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POPCORN IN BRAIN" –TYPICAL AND ATYPICAL LOCATIONS OF CAVERNOUS ANGIOMAS IN BRAIN: CASE SERIES IN A TERTIARY CARE HOSPITAL.

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ABSTRACT

INTRODUCTION: Cavernous haemangiomas (CH) are relatively uncommon non-shunting vascular malformations of the central nervous system and can present with seizures or with neurological deficits due to haemorrhage. Radiologists can often suggest the diagnosis of Cavernous haemangiomas based on characteristic magnetic resonance imaging (MRI) features, thus avoiding further invasive procedures such as digital subtraction angiography or surgical biopsy. Although typical MRI appearance combined with the presence of multiple focal low signal lesions on T2*-weighted images or the presence of one or more developmental venous anomaly within the brain can improve the diagnostic confidence, serial imaging studies are often required if a solitary Cavernous haemangioma presents at a time when the imaging appearances had not yet matured to the typical "popcorn" appearance. [1]

Aims and Objectives: To study the common and uncommon locations of intracranial cavernous angioma (CA) with various clinical presentation. **Materials And Method:** Retrospective study of 20 cases of cavernous angiomas in brain (case series) was studied using advanced cross sectional imaging modalities like Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) over a period of 2 years (Feb 2011-Oct 2013). **Result:** Cases studied were in the age group of 20-35yrs (12), 55-65yrs (8) with male: female ratio of 6:14. Most common clinical feature was headache and seizures (5/20), involuntary

movements (3/20). Other features were sudden loss of speech, left sided hemiplegia, memory loss and incidental finding (12/20). Magnetic resonance imaging- Findings Typical- Reticulated "popcorn-like" lesion, mixed signal core, complete hypointense hemosiderin rim, locules of blood with fluid-filled levels, surrounding edema. No enhancement.

KEYWORDS: Cavernous angiomas, magnetic resonance imaging, computed tomography. Seizure.

INTRODUCTION

Cavernous haemangiomas (CHs) are uncommon, angiographically occult vascular malformations of the central nervous system (CNS), with an incidence of 0.5 and 0.7% at autopsy and magnetic resonance imaging (MRI), respectively. CHs, also called cavernous malformation, cavernous angioma, or cavernoma, are composed of abnormally enlarged, discreet, thin-walled vascular structures within the parenchyma of the CNS. There is no brain parenchyma between the vascular channels, and the presence of blood products in various stages of degradation is responsible for the typical MRI signal characteristics of these lesions.[1] Although the majority of CHs develop de novo, rare associations with viruses, pregnancy, family history, and previous irradiation have been described. Symptomatic cerebral CH typically come to clinical attention in the third to fifth decades of life with Headaches, seizures, or focal neurological deficits.[2] Clinical manifestations in the paediatric age group are similar to those in adults. There is no sex predilection. Cerebral CH is increasingly detected at computed tomography (CT) and MRI performed in asymptomatic patients as well as those with headache, seizures, or intracranial haemorrhage. Although the typical imaging appearances of CH have been well described in the literature, these lesions can pose a diagnostic dilemma for radiologists, especially when atypical appearing lesions are encountered in unusual locations, or following recent intralesional haemorrhage. Hence, radiologists should be familiar with typical and atypical features of CH of the brain and spinal cord. [1]

Typical imaging appearances of cerebral CH

At CT, typical cerebral CHs may be identified as focal hyperdense lesions with indistinct margins, containing areas of blood or speckles of calcific density (Fig 1) These CT appearances are, however, non-specific and differential diagnosis such as granuloma, haemorrhage from other causes, vascular malformation, and neoplasm should be consider.[1] Perifocal oedema and mass effect are not typical features of uncomplicated CH, and usually

signify recent haemorrhage. MRI is the technique of choice for diagnosis of cerebral CHs, and they exhibit characteristic features that have been described as resembling a "mulberry" or "popcorn" appearance. This comprises a well- circumscribed, lobulated lesion with a reticulated core of heterogeneous signal intensity on both T1 and T2-weighted sequences histologically, the central mixed signal intensity comprises thrombosis, fibrosis, calcification, and various blood breakdown products. In addition, there is also a characteristic peripheral ring of hypointensity which corresponds to haemosiderin and iron deposition in the surrounding brain parenchyma.[1] On contrast-enhanced MRI sequences, the appearances are variable with CH appearing as non-enhancing lesions or at most, demonstrating minimal enhancement. Gadolinium administration is generally not indicated in patients with a suspected CH unless the appearance is significantly atypical and the imaging differential diagnoses include other conditions such as infections and malignant neoplasms. [2] Cerebral CH may also be associated with a nearby developmental venous anomaly .[1] Studies have reported an incidence of CH in up to 33% of individuals with DVA and co-existence of DVA in 23% of patients with CHs.[2] DVA comprise normal but variant venous vasculature (in contrast to CH, they are vascular anomalies and not abnormal malformations), and their synchronous presence can increase the diagnostic confidence for the adjacent cerebral CH.[3]

TABLE 1:

ZABRAMSKI CLASSIFICATION	MAGNETIC RESONANCE IMAGING APPEARANCES
TYPE 1 – SUBACUTE HEMORRHAGE	T1 : HYPERINTENSE T2 : HYPER/HYPOINTENSE
TYPE 2 – DIFFERENT AGE OF HEMORRHAGES: POPCORN - like	T1 AND T2 : MIXED SIGNAL INTENSITIES
TYPE 3 – CHRONIC HEMORRHAGE	ISO/HYPOINTENSE ON T1 AND T2 WEIGHTED IMAGES
TYPE 4 – PUNCTATE HEMORRHAGE	BLOOMING BLACK DOTS ON GRE AND SWI

METHODOLOGY

Retrospective study of 20 cases of cavernous angiomas in brain (case series) was studied using advanced cross sectional imaging modalities like Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) over a period of 2 years (Feb 2011- Oct 2013).

RESULTS

AGE GROUPS (YEARS)	NUMBER OF PATIENTS
15-25	12 (60%)
45-65	08 (40%)
GENDER	
MALE	06 (30%)
FEMALE	14 (70%)

TABLE 2:

CLINICAL PRESENTATION	
HEADACHE AND SEIZURES	05 (25%)
INVOLUNTARY MOVEMENTS	03 (15%)
SUDDEN LOSS OF SPEECH ,	
LEFT SIDED HEMIPLEGIA ,	12 (60%)
MEMORY AND BEHAVIOURAL DISTURBANCE	
INCIDENTAL FINDING	
TOTAL	20

TABLE 3:

OUT OF THE 20 PATIENTS

	TOTAL NUMBER OF PATIENTS	SYMPTOMATIC PATIENTS	AYSYMPTOMATIC PATIENTS
SUPRATENTORIAL REGION	15	10	05
INFRATENTORIAL	03	01	02
MULTIPLE SITES	02	01	01
TOTAL	20	12	08

TABLE 4:

Supratentorial

Out of 15 patients

TYPICAL LOCATION	NUMBER OF CASES	SYMPTOMATIC PATIENTS	ASYMPTOMATIC PATIENTS
FRONTAL LOBE {FIGURE 1}	06 (30%)	04	02
TEMPORAL LOBE	03 (15%)	02	01
OCCIPITAL LOBE	02 (10%)	01	01
ATYPICAL LOCATION			
RIGHT GANGLIO-CAPSULAR REGION WITH INSULAR CORTEX AND EXTERNAL CAPSULE	01 (5%)	---	01
HYPOTHALAMUS {FIGURE 2}	01 (5%)	01	---
LEFT THALAMO-CAPSULAR REGION	01 (5%)	01	---
BASAL GANGLIA	01 (5%)	01	---

TABLE 5

OTHERS

OUT OF 5 PATIENTS

INFRTENTORIAL REGION	NUMBER OF CASES	SYMPTOMATIC PATIENTS	ASYMPTOMATIC PATIENTS
CEREBELLUM AND VERMIS {FIGURE 3}	02(10%)	01	01
PONS {FIGURE 4}	01(5%)	---	01
MULTIPLE {FIGURE 5}	02 (10%)	01	01

COMPUTED TOMOGRAPHY	FINDINGS
TYPICAL	WELL-DELINEATED ROUND/OVOID CENTRAL HYPERDENSE LESION. CALCIFICATION +/-NO MASS EFFECT/ PERILESIONAL EDEMA NO ENHANCEMENT
ATYPICAL	MASS EFFECT - ACUTE HEMORRHAGE

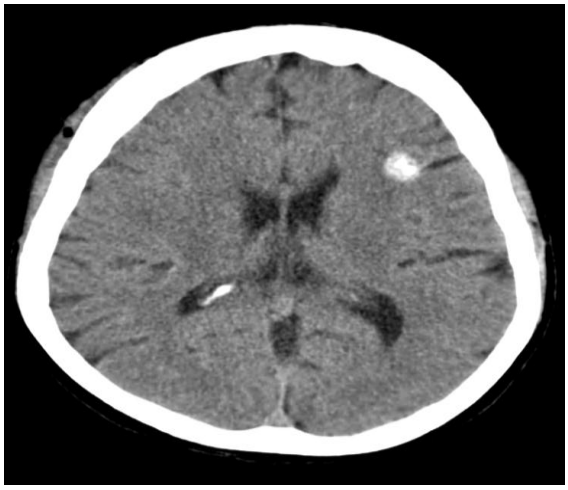

Figure 1: Left Frontal Lobe: Axial Non-Contrast Computed Tomography–Shows Hyperdense Nodular Lesion in the Left Frontal Lobe with Punctate Calcification (Arrow) Without.

SURROUNDING EDEMA OR MASS EFFECT. HYPOTHALAMUS

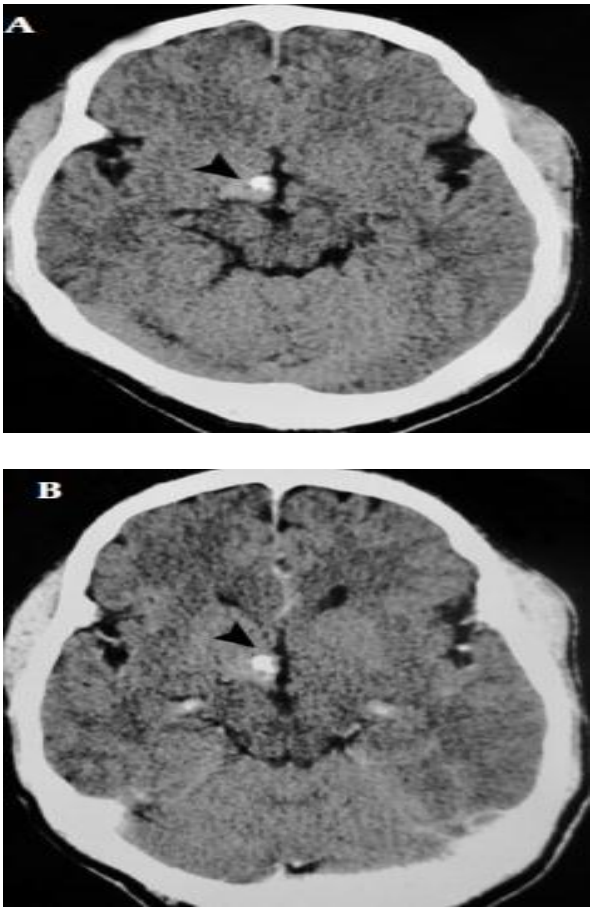

Figure 2: Axial Non-Contrast Computed Tomography, B – Axial Contrast Enhanced Computed Tomography – Shows A Mildy Enhancing Hyperdense Nodular Lesion On The Right Side Of The Hypothalamus With Punctate Calcification (Arrowhead) Without Surrounding Edema Or Mass Effect.

CAVERNOMA IN THE VERMIS

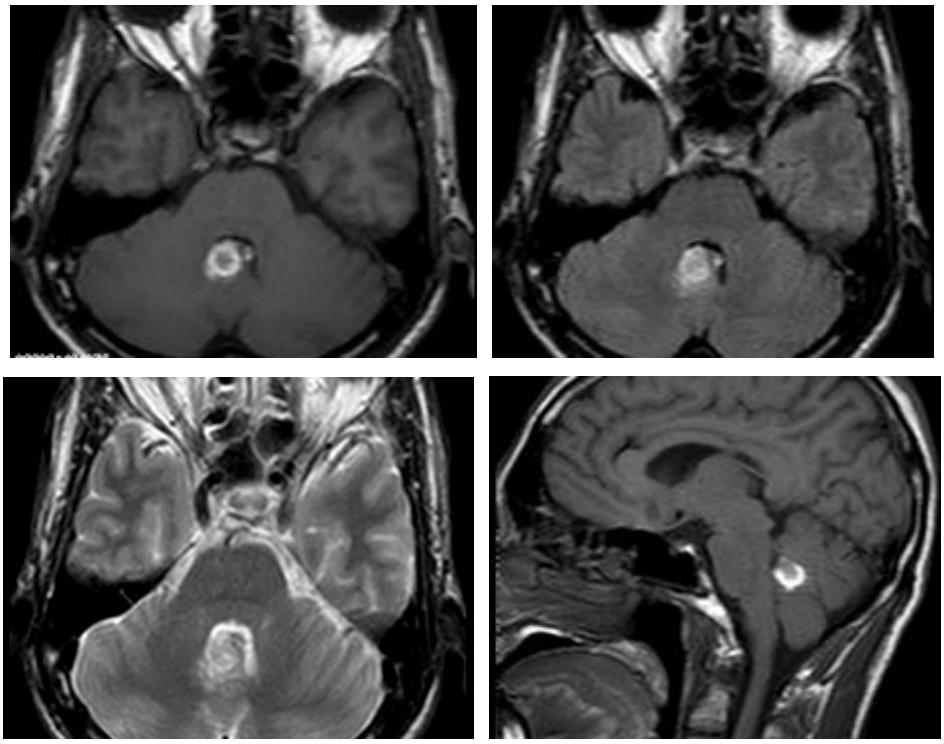

Figure 3:– Axial T1 Weighted, C–Axial T2 Weighted, D- Sagittal T1weighted Magnetic Resonance–Shows A Well Defined Mixed Intensity Lesion In The Vermis On T2 Weighted Imageswith Central Hyperintensity And Peripheral Hypointense Rim.

LEFT SIDE OF THE PONS

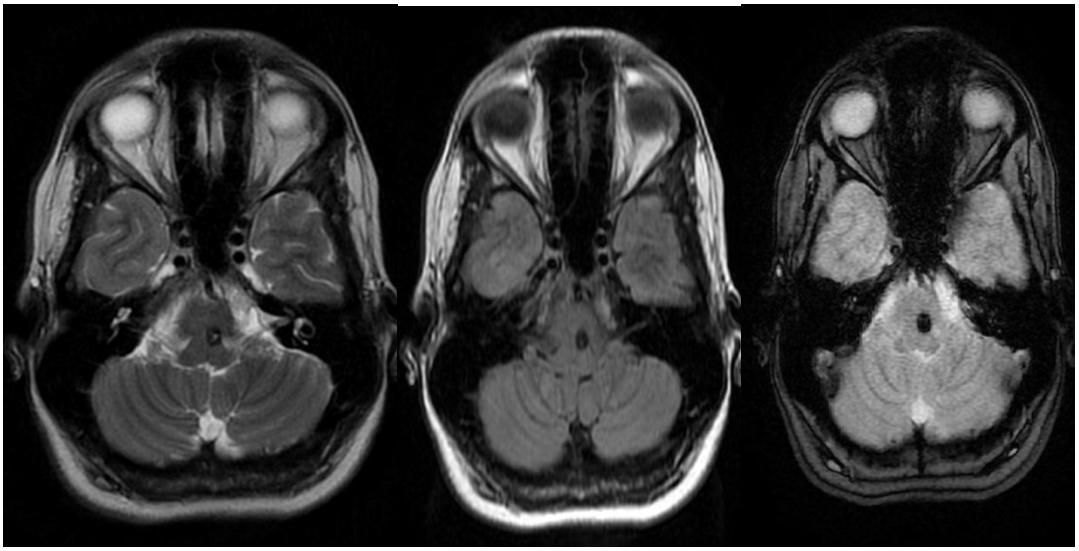

Figure 4: Axial T2 Weighted, B- Axial T2 Flair, C – Axial Gradient Magentic Resonance – Shows Well Defined Rounded Area Of Altered Signal Intensity Appearing Mixed Intensity On T2 Weighted IMAGES WITH HYPOINTENSE RIM ON T2 WEIGHTED AND GRADIENT IMAGES IN LEFT SIDE OF PONS.

MULTIPLE CAVERNOMAS

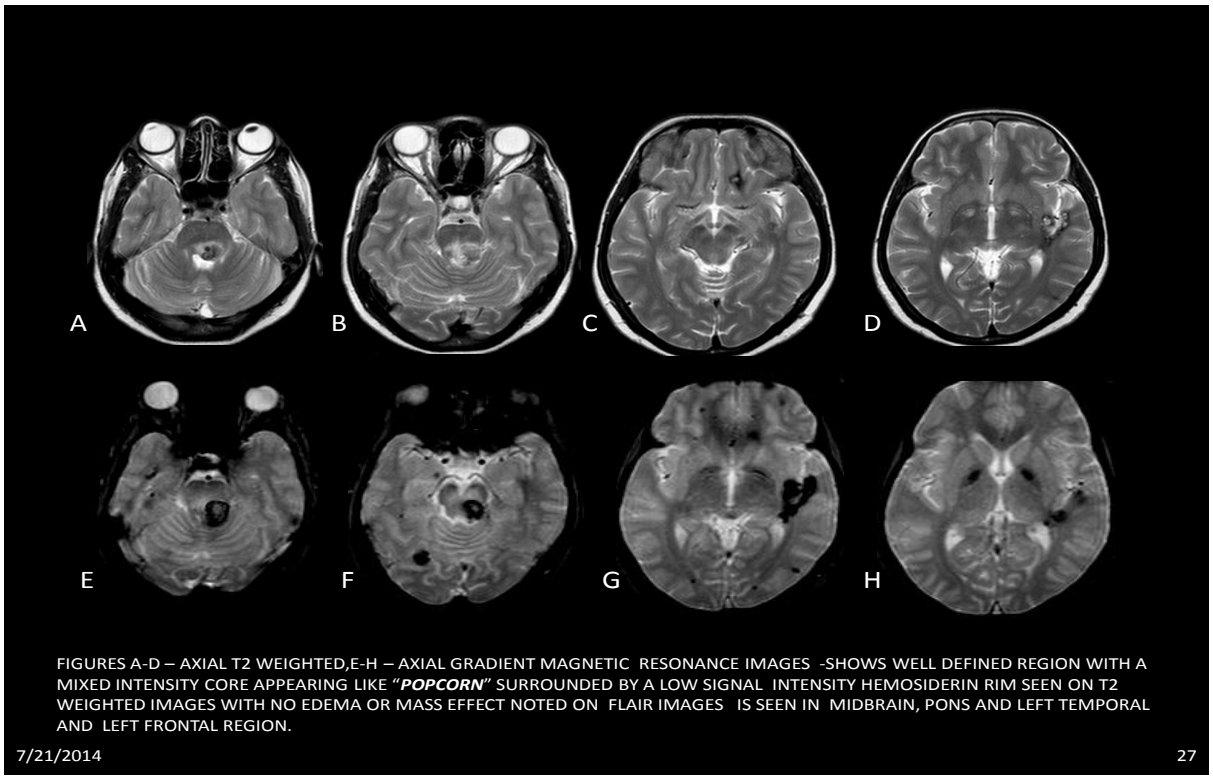

FIGURE 5:

DISCUSSION

A cavernous angioma (also known as a cavernous malformation or cavernoma) is a type of vascular malformation. Consisting of a low-pressure collection of vein-like vessels in the shape of caverns, cavernous angiomas can occur anywhere in the brain, brain stem or spinal cord. Cavernous angiomas may cause seizures and frequently leak blood into the surrounding brain. Severe or frequent bleeding may lead to the development of a neurological deficit such as weakness, numbness or double vision. Rarely they may cause a large, life-threatening hemorrhage. [1]

Cavernous angiomas are best recognized on an MRI scan of the brain. They appear as "popcorn"-shaped lesions surrounded by a dark ring made up of old blood from previous leaks. On CT they can be identified as small hemorrhage areas. The malformations are not visible by a conventional angiogram, and for this reason, cavernous angiomas are often referred to as angiographically occult vascular malformations (AOVM). [2]

The sensitivity of magnetic resonance imaging (MRI) to flowing blood and blood products of varying ages, as well as the greater contrast resolution of MRIs, greatly increases the specificity of MRI compared with that of CT scanning. Combining multiple MRI sequences

has largely eliminated misdiagnosis of cavernous angiomas, because they have relatively specific signal characteristics. [6]

Additionally, gradient-echo imaging, with its increased sensitivity to susceptibility artifact, is useful in the detection of smaller and concomitant lesions, which may not be detected with traditional sequences. [4]

CT scanning and MRI can be used in the follow-up monitoring of patients with known cavernous angiomas, particularly when hemorrhagic events are suspected. Although the MRI appearance of cavernous angiomas is not helpful in predicting future bleeds, MRI is the method of choice for the long-term follow-up of patients with cavernous angiomas and for the assessment of family members in whom similar lesions are suspected. In addition, MRI is extremely helpful in presurgical planning to assess the extent of the lesion, define borders, and plan the surgical approach and exposure. 1Most cavernous malformations are angiographically occult, and when they are evident on angiograms, the findings are nonspecific. When the lesions occur in combination with other vascular malformations, as they do in as many as 30% of patients with venous malformations, MRI characteristics become more complicated and less specific. In these patients, angiography can be helpful in further defining the lesions. [2]

Limitations of techniques

CT scanning has only a limited role in the diagnosis of cavernous angiomas, largely because of its relative lack of specificity. CT scan findings are compatible with low-grade gliomas, hematomas, granulomas, and inflammatory conditions such as tuberculomas and sarcoidomas. When calcified and located near the dura, cavernous angiomas can even resemble meningiomas. CT images also cause small lesions to be missed altogether, and cavernomas, when they present as acute intracerebral hematomas, may not be detected by using nonenhanced CT scanning. [4]

MRI may cause small lesions to be missed if T2-weighted pulse sequences, such as T2-weighted fast spin-echo sequences, are used because these can be less sensitive to chronic hemorrhage. Additionally, even standard T1- and T2-weighted images can fail to depict minute concomitant lesions. Therefore, T2-weighted gradient-echo sequences, with their increased magnetic susceptibility effects, always should be performed during an evaluation for smaller or multiple lesions that may not be visible on standard spin-echo images.1

CONCLUSION

• Cavernous angiomas are increasingly detected with advanced cross sectional imaging performed in asymptomatic patients and those with headache and seizures.

• It can pose a diagnostic dilemma for radiologists, when they are in atypical locations, or following recent intralesional haemorrhage as they have to be differentiated from hemorrhagic metastases and other occult arterio-venous malformations.

• Hence, radiologists should be familiar with typical and atypical clinical presentation and locations of cavernous angioma of the brain.

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