

Psammomatous Arteriovenous Malformation-Brain: A Rare Histological Presentation Leading to Diagnostic Dilemma

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Abstract

Occurrence of psammoma bodies (PB) in CNS is strongly associated with meningiomas. Its occurrence with arterio-venous malformation (AVM) is rare. We report a case of an unconscious 70 year old male who presented in the emergency department with mass lesion and on histopathology revealed a lesion composed of variable sized dilated and congested vascular channels which were mainly thin walled but with few thick walled vascular channels as well. Also noted were numerous lamellated calcified bodies (PB) which were present both intra-vascularly in small vascular channels as well as extra-vascular glial tissue. The minimal glial tissue included in the biopsy showed astrocytic proliferation showing mild anisonucleosis and hyperchromasia. Hence on light microscopy a differential diagnosis of Psammomatous cavernous haemangioma with a possibility of concurrent meningioma was suggested, as the synchronous presence of both the lesions is well documented in literature. A possibility of angiomatous variant of meningioma was also suggested. For confirmation of diagnosis, immunohistochemical studies were recommended which confirmed the lesion to be arising from vascular endothelial cells; so thereby conclusively ruling out meningotheliomatous neoplasm. Hence a final diagnosis of psammomatous arteriovenous malformation was rendered. Present case report is worth publishing as it not only documents the presence of PB in setting of AVM, but also highlights the importance of utilizing IHC tool to conclusively diagnose or rule out meningiomas in all such settings. It is important because angiomatous variant of meningioma with presence of PB can closely mimic AVM leading to a wrong diagnosis and further wrong management of the patient.

Index terms— av malformation, cns, psammoma bodies.

Psammomatous Arteriovenous Malformation-Brain: A Rare Histological Presentation Leading to Diagnostic Dilemma Sanjay Piplani¹, Rahul Mannan², Manas Madan³, Harjot Kaur⁴, Saumil Garg⁵ & Monika Lalit⁶
Abstract—Occurrence of psammoma bodies (PB) in CNS is strongly associated with meningiomas. Its occurrence with arterio-venous malformation (AVM) is rare. We report a case of an unconscious 70 year old male who presented in the emergency department with mass lesion and on histopathology revealed a lesion composed of variable sized dilated and congested vascular channels which were mainly thin walled but with few thick walled vascular channels as well. Also noted were numerous lamellated calcified bodies (PB) which were present both intra-vascularly in small vascular channels as well as extra-vascular glial tissue. The minimal glial tissue included in the biopsy showed astrocytic proliferation showing mild anisonucleosis and hyperchromasia.

Hence on light microscopy a differential diagnosis of Psammomatous cavernous haemangioma with a possibility of concurrent meningioma was suggested, as the synchronous presence of both the lesions is well documented in literature. A possibility of angiomatous variant of meningioma was also suggested. For confirmation of diagnosis, immunohistochemical studies were recommended which confirmed the lesion to be arising from vascular

endothelial cells; so thereby conclusively ruling out meningotheliomatous neoplasm. Hence a final diagnosis of psammomatous arteriovenous malformation was rendered. Present case report is worth publishing as it not only documents the presence of PB in setting of AVM, but also highlights the importance of utilizing IHC tool to conclusively diagnose or rule out meningiomas in all such settings. It is important because angiomatous variant of meningioma with presence of PB can closely mimic AVM leading to a wrong diagnosis and further wrong management of the patient. Keywords: av malformation, cns, psamomma bodies.

1 I. Introduction

sammoma body (from Greek word psammos meaning "sand") is a lamellated round collection of calcium. It is an example of dystrophic calcification seen mainly in various neoplastic and non associated with meningotheliomatous malignancies (meningiomas).

Author ? ? ? ? ¥ §: Sgrdimr, Amritsar, Punjab (India). e-mails:sanjaymikki@gmail.com, rahulmanan@gmail.com manasmadan@gmail.com, harjotbhandari@gmail.com monika.lalit@yahoo.com omatous calcification in angiomatous malformation (AVM) leading to a diagnostic dilemma.

2 II. Case Report

A 70 year old male presented in the emergency department of a tertiary care teaching hospital post trauma head after a road side accident. On admission, patient was unconscious. His Glasgow coma scale was 9. Patient was taken up for routine haematological, biochemical and serological investigations which were within normal limits. On ophthalmological examination, evidence of increased intracranial tension was suggested and patient was taken to the radiodiagnosis department for CT Head. Radiological opinion revealed a hyperdense lesion of size 7.2x3.4x5.7cm associated with hemorrhage seen in the region of right temporal lobe, right basal ganglia and corona radiata. Effacement of sylvian fissure with compression of right lateral ventricle and midline shift of 6 mm towards left side was noted.[Figure ??] To relieve the intracranial tension and to reach a conclusive diagnosis regarding the nature and etiology of mass lesion, craniotomy was done. The blood clots along with tissue fragments were evacuated and the specimen thus collected was sent to the pathology department for histopathological examination (HPE). The tissue was processed; 3-4 micron thick sections were cut and stained with haematoxylin and eosin stain. The sections processed showed variable sized dilated and congested vascular channels which were mainly thin walled but with few thick walled vascular channels as well. Also noted were numerous lamellated calcified bodies (psammoma bodies) which were present both intra-vascularly in small vascular channels as well as extra-vascular glial tissue.

3 C

We present an interesting case of CNS lesion in a 70 year old male presenting with increased psamm -neoplastic lesions. [1] Its occurrence in CNS is strongly The minimal glial tissue included in the biopsy showed astrocytic proliferation showing mild anisonucleosis and hyperchromasia. Slightly angulated nuclei were also noted. However; no obvious paranuclear vacu -oles or grooving was noted. Hence on light microscopy a differential diagnosis of Psammomatous cavernous haemangioma with a possibility of concurrent meningioma was suggested, as the synchronous presence of both the lesions is well documented in literature. A possibility of angiomatous variant of meningioma was also suggested.

For confirmation of diagnosis, immunohistochemical studies(IHC studies) were recommended which demonstrated immunoreactivity with CD31 and CD34 and immuno-negativity for EMA and CK-18. IHC therefore confirmed the lesion to be arising from vascular endothelial cells; so thereby conclusively ruling out meningotheliomatous neoplasm. The exuberant changes noted in the glial tissue were attributed to the phenomenon of reactive gliosis arising in setting of haemorrhagic diathesis of a vascular neoplasm post trauma. Hence a final diagnosis of psammomatous arteriovenous malformation was rendered.

4 III. Discussion

Psammoma bodies (PB) are lamellated basophilic structures which stain for mucin, calcium and iron.

Osteopontin produced by the macrophages is closely linked to their pathogenesis in the setting of dystrophic calcification. [2] Although conventionally presence of PB is harbinger of malignancy; for example, its presence in locations such as thyroid or cervical lymph node, almost always points towards a diagnosis of papillary carcinoma thyroid. Its presence is documented in various tumours such as papillary renal cell carcinoma, papillary serous cystadenocarcinoma, endometrial adenocarcinoma, mesotheliomas, somatostatinomas of pancreas and prolactinomas in pituitary gland. In central nervous system (CNS), its presence in mass lesions is often a red-herring for neuro-pathologist towards a diagnosis of meningioma as seen in present case report.

PB can however also be seen in certain nonneoplastic lesions such as endosalpingiosis of female genital tract [3] , psammomatous melanotic schwannoma and even melanocytic naevi.

Apart from meningiomas, in CNS; PB are associated with a rare benign vascular malformation arising in capillary telangiectasis-"Calcified telangiect -atic hamartoma" or "hemangioma calcificans." [4] Its association with (AVM) is rare as noted in present case report. The situation is confounded further by the fact that AVM have been found in association with a variety of intracranial neoplasms. The simultaneous occurrence is

thought to be coincidental but at many times it may also reflect common origin. [5] Various hypothesis regarding synchronicity have been postulated such as AVM induce tumour development and vise-a-versa. Also it has been proposed that certain humoral factors secreted by tumours can induce AVM as an acquired condition and that any focus of chronic irritation arising out of haemorrhagic diathesis on the arachnoid cells could cause a meningioma to develop. [6,7] Although CT report did point towards a diagnosis of AVM, the presence of atypical looking meningothelial cells (later attributed to reactive gliosis) did create a diagnostic dilemma in setting of large number of PB on biopsy.

It was important to rule out a possibility of meningioma as the management is different in the setting of intracranial AVM associated with meningioma. IHC is a useful tool in such cases. Meningiomas exhibit (at least focally) membranous as well as diffuse cytoplasmic immunolabelling for EMA, a feature foreign to vascular neoplasms of CNS such as hemangiopericytoma, nerve sheath tumours, solitary fibrous tumour and other fibroblastic tumours. Also on immunolabelling for cytokeratins, it is typically positive for CK-18 negative in the present case thereby comprehensively ruling out any possibility of meningioma. Also all the endothelial markers were positive on IHC. The optimal therapy for AVM has many options such as surgery, neuro-surgical treatment with radiosurgery or embolization or a combination of surgical and non surgical methods.

Present case report is worth publishing as it not only documents the presence of PB in setting of AVM, but also highlights the importance of utilizing IHC tool to conclusively diagnose or rule out meningiomas in all such settings. It is important because angiomatous variant of meningioma with presence of PB can closely mimic AVM leading to a wrong diagnosis and further wrong management of the patient.



Figure 1: Figure 1 :Figure 2 :

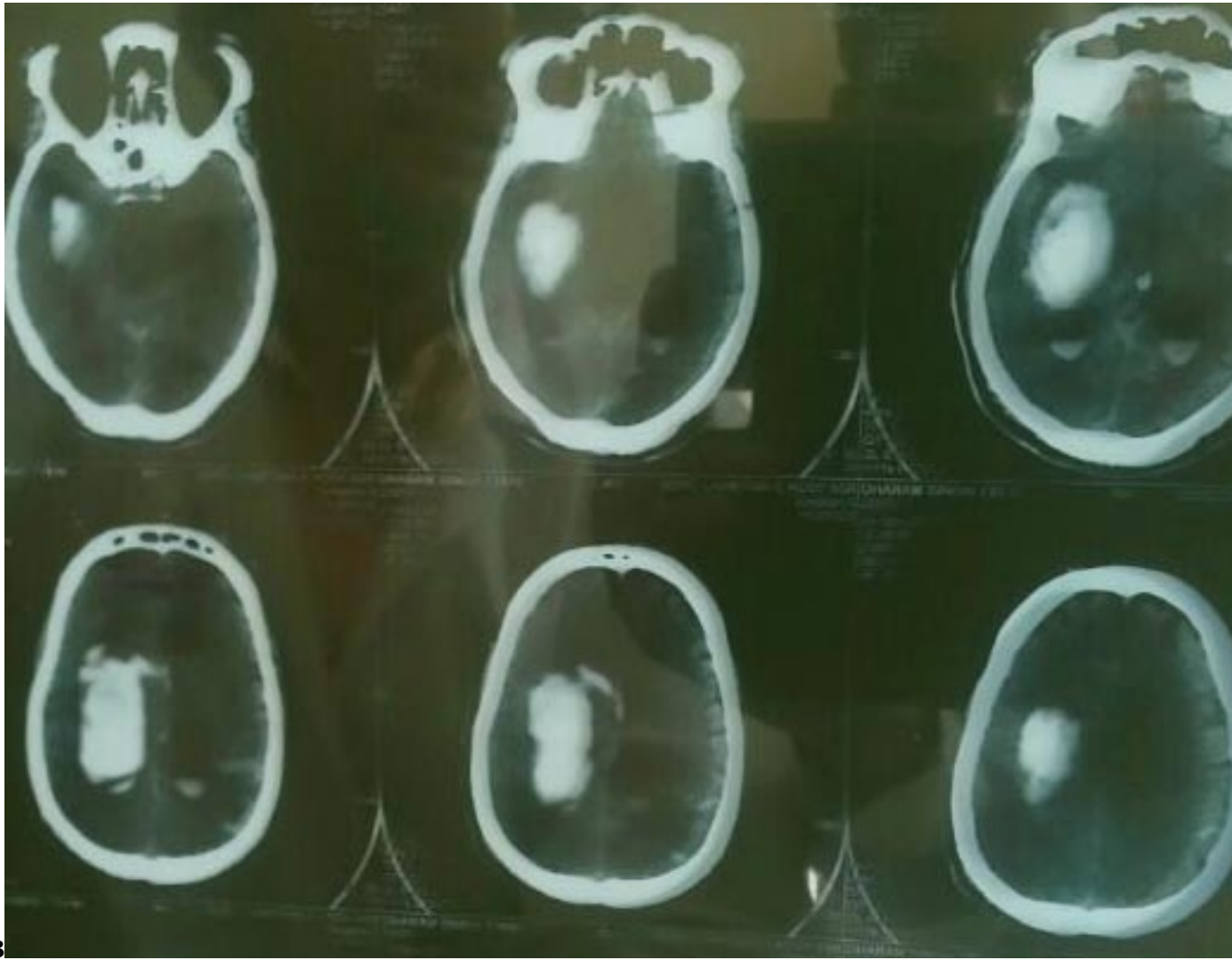


Figure 2: Figure 3 :

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