Cerebral Vascular Malformations in Hereditary Hemorrhagic Telangiectasia

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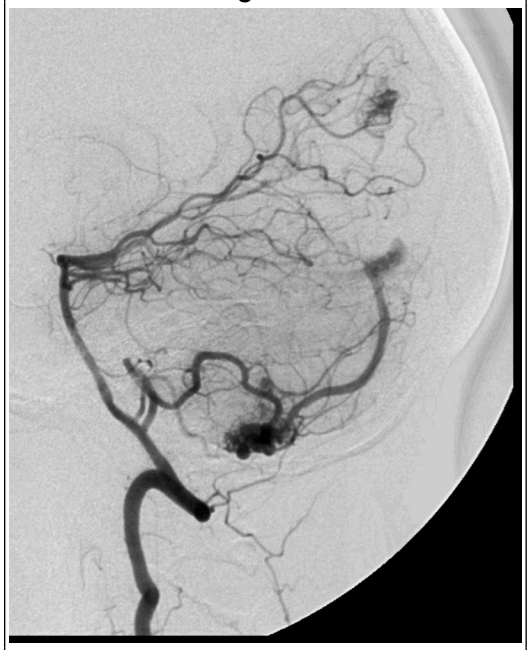
Introduction

Hereditary Hemorrhagic Telangiectasia (HHT) is a hereditary disorder characterized by mucocutaneous telangiectasias, frequent nosebleeds, and visceral arteriovenous malformations. Few reports have outlined the prevalence of the various cerebral vascular malformations found in HHT patients. We set out to compare the prevalence of cerebral vascular malformations in HHT to that of a control population imaged with 3-Tesla Magnetic Resonance Images (MRI) of the brain.

Figure 1



Lateral Vertebral Artery Injection in an HHT Patient

Methods

A retrospective review of prospectively collected data was carried out using a database of

372 patients seen and examined at the Georgia Health Sciences University HHT Center. For control, 413 consecutively collected MRI examinations of the brain were reviewed for the presence of cerebral vascular malformations in a blinded fashion.

Vascular Malformations in HHT versus Controls

	Definite HHT* (%)	Controls (%)	*p*-value
	n = 209	n = 413	
AVM**	7.7	0.5	<0.001
AVF***	0.5	0	NS****
Cavernous Malformation	1	2.7	NS
Telangiectasia	1.9	0.7	NS
DVA*****	4.3	1.2	<0.025
Aneurysm	2.4	0.5	<0.05

Table 1. Types of Cerebral Vascular Malformations

*HHT = Hereditary Hemorrhagic Telangiectasia

**AVM = Arteriovenous malformation

***NS = Not Statistically Significant

****AVF = Arteriovenous fistula

*****DVA = Developmental venous anomaly

Results

There was a significant difference in the number of arteriovenous malformations (AVM) (7.7%, $p < 0.001$), developmental venous anomalies (DVA) (4.3%, $p < 0.025$), and cerebral aneurysms (2.4%, $p < 0.05$) in the HHT group as compared to the control group (0.5%, 1.2%, and 0.5% respectively). To our knowledge, this is the first report of an increased prevalence of cerebral aneurysms in the HHT population. The prevalence of multiple arteriovenous shunting lesions was also increased in the HHT group ($p < 0.05$). HHT AVMs tended to be supratentorial, small, and cortical in this series – findings consistent with other recent studies in the literature. There was no significant difference in the number of arteriovenous fistula, cavernous malformation, or capillary telangiectasia when comparing the HHT group to the control group.

HHT AVM Characteristics

	Woodall et al.`	Bharatha et al.	*P*
AVM Number	13	89	
Supratentorial	85% (11)	93% (52 of 56)	NS``
Size < 3cm	100% (13)	88% (37 of 42)	NS
Eloquent Location	54% (7)	45% (30 of 67)	NS
Cortical Location	85% (11)	90% (53 of 59)	NS

Table 2. HHT AVM Characteristics
Expressed as Percentage (number of lesions)
`Present study
``NS = Not statistically significant ($p < 0.05$)

Conclusions

The prevalence of arteriovenous malformations, developmental venous anomalies, and cerebral aneurysms is increased this population of HHT patients when compared to controls screened with 3-Tesla MRI examinations of the brain. HHT AVMs are more likely to be multiple, and have a tendency toward small size and cortical location. As such, they are often treated with a single-modality therapy. Current consensus guidelines call for MRI/MRA screening examinations of the brain in patients diagnosed with definite HHT by modified Curacao criteria.

Learning Objectives

1) Review vascular malformations in HHT 2) Discuss AVM characteristics in the HHT population 3) Discuss screening recommendations for brain vascular malformations in the HHT population