: extradural partially hemorrhagic lesion in the left petrous apex and Meckel's cave. | Left Meckel's cave. | subtotal | none | Residual mass 10 mos postoperatively |
| Cagli et al., [3] | 16 yrs, F | Unilateral deficit of CNs III and IV | MR: small enhanced left intracavernous mass. | Left cavernous sinus. | Subtotal | None | Residual intracavernous mass at 3 yrs |
| | 18 yrs, F | Unilateral deficit of CN IV | MR: small enhanced right intracavernous mass. | Right cavernous sinus. | Subtotal | None | Residual intracavernous mass at 3 yrs |
| | 28 yrs, F | Unilateral deficit of CNs III, V and VI | MR: strongly enhanced left intracavernous mass. | Left cavernous sinus. | Complete | None | No recurrence at 2 yrs |
| | 24 yrs, F | Seizure | MR: enhanced left parietal mass. | Left parietal lobe. | Complete | None | No recurrence at 2 yrs |
| Zhang et al., [14] | 49 yrs, F | Left loss of hearing and facial palsy | MR: enhanced left petrous mass. | Left APC. | Subtotal | None | No follow-up available |
| Shih et al., [11] | 2 days | Proptosis | MR: supracellar, orbital and cerebellar. | Multiple supra and infratentorial. | Subtotal | None | Resection at 9 mos, died 6 mos later |
| Present case | 84 yrs, M | Hemiparesis, seizures | MR imaging: small enhancing parietal mass. | Right parietal-mass. | Total | None | No evidence of recurrence for 6 mos |

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