Case report

Mural nodules in multiple cavernous haemangioma

Shalini Rajandran Nair¹, Kartini Rahmat¹, Sharifah Majedah Idrus Alhabshi a, d, *, Norlisah Ramli¹, Mun Kein Seong b, Vicknes Waran c

¹ Department of Biomedical Imaging, University Malaya Research Imaging Centre, University of Malaya, Malaysia
b Department of Pathology, University of Malaya, Kuala Lumpur, Malaysia
c Department of Radiology, University Kebangsaan Malaysia, Malaysia

A R T I C L E  I N F O

Article history:
Received 16 June 2012
Received in revised form 30 August 2012
Accepted 9 September 2012
Available online xxx

Keywords:
Cavernous haemangioma
Mural nodule
Brain cyst
Cysticercosis
Imaging

1. Introduction

Mural nodules are round or plaque-like solid components arising from the wall of a cyst. These nodules may be hypervascular, neoplastic or infective. They do not always indicate a malignant aetiology, although they are commonly associated with tumours such as astrocytoma, meningioma, ependymoma, neuroblastoma and haemangioblastoma. Mural nodules have been noted in cavernous malformation as well. Cavernous malformations also known as cavernous angioma or cavernous haemangioma are rare vascular malformations characterised by hamartous dysplasias formed by abnormally dilated vascular channels with a single layer of endothelial lining and peripheral gliosis [1]. Absence of intervening normal neural tissue is a distinguishing feature. They represent 5–13% of all the vascular malformations [2]. They are commonly solitary with sporadic occurrence [3]. Interestingly, multiple lesions have been associated with familial form [3] with recent discovery of autosomal dominant inheritance pattern [2]. With the availability of high field magnetic resonance imaging (MRI), the detection and characterisations of cavernous malformation are becoming more common place. Cavernous malformation may vary in sizes. Repeated haemorrhages and cystic degeneration may give rise to larger lesions. To our knowledge, there were only two previously reported cases of giant cavernous haemangioma presenting with a mural nodule. We report a rare case of multiple cerebral cystic cavernous haemangiomas with mural nodules. Characteristic imaging findings and differential diagnosis are discussed.

2. Case report

Madam CPK is a 33-year-old female who had history of recurrent falls and abnormal gait at the age of 13 years. Imaging done at the time revealed presence of brain cysts. Following that, she also developed meningitis complicated with hydrocephalus and a ventriculo-peritoneal shunt was inserted. Over the next few years, she was asymptomatic until 4 years ago when she had recurrent unsteady gait. A new left perteiventricular cyst was noted and an Omaya reservoir was performed to assist in regular aspiration of the cyst.

The repeated aspiration gave temporary resolution of her symptoms until early this year. Subsequently, she was referred to our centre for further management. At presentation, she had difficulty in walking and weakness of the right side of the body along with progressive, painless right eye blurring of vision. Physical examination revealed power of 4/5 in both upper limbs. Ophthalmologic assessment showed poor visual acuity in the right eye (6/60) compared to the left eye (6/36). There was no retinal haemangioma.

* Corresponding author at: Department of Radiology, Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. Tel.: +603-91455555; fax: +603-9145 6682.
E-mail address: shmajedah@yahoo.com (S.M.I. Alhabshi).

0303-8467/– see front matter © 2012 Elsevier B.V. All rights reserved.
http://dx.doi.org/10.1016/j.clineuro.2012.09.014

Please cite this article in press as: Nair SR, et al. Mural nodules in multiple cavernous haemangioma. Clin Neurol Neurosurg (2012),
http://dx.doi.org/10.1016/j.clineuro.2012.09.014
The rest of the physical examination was unremarkable. The blood investigations were also normal.

Magnetic resonance imaging (MRI) of the brain showed multiple rim enhancing cysts in the pons and both cerebral hemispheres with the largest measuring 2.9 cm × 3.1 cm in the pons (Fig. 1). On the MRI scan, enhancing mural foci (Fig. 1) and blood-fluid levels (isointense on T1-weighted and hypointense on T2-weighted indicating haemorrhage) were noted within the cystic lesions. Extensive perilesional oedema was present but no areas of restricted diffusion were noted. In view of multiple cystic lesions with mural nodules, a diagnosis of haemangioblastoma with association to von Hippel–Lindau syndrome was suspected with differential diagnosis of cysticercosis and metastases. However, ultrasound and CT scan of the abdomen and pelvis were normal. There was no family history of similar disease.

Surgical decompression and image guided biopsy of the pontine cyst was done. However, histopathological examination revealed reactive gliosis with abundance of Rosenthal fibres. In view of her persistent walking difficulty, a repeat MRI done 6 months after the first surgery and showed an enlarging left frontal lobe cyst measuring 2.4 cm × 2.8 cm with a calcified coarse mural nodule and acute intracystic haemorrhage (Fig. 2). The large pontine cyst was now replaced by small clusters of cysts. Some of the other cysts with mural calcifications have also increased in size. There was also marked bilateral white matter oedema.

An unenhanced CT scan brain done for image guided surgery revealed calcified mural nodule in the left frontal cystic lesion and intraparenchymal calcifications. The patient underwent surgical excision of the left frontal lobe cyst. Intraoperatively, the cyst fluid was xanthochromatous and a 1.0 cm intracystic nidi was completely excised. The histopathological review showed numerous distended vascular channels formed by endothelial cells devoid of intervening neural tissue compatible with cavernous haemangioma (Fig. 3). Post surgery, patient was asymptomatic and is currently on neurosurgical follow-up.

3. Discussion

The aetiology of cysts with mural nodules can be broadly categorised into neoplastic, vascular malformations and infective. Cavernous malformations, although rare, should be considered in patients with features of progressive intracystic haemorrhage with cerebral calcifications. Patients can have a spectrum of neurological symptoms, most times they present with intractable epilepsy. According to Chahine et al., the lesions themselves do not form epileptic foci as they do not have neural tissues within [4]. Frequent intrarheostal haemorrhages especially in larger lesions resulting in ischaemia, inflammation and gliosis of the surrounding brain parenchyma which eventually forms epileptic nidi [4]. Interestingly, our patient did not develop seizure despite having intrarheostal microhaemorrhages with extensive perilesional oedema. Rather, she presented with weakness and difficulty in walking. Location of the lesions may be an influencing factor of the presenting symptoms as well. As the patient’s largest lesion was in the pons, we concluded that the inflammatory changes and gliosis could have involved the corticospinal fibre tracts resulting in her symptoms. Cavernous haemangioma has variable imaging appearances which include solid lesions, cystic lesions with mural nodules and heterogeneous lesions with both solid and cystic components [3]. Recurrent haemorrhage from the sinusoids of the malformation or from the neocapillaries of the cyst wall may result in cyst formation [5]. The haemorrhagic potential may be the result of presence of angiogenic activity from vascular endothelial growth factor which were detected in several immunohistochemistry studies [6,7]. A review of the literature showed that the cystic form is very rare with only 24 cases reported in the past [5]. Greater female and higher incidences in older patients have been documented in the cystic form [5].

These lesions are recognised to have a variety of sizes and growth rate due to repeated microhaemorrhages and/or recanalisation after intraluminal thrombosis [8]. Larger lesions
of more than 6 cm in diameter are defined as giant cavernous malformations [3]. To our knowledge, giant cystic cavernous haemangiomas with mural nodules are rare with only two cases reported in the existing literatures [3]. Our patient also showed a rare manifestation of cavernous haemangioma presenting with multiple cystic lesions associated with mural nodules and intratumoral haemorrhages. This posed a challenge in the radiological diagnosis as they have atypical features and mimicked other common pathologies.

In MRI scan, cavernous haemangiomas have a heterogeneous intensity from blood degradation products with a typical haemosiderin rim. In the cystic form, iso- to high-intensity cyst on T1-weighted images and a high intensity cyst on T2-weighted images are seen representing haemorrhage. Intramural nodules and cyst walls may enhance post gadolinium. The enhancement of the cysts wall is likely due to inflammatory reaction from recurrent haemorrhage. Calcification is seen in 20% of cases. A typical cavernous haemangioma usually has very little or no surrounding

---

oedema. However, giant cavernous haemangiomas or acute intralelional haemorrhage are noted to cause perilesional oedema and mass effect [3]. Gradient echo sequence is also very useful as it is sensitive in detecting haemorrhages and can be used in screening family members in suspected familial cases of cavernous malformation [2]. In our patient, the radiological diagnosis was difficult as apart from being a rare diagnosis, the imaging findings of mural nodules as well as extensive perilesional oedema resembled neoplasm.

Malignant transformation of a cavernous malformation has been reported by Schreuder et al, with several interesting hypothesis [6]. One of the theories suggested oligodendrogial cells proliferation in an existing cavernous malformation [6]. However, as cavernous malformations do not have neural tissue, its neoplastic potential is still debatable.

Primary neoplasms which have cystic components with a mural nodule and a tendency to haemorrhage are pilocytic astrocytoma, pleomorphic xanthoastrocytoma, glioblastoma multiforme and haemangioblastoma. On imaging, although some of the tumours have pathognomonic features, distinguishing them from one another may be challenging. Age of presentation and location of the lesions may assist in the diagnosis. Pilocytic astrocytoma occurs in childhood and posterior fossa is the most common location. It is rarely multiple. Pleomorphic xanthoastrocytoma is seen around the second decade. It usually occurs in the supratentorial region and multiplicity has not been documented.

Haemangioblastoma is a vascular tumour with an infratentorial preponderance and seen in adulthood. Relation to von Hippel–Lindau syndrome needs to be excluded in multiple lesions. This tumour has a high tendency to haemorrhage but almost never calcifies. Malignant tumour such as glioblastoma multiforme is seen in the older age group and tend to progress rapidly with a possibility of metastasis. They can cross the midline and occur mostly in the supratentorial region. This tumour rarely calcifies. Although cystic lesions with mural nodules were reportedly seen in meningioma, ependymoma and neuroblastoma it is not a usual feature as majority tend to be solid tumours. Our patient did not have the typical features of these tumours.

Cystic intracerebral lesions are also associated with metastasis from breast, liver, ovary and lung with tendency to haemorrhage. In younger children, multiple cystic lesions with calcified mural nodules are strongly associated with neuroblastoma metastasis. Since this was a long standing case, brain metastasis was excluded.

We feel that it is also worth discussing about neurocysticercosis as it is the most common parasitic infection in the world with similar imaging findings. Neurocysticercosis is seen in 60–90% of all cases of systemic cysticercosis. Radiographic features depend on the phase of the disease and MRI is the best imaging modality. Recognised features are enhancing mural nodules consisting of viable larval scolex. Extensive perilesional oedema may be seen in the inflammatory phase. Intracystic haemorrhage is an unusual presentation, differentiating it from a cavernous haemangioma.

The initial histopathology revealed presence of gliosis with Rosenthal fibres which may have been obtained from the periphery of the lesion. The subsequent histopathological examination further confirmed the presence of vascular channels of various sizes. No remarkable presence of intervening neural tissue was seen. The cystic fluid noted intraoperatively was noted to be xanthochromic indicating blood stained CSF-like fluid. The evolution of brain MRI findings in our patient correlated well with the histopathology reports. The treatment of intracranial cavernous haemangiomas may be medical or surgical which primarily depends on the progression of disease, size, location of lesions and presence of secondary complications. A detailed guideline and treatment algorithm has been developed to facilitate management of cavernous haemangiomas at the Northwestern Memorial Hospital [8].

4. Conclusion

Our case report highlights the importance of diagnostic imaging clues in differential diagnosis of multiple intracranial cysts. The presence of complicated intracystic haemorrhage, calcifications, disruption of blood brain barrier and evolving signal changes should alert the clinicians to the diagnosis of cystic cavernous malformations.

Conflict of interest

None.

Acknowledgement

The authors gratefully acknowledge the financial support from University Malaya Research Grant (RG390/11HTM).

References