Cerebral cavernoamas: radiological prevalence and clinical features in a 10 year retrospective study in a teaching hospital.

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Abstract

Objectives: Cerebral cavernous angiomas remain one of the most negotiable and controversial topics in neurological and neurosurgical practice. It can present with symptoms or can be found incidentally. The aim of this study is to evaluate the prevalence, clinical presentation, treatment options and outcome of patients with cerebral cavernoma.

Materials and Methods: Brain MRI for (14372) patients done in our university teaching hospital during 10 -year period from 2001 to 2011 were reviewed, 33 cases of cerebral cavernoma were found. The medical charts of these patients were reviewed and analyzed for their clinical presentation, treatment options and outcome.

Results: We identified 33 cases of cavernoma with a mean age of 33.3±16.8 years. There were18 females and 15 males. Four cases were excluded from analysis (but included in prevalence calculation) due to unavailability of clinical data. In the 29 other cases there were 35 cavernomas. The radiologic prevalence of cavernomas in this population was 0.23%. Most cavernomas were found in the supratentorial regions mostly found in the frontal lobes. The most common presenting symptoms were seizures in males and headache in females.

Conclusion: The radiologic prevalence of cerebral cavernomas was less than previous series, while clinical presentation was the same. Performing MRI at higher resolution and field strength with more sensitive sequences may lead to the detection of subtle or small brain abnormalities that would not have been detected previously.

Keywords: Cerebral Cavernoamas, Teaching Hospital.

Introduction

Intracranial vascular malformations are divided into four groups: capillary telangectasia, cavernous angioma (cavernoma), arteriovenous malformation and venous malformation. In

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large series, the mean age was reported to be about 37 years [1, 2]. Cavernomas are benign vascular hamartomous dysplasia characterized by abnormally dilated vascular channels lined by a single layer of endothelium surrounded by hemosiderin deposits and gliosis. Cavernous angioma occurs in 0.5% of the general population and compromise about 5-13% of all vascular malformations. There is no difference in the incidence between men and women. [1, 2]

Cavernous angiomas are divided into sporadic and familial forms. The frequency of detection of multiple lesions in familial cases is greater than that in sporadic ones [3]. Majority of multiple lesions were found in women [4]. Based on MRI images Zbramski et al have described four types of lesions associated with cavernous angioma. Type I lesions show hyperintense core on T1 weighted images and hyper or hypointense core on T2 weighted images. Type II lesions show reticulated mixed signal core on T1 and T2 weighted images with surrounding hypointense rim. Type III lesions are iso- or hypointense on T1-weighted images and surrounding hypointense rim on T2 weighted images. Type IV lesions are not seen or poorly visualized on both T1 and T2 weighted images [5].

Materials and Methods

Brain MRI for (14372) patients done during 10 year period from 2001 to 2011 were reviewed. Thirty three cases of cerebral cavernoma were found. The images were reviewed by experienced radiologists. The protocol for Brain MRI included sagittal T1, axial T1 and T2 and coronal FLAIR images, in some cases there was axial T2 gradient echo images and in 11 cases contrast media was given. The clinical data of these patients were reviewed and analyzed by an experienced neurologist regarding the clinical presentation, treatment options and outcome of these patients.

Results

We found thirty three cases of cavernoma; 18 females and 15 males. Four patients (all males) were excluded due to unavailability of clinical data, and in the remaining 29 cases there were 35 cavernomas. The mean age was 33.3 ±16.8 years, 18 (62%) were females and 11(38%), were males. Multiple cavernomas were seen in 3(27%) of males and 5(28%) of females. Of the 35 cavernomas 25 (71%) were supratentorial while 10(29%) were infratentorial (Table 1). The most common location was the frontal lobes 14/35 (40%). Temporal lobes were the second most common location 5 (15%) cases. Three cases (8%) were located in the deep grey matter nuclei and 2 cases (5%) were in the parietal lobes. None of the patients had cavernoma in the occipital lobes. Infratenotrially, 6 (17%) cases were found in the cerebellum while 4 (11%) cases involved the brain stem. Table 1 summarizes the location of the cavernomas. The clinical presentation (table 2) included headache, seizures or focal neurological deficits. The most common symptom was seizures in 11(38%) patient, 9 (31%) patients had headache and focal deficit was present in 14 (48%) patients. Four (14%) patients were asymptomatic (incidental cavernoma).

The most common symptom in males was seizures which was present in 7 (64%) patients, while in females the most common symptom was headache which affected 7(39%) patients. Seizures were present in 11 patients: 7 males and 4 females. Scalp electroencephalogram (EEG) was performed for 6 males and was

normal in all of them, however it showed focal abnormal activity in 3 female patients.

Table (1) location of cavernomas

	Supra-tentorial	Infra-tentorial	Frontal	Temporal	Parietal	Brainstem	Cerebellum	Deep Nuclei
Males	13	4	10 (7 right and 3 left)	1(left)	2(Right)	0	4 (1 right and 3 left	0
Females	12	6	4(1 right and 3 left	4(2 right and 2 left	0	5	2(1 right and 1 left	3(2 right and 1left)
Total No. of Patients	25	10	14(8 right and 6 left)	5(2 right and 3 left)	2(right)	5	6(2 right and 4 left)	3(2 right and 1left)

Table (2) clinical presentation

	Headache	Seizures	Focal deficit	Asymptomatic
Males	2	7	9	1
Females	7	4	5	3
Total No. of Patients	9	11	14	4

Headache was present in 7 female patients and 2 males while focal neurological deficits were found in 9 females and 5 males and ranged from numbness to quadriparesis. Five (17%) patient presented with hemorrhage; three of them were frontal hematoma and the other two with brainstem hemorrhage. The two cases of brain stem hemorrhage were females one of them was associated with hydrocephalus and both cases ended with poor outcome. Of the three cases of frontal hematoma one was found to have internal carotid artery (ICA) aneurysm on the same side of hematoma on conventional angiography. Eight patients (5 males and 3 females) were treated surgically while 15 patients (5 males and 10 females) were not candidate for or refused surgical treatment and were treated medically. Endovascular treatment was used in one case while one patient had ventriculoperitoneal (VP) shunt insertion. Medical treatment was offered from the beginning to one patient while 3 patients didn't receive any kind of treatment and were just followed up. The outcome was good in 13 patients (12 females and one male), stable in 10 (3 females and 7 males) and poor in two female patients. Four patients had variable non-resolving symptoms: quadriparesis, developmental delay, and obsessive compulsive disorder and hand tremors.

Imaging: Type II was the most common, present in 17 cases, 10 cases were of type I. Two cases were of type III and none of cases were type IV. (Figures 1and 2)
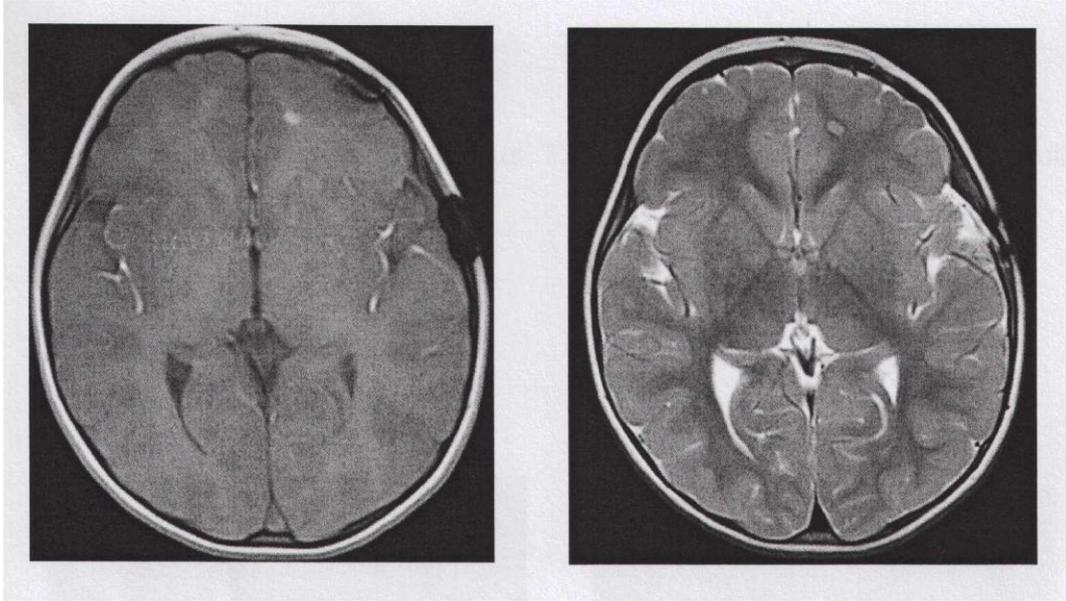
Figure (1) (Type I Cavernoma): Axial T1 weighted (left) and T2 weighted (right) show hyperintense lesion on both sequences in left frontal lobe
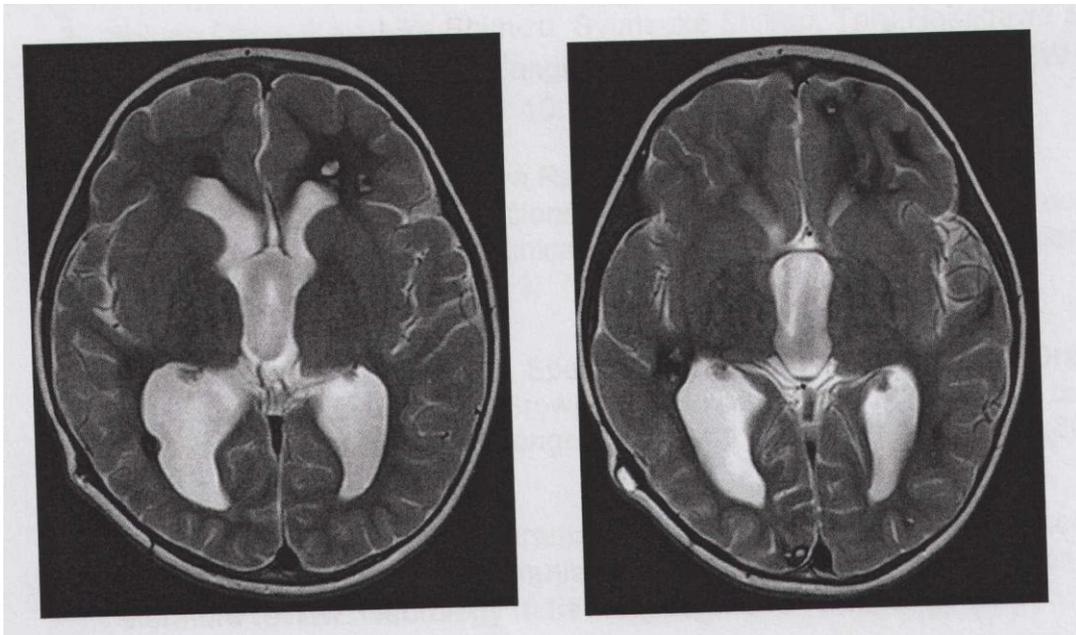

Figure (2) (Tupe II cavernoma): Axial T2 two consecutive images show multiple reticulated lesions

Discussion

The etiology of cavernous angioma is unknown .Cranial radiation, immunosuppressive treatment, genetic and hormonal factors have been implicated for cavernous angioma [6] .Clinical behavior vary from asymptomatic to sudden death related to intracerebral hemorrhage. No correlation was found between size of cavernous angioma and initial presentation[4] .Risk factors for aggressive clinical behavior of cavernous angioma include familial or multiple forms, previous brain radiotherapy, incomplete removal, associated venous malformation and previous hemorrhage. Cavernous angioma radiological appearances is regarded as pathognomenic but some pathologies can mimic cavernous angioma, most common are hemorrhagic neoplasm especially metastatic cancer while others include meningioma, gliomas, other vascular malformation and cysticercosis[7]. Cavernous angioma are most commonly found in supratentorial region (80%) and are preferentially located cortically at subcortical white matter, a deep location in basal ganglia, hypothalamus or ventricular system is infrequent[4]. Cavernous angioma size vary from few millimeters up to giant cavernous angioma(up to 11cm) which is due to recurrent hemorrhages, clot organization, pseudocapsule formation and secondary expansion. The mechanism behind this is the osmotic effect of the blood breakdown products [8]; Cavernous angioma reaches larger size in children more than in adults [4]. Cavernous angioma related intracerebral hemorrhage occurs more frequently in female patients especially during pregnancy. Those located in third ventricle and pontomesencephalon [7] are also at higher risk of bleeding. Calcification is believed to be associated with hyalinization of sinusoidal walls of the cavernous angioma [8] .T2 Gradient echo and post contrast images aid in increasing the sensitivity to detect cavernoma and determining if there are single or multiple lesions. Lack of contrast enhancement may be due to slow perfusion at cavernous angioma but some authors advocate the use of high dose and time-delayed contrast MRI studies [8]. Higher MRI field strengths with increased resolution, and the use of new MRI sequences that are more sensitive to subtle structural changes, will probably increase the number of small brain abnormalities detected[9]. Prior to MRI cavernous angioma were thought to be rare. MRI is the most sensitive modality to detect cavernous angioma which is considered now the most common identified vascular brain malformation[4] and T2 weighted gradient –echo images (T2*GRE) is considered the gold standard MRI sequence[10]. Inclusion of a FLAIR sequence is recommended because it is very sensitive for the detection of white matter lesions or vascular abnormalities; also 3-T MRI is more susceptible to subtle findings [11]. However, for clinically symptomatic cavernous angioma conventional MRI (T1 and T2 weighted images) has specificity and sensitivity near 100% [10]. The sensitivity may be lower when scans are read by professionals who are not medically qualified as neuroradiologists [9]. Limitations of our study include retrospective analysis, relatively small number of patients , not using high field MRI nor susceptibility weighted MRI which can better delineate cavernous angioma and further reveal additional lesion nearly twice as many lesion compared to T2* GRE.

Conclusion

We described our experience over 10 years of

cavernomas in a retrospective manner. Our findings did not differ from clinical aspects as compared to previous series [1, 2, 4], however, the radiological prevalence of cavernoma in our series was lower (0.23%) than what was reported in two large retrospective series of 0.5% [2] and the 0.4% in general population [9] respectively. This may be due to our small size of the sample, one site study, less sensitive techniques or a reflection of genetic variation. Thus, performing MRI at higher resolution and field strength and with more sensitive sequences may lead to the detection of subtle or small brain abnormalities that would not have been detected previously.

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الأورام الوعائية الكهفية الدماغية: دراسة انتشار شعاعية مع المظاهر السريرية: دراسة استعادية لفترة عشر سنوات في مستشفى تعليمي

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الملخص

الأهداف: تظل الأورام الوعائية الكهفية الدماغية مثاراً للجدل في أوساط أطباء الجملة العصبية. يمكن لهذه الأورام أن تكتشف صدفةً، أو يمكن لها أن تظهر بأعراض واضحة. هدف هذه الدراسة: هو معرفة مدى انتشار مثل هذه الأورام في عينة من المرضى خلال عشر سنوات تمّ خلالها إجراء صور الرنين المغناطيسي للدماغ لهم لأسباب مختلفة. طريقة البحث: تمت دراسة 14327 تصويرا بالرنين المغناطيسي خلال الفترة بين عامي 2001 و2011م. تم اكتشاف 33 مريضا (18 إناثا، 15 ذكورا) يعانون من هذه الأورام وكان معدل العمر عند اكتشافها 32.3 ±16.2عاماً، مما يشير الى أن نسبة الانتشار تساوي 0.23% خلال هذه السنوات. تمّت دراسة النمط الشعاعي والسريري لهذه الحالات، وتمّ استثناء 4 حالات بسبب عدم وجود المعلومات السريرية في الحالات الـ 29 المتبقية، وكان هناك 35 ورماً، وأغلب هذه الأورام كانت في منطقة فوق الخيمة المخيخية خاصة في الفص الأمامي. وكانت نوبات الصرع أهم مظهر سريري عند الرجال، في حين كان الصداع أهم مظهر عند السيدات. الاستنتاج: لم تختلف هذه العينة عن غيرها من العينات السابقة باستثناء نسبة انتشار أقل ويمكن أن يسهم التصوير بالرنين المغناطيسي مع استعمال طرق كشف أكثر حساسية في الكشف عن مزيد من هذه الأورام.

الكلمات الدالة: الأورام الوعائية الكهفية الدماغية، مستشفى تعليمي، مظاهر سريرية.