Ivy Sign in Moyamoya Disease

Abstract

Moyamoya disease is an idiopathic disease characterized by the progressive stenosis and collateral development of the distal internal carotid arteries. In this disease, several collateral vascular structures develop following stenosis and occlusion. The “Ivy sign”, which was described in Moyamoya disease, is a characteristic Magnetic rezonance imaging (MRI) finding frequently encountered in patients with moyamoya. It can be observed both in post contrast T1-weighted images and Fluid attenuated inversion recovery (FLAIR) images. While this sign manifests in the form of contrasting on the cortical surfaces due to the formation of leptomeningeal collateral development and increased numbers of pial vascular webs on post contrast images, in FLAIR images it originates from the slow arterial flow in the leptomeningeal collateral vascular structures. In this case, we presented the Digital subtraction angiography (DSA) signs of moyamoya disease and “Ivy sign” in MRI and its development mechanism in a 16 years old female patient.

Keywords: DSA, MRI, Moyamoya Disease, Ivy Sign

Introduction

Moyamoya disease is an idiopathic disease characterized by the progressive stenosis and collateral development of the distal internal carotid arteries. In this disease, several collateral vascular structures develop following stenosis and occlusion. The “Ivy sign”, which was described in Moyamoya disease, is a magnetic resonance imaging (MRI) image finding and indicator of the developed leptomeningeal collaterals. In this case, we presented the Digital subtraction angiography (DSA) signs of moyamoya disease and “Ivy sign” in MRI and its development mechanism in a 16 years old female patient.

Case Report

A 16 years old female patient with recurrent bilateral paresis complaints applied to our department for cranial imaging. Her medical history had no characteristic features except for a continual frontal headache for the past 1-2 years. The neurological examination was normal except for bilateral loss of strength in the lower extremities, being more notable on the left side. On T2-weighted (T2W) and Fluid attenuated inversion recovery (FLAIR) images in the contrast-enhanced cranial MRI (Figures 1 a, b), there was loss of volume and increased signal compatible with chronic infarction in the frontal-parietal region at convexity level consistent with the Middle cere-
bral artery (MCA) irrigation area and the left temporal lobe posterior area. On the post-contrast images, leptomeningeal contrasting was noted at the convexity level in both hemispheres and basal cistern, particularly on the left. Contrasting left convexity level was evaluated as being compatible with the typical ‘ivy sign’ (Figure 2 a, b.). In addition, loss of ‘flow-void’ which was compatible with stenosis-occlusion in both Internal carotis artery (ICA) cavernous sinus segments was observed in T2W images. On cranial TOF angiography and contrasted cervical MR angiography, occlusions were present in both ICA cranial segments. On the DSA examination performed, both internal carotid arteries appeared completely occluded from the ophthalmic segments. Widespread collateral development was observed at the frontal-basal region bilaterally and the posterior parietal occipital region. Diffuse osteodural anastomosis from the bilateral external carotid artery to the internal carotid artery branches drew attention (Figure 3 a, b.). On right vertebral artery injections, both MCA distal territory regions, especially the left side, appeared to be reconstructed with the developed collaterals (Figure 4 a, b.). Consequently, all these findings were evaluated as being compatible with idiopathic moyamoya disease.

Discussion

Moyamoya disease is an idiopathic disease characterized by the progressive stenosis-occlusion and collateral development of the distal internal carotid arteries [1]. The incidence of Moyamoya disease is high in eastern Asian countries such as Korea and Japan. Its estimated incidence in Japan is 0.35-0.94/100,000 [2]. The incidence in the USA and Europe has been reported as being about 10% of that of Japan [3]. It is observed as being 1.8-2.2 times more common in women than in men. Bimodal age dissemination in Moyamoya disease has been reported to show high peaks at ages over 5 and low peaks at age 40 [4]. Familial transmission is observed in 15% of these patients [5]. The increased levels of serum EBV DNA and antibodies found in these patients suggest an infectious aetiology [6].

Moyamoya disease shows different clinical features in children and adults. While it typically presents with subarachnoid and intra parenchymal bleedings in adults, in children it presents with transient ischemic attacks with infarcts developing predominantly in the ICA territory in the frontal lobe. The typical symptoms in children include monoparesis, hemiparesis, aphasia and dysarthria [6].

Several collaterals develop in these patients in order to compensate the stenosis occlusion. The first collateral is known as the basal moyamoya. It includes abnormal dilatation of the lenticulostriate and thalamoperforating arteries in the thalamus and basal ganglia (moyamoya vessels). The second collateral includes dilatation of the anterior coroidal and posterior pericallosal arteries. The third collateral is known as the ethmoidal moyamoya. It includes the abnormal dilatation of the anterior and posterior ethmoidal arteries that supply blood to the ACA branches from the ophthalmic arteries. The last collateral path is between the dural arteries and the pial arteries and is called ‘vault moyamoya’ [6]. Stenotic occlusive changes in the proximal posterior cerebral artery are observed in 25% of moyamoya patients [7]. The hazy appearance in angiography simulates cigarette smoke (moyamoya in Japanese). Moyamoya disease was first given a name by Suzuki and Takaku [8].

The diagnosis of moyamoya disease can be made when all other causes of cerebrovascular disease have been elmi-
nated. If diseases such as Down Syndrome, NFM type-1, Sickle Cell Anaemia, radiation treatment, glycogen storage disease type-1a, hereditary spherocytosis, tuberculosis meningitides and SLE are associated with the moyamoya vessels, this is called moyamoya syndrome [6, 9].

The golden standard method for the diagnosis of moyamoya disease is cerebral angiography. However, since cerebral angiography is invasive and not easy to perform, particularly in paediatric age groups, the primarily preferred method is non-invasive MRI and MR angiography. In FLAIR and post contrast T1W images of moyamoya patients, compensatory pial vessels and leptomeningeal anastomoses and dilatation of the medullary vascular structures cause the 'ivy sign' and 'medullary streak sign' images respectively.

The "ivy sign" described in moyamoya disease is a finding on MR image. This finding is described in both post-contrast series and FLAIR images. In post contrast images, it manifests as contrasting in the cortical surfaces due to the development of leptomeningeal collaterals because of the increased number of obvious and pial vascular network formations [10]. The 'ivy sign' was first described in post contrast MR sequences as a gadolinium accumulation in the pial vascular network appearing as leptomeningeal and cortical contrasting images. This finding has been described in post contrast MR images of approximately 70% of moyamoya patients. However, in later reports, the same finding was also reported as a cortical and leptomeningeal signal increase in FLAIR images [10, 11]. This signal increase originates from the slow arterial flow of the leptomeningeal collateral vascular structures. This leptomeningeal contrasting on the cortical surface and FLAIR signal increase resembles ivy growing on a rock, this appearance is called the "ivy sign". Maeada and Tsuchiada [12] have also suggested that this finding may originate from leptomeningeal thickening due to vascular congestion. MR images of the 'ivy sign' decrease following successful bypass operations [10].

Contrast-enhanced FLAIR MR imaging has been reported as an examination that can be used in the diagnosis of patients with cerebral glioma and metastasis [13]. In some other studies, it has been reported that contrasted T1W images are superior to contrasted FLAIR images for showing intraparenchymal lesions [14]. Yoon et al. [10] have mentioned that post contrast T1W images are more successful than post contrast FLAIR images for showing the ivy sign. Nevertheless, post contrast FLAIR imaging is a technique that can be used in the evaluation of meningeal diseases since it does not display vessel contrasting.

There is no effective medical treatment for moyamoya disease. Surgical revascularization improves cerebral perfusion and reduces the risk of stroke development in both paediatric and adult patients [6]. In paediatric patients, the incidence of transient ischaemic attack decreases rapidly following surgery. Surgical procedures are classified in three groups. Direct bypass extending from the superficial temporal artery to the MCA, encephaloduoroarteriosynangiosis (EDAS) and encephalomyosynangiosis (EMS) including indirect bypass and combined bypass techniques.

The ivy sign is a characteristic finding frequently encountered in patients with moyamoya. It can be observed both in post contrast T1W images and in FLAIR images. While this sign manifests in the form of contrasting on the cortical surfaces due to the formation of leptomeningeal collateral development and increased numbers of pial vascular webs on post contrast images, in FLAIR images it originates from the slow arterial flow in the leptomeningeal collateral vascular structures.
**Informed Consent:** Written informed consent was obtained from patients/patients’ parents/ the parents of the patients/patient who participated in this study.

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**References**