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Homonymous Hemianopia Secondary to A Long *Fusiform Aneurysm* of Posterior Cerebral Artery in A Patient with Connective Tissue Disease

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ABSTRACT

Introduction: Fusiform aneurysms are uncommon, accounting for 1% of all intracranial aneurysms. Dissection and atherosclerosis are the main causes of this vasculopathy, but connective tissue disease is a very uncommon cause. Ehlers-Danlos Syndrome is the most common connective tissue disease, accounting for 11% of all cases. Symptoms depend on the location and size of the aneurysm, including headaches, blurred or double vision, and focal neurological deficits. **Case:** A 36-year-old man suddenly experienced blurred vision in both eyes on the right, starting with a chronic left-sided headache and no history of cardiovascular disease. In the confrontation test, Humphrey gave the right homonymous hemianopia. A head CT scan showed a lobulated lesion which showed enhancement in the left suprasellar region, and cerebral digital subtraction angiography (DSA) gave the impression of a long fusiform aneurysm L PCA. Clinically, the patient's skin on the left side of his face was darker than on the right, his skin was more elastic, and his blood vessels were wider and more prominent on the side of the fusiform aneurysm. **Conclusion:** Posterior circulation involvement is only 3-13% of cases of intracranial aneurysms. Many cases of intracranial aneurysms are not detected before rupture, resulting in delays in treatment. Surgical or endovascular surgery can be performed if the size is >10 mm and causes clinical symptoms. Symptoms of ischemia are managed with antiplatelets or anticoagulants. *I*ncidentally detected unruptured aneurysms are generally managed conservatively because of the highly friable nature of the blood vessels in patients with connective tissue diseases.

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INTRODUCTION

An aneurysm is defined as a widening or dilatation of the blood vessels.[1] The most common intracranial aneurysm is a saccular arterial aneurysm, a progressive degenerative process involving the arterial wall.[2] Fusiform aneurysms are uncommon, accounting for only 1% of total intracranial aneurysms, although the number of cases has increased in recent years, especially in younger patients.[3] It is most common in the vertebrobasilar system, followed by the middle cerebral artery (MCA), internal carotid artery (ICA), and anterior cerebral artery (ACA).[3,4]

Aneurysmal processes in arteries that supply the central nervous system can be classified based on their morphology (saccular, fusiform, and dissecting), size (non-giant or giant, with a maximum diameter of > 2,5 cm), vascular type (arterial or venous), etiology (acquired or familial/genetic), underlying disease (traumatic infection, inflammation, neoplastic) and location (intracranial, cranial bases, extracranial, spinal, and systemic).[4,5] Fusiform aneurysms have different characteristics in pathology, hemodynamics, anatomic distribution, formation process, and treatment than the saccular type. The two leading causes of this type of aneurysm are dissection and atherosclerosis. Dysfunction of collagen and elastin metabolism, infection, neoplastic invasion of the arterial walls, and iatrogenesis are other causes of this vasculopathy.[1,3,6]

Patients with connective tissue diseases are at a higher risk for cerebrovascular diseases such as intracranial aneurysms, dissections, and acute ischemic strokes. The most commonly accepted causal explanation for this association is that genetic mutations involved in such connective-tissue diseases affect the collagen and proteoglycans that construct the extracellular matrix, thus weakening the vessel wall. Neurovascular specialists should be aware of these diseases' various cerebrovascular manifestations because there is a higher prevalence of these neurovascular complications in patients with connective-tissue diseases.[7]

A dissecting aneurysm is an aneurysm secondary to separate layers in the arterial wall after the intimal layer of the arteries is torn. The gap formed by splitting the artery wall's layers becomes the entry point for blood flow. The gap progressively gets larger, creating a balloon-like structure that may be painful. Any pain or symptoms that arise will be fatal if this aneurysm ruptures. Some symptoms include headaches, blurred or double vision, nausea, vomiting, and focal neurological deficits.[2,6]

CASE

A 36-yer-old male working as a private employee who had previously been in relatively good health complained of visual disturbances such as blurry vision, particularly when looking in a specific direction. Blurred vision is felt in both eyes right side suddenly. An additional complaint that is felt is a mild headache on the left side that has been felt for a long time.

The confrontation test revealed right homonym hemianopsia on physical examination, while other neurological examinations were normal. This result is supported by the *Humphrey* test, which showed a similar result. On local examination in the left facial area, the skin was brown, elastic, and wrinkled with prominence and widening of the blood vessels, which leads to connective tissue disease.

Laboratory investigations showed normal values. The ECG examination was also within normal limits, allowing cardiac risk factors to be ruled out. A head CT scan showed lobulated lesions exhibiting enhancement post-contrast in the left suprasellar region with a size of 36x23x25,5 mm adjacent to the left posterior communicating artery (Figure 1).

The results of the MRI examination of the head showed a lesion measuring 4,06 x 2,66 x 3,70 cm of the left temporal on T1W1 and hypo-iso intensity of T2W1, predominantly hypointense on FLAIR, unrestricted DWI, which most likely suggests an AVM dd/aneurysm (less likely) (Figure 2a). An MRA examination of the brain suggested a dilatation of the left PCA, giving the impression of an aneurysm (Figure 2b).

Cerebral digital subtraction angiography (DSA), a modality that can visualize intracranial vascular, revealed dilated L PCA from P1 to P2 segment, accompanied by a late arterial draining phase in those regions. A late arterial L PCA on the P3 and P4 segments is late perfusion of L PCA blood flow to the occipital. No early venous phase drainage is visible, which gives the impression of a long fusiform aneurysm of the L PCA of segments P1 to P2 (Figure 3).

In this case, the diagnosis of connective tissue disease was based on clinical and investigational data, but skin biopsies are the gold standard for establishing connective tissue disease.

DISCUSSION

EDS (Ehlers-Danlos Syndrome) describes a group of disorders with shared primary characteristics of skin hyperextensibility, joint hypermobility, and tissue fragility. One of 5000 people is affected by this illness, though some types are more uncommon than others. Each of the six main types of EDS has distinctive genetic flaws and inheritance patterns. Classic (EDS types I and II), hypermobility (EDS type III), and vascular (EDS type IV) are the most observed of these types, with vascular EDS having the most prominent neurovascular manifestations.

The diagnosis of EDS and its various types

should be suspected when some combination of their shared features, such as joint hypermobility, multiple joint dislocations, translucent skin, poor wound healing, easy bruising, unusual scars, spontaneous ruptures of organs, and dissections of blood vessels. In addition, genetic testing for specific genes is available for all EDS types except for the hypermobility type.[8]

Type III collagen, mutated in EDS IV, is essential in providing structure and strength to connective tissue (skin, blood vessels, and internal organs) as a major component of the extracellular matrix. It is hypothesized that the poor assembly of these type III collagen fibers in EDS IV results in the neurovascular manifestations of EDS IV.[8,9] The potential relationship between collagen deficiencies in EDS and cerebral aneurysms has long been hypothesized but never tested in a prospective screening study. In EDS patients, intracranial aneurysms can be saccular or fusiform and are typically located in the cavernous sinus. Studies on the prevalence of aneurysms and subarachnoid hemorrhage in EDS patients have shown varying results. In the largest study of EDS patients with intracranial vascular imaging, the prevalence of unruptured intracranial aneurysms was 11%.[10]

This suggests that the site of the associated lesion may be in the left optic tract. Based on the initial diagnosis of non-hemorrhagic cerebrovascular disease, the process may take the form of vascular ischemic of the blood vessels that supply the affected area, which can happen when the blood vessels are blocked or narrowed. It could also be due to the mass effect or dilated blood vessels compressing the left optic tract.[11] The left optic tract is in the left temporal area, near the suprasellar region. So, if occlusion or ischemia exists in the area, there should be an additional neurologic deficit. The most likely diagnosis is the mass effect or dilated blood vessels compressing the left optic tract, which is accompanied by a headache in the patient.

The cause of fusiform aneurysms can be caused by vascular factors, namely hypertension and atherosclerosis. This patient has no history of hypertension or atherosclerosis, so the cause of vascular factors can be ruled out. There is no risk factor of infection, symptoms of infection, or a history of previous infections found in this case. According to some studies, abnormal connective tissue, particularly collagen, is the most common cause of fusiform aneurysms after vascular events and infections.[9,10]

The most common collagen abnormalities are Ehlers-Danlos Syndrome type IV and neurofibromatosis type I. The clinical features of café-au-mark type I neurofibromatosis lit spot, a pigmented spot on the skin with a predilection for the arm or thigh area, are associated with skeletal abnormality and peripheral neural sheath tumor.[9] The patient did not indicate those clinical manifestations. Therefore, neurofibromatosis could be ruled out. Ehlers-Danlos Syndrome type IV is a hereditary collagen tissue disease characterized by clinical manifestations such as elastic skin with prominent vascular on the skin surface, with visible premature aging.[10] These clinical diagnoses were discovered in the patient. On the same side as the fusiform aneurysm (left side of the face), there was hyperpigmentation, more elastic, with dilated and prominent blood vessels compared to the opposite side of the face, which has been there since birth (Figure 4).

According to the case report, the patient is thought to have Ehlers-Danlos syndrome because the clinical diagnosis of vascular Ehlers-Danlos syndrome is based on four criteria: a characteristic facial aspect (acrogeria) in most patients; thin and translucent skin with obvious subcutaneous vessels; ecchymoses and hematomas, and arterial, digestive and obstetrical complications. Based on that, the diagnosis of Ehlers-Danlos Syndrome type IV is most likely. Of course, clinical examination is not enough to establish a diagnosis, so further investigation must be conducted, including histopathology tests to find the type III collagen deficiency.

CONCLUSION

In this case, a fusiform-type aneurysm was found to be >10 mm, which had already caused a mass effect. Therefore, the patient required an endovascular treatment by sacrificing the parent vessel with L PCA embolization from the bottom, expecting it to form collateral vessels.[12–15] It was preceded by a balloon test occlusion for about 5-20 minutes, followed by an evaluation of the movement and other neurological functions that would be sacrificed if the results were good or the collateral was formed. There is a risk of ischemia in the left posterior circulation and occipital mesencephalon if the collateral is not formed. The DSA results show that the areas supplied by L PCA are mesencephalon to the occipital and L PCA itself, even with late perfusion. With that in mind, endovascular treatment was not performed. Symptoms of ischemia are managed with antiplatelets or anticoagulants.[16,17,18]

Furthermore, the risks and benefits of various conservative, endovascular, and surgical management techniques for these lesions also poorly understood.

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ATTACHMENT

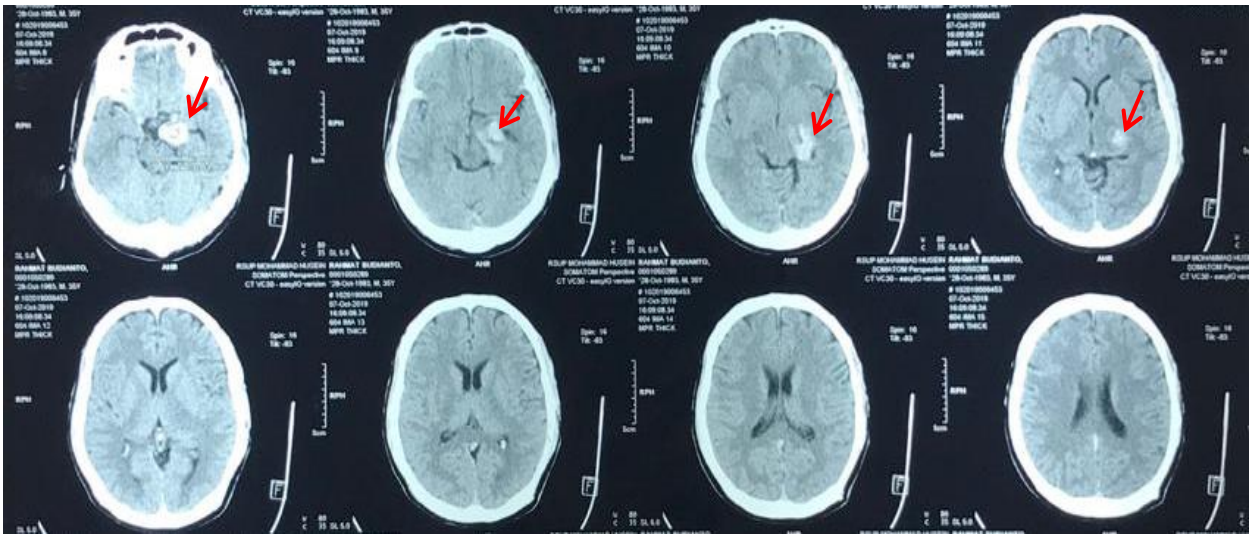

Figure 1. A head-contrast CT scan showed lobulated lesions, with enhancement post-contrast on the left suprasellar region (red arrows)

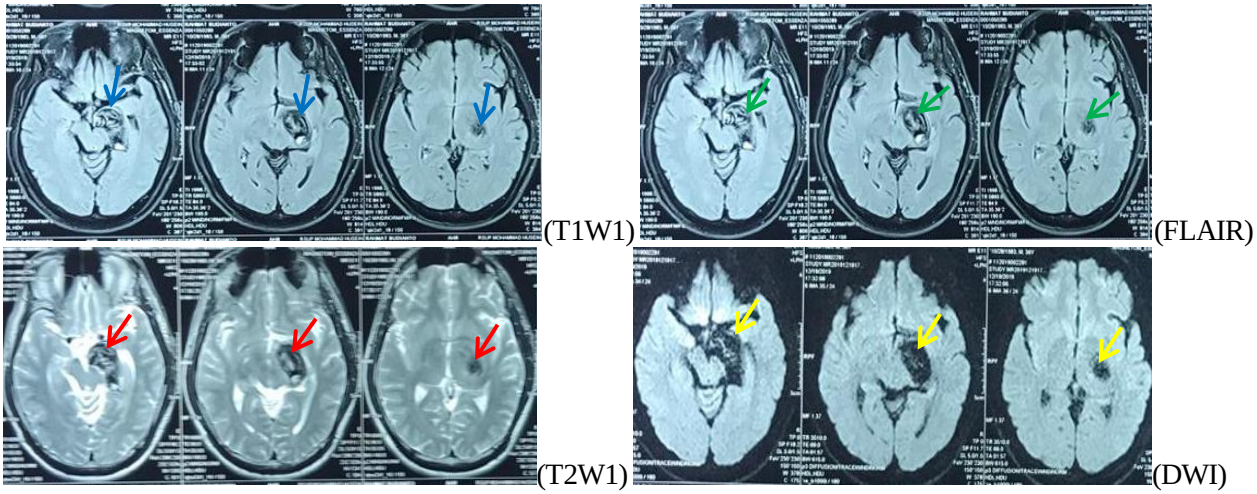

Figure **2a**. A head MRI showed a lesion measuring 4,06 x 2,66 x 3,70 cm of the left temporal on T1W1(blue arrows), hypo-iso intensity of T2W1 (red arrows), predominantly hypointense on FLAIR (green arrows), unrestricted DWI (yellow arrows)

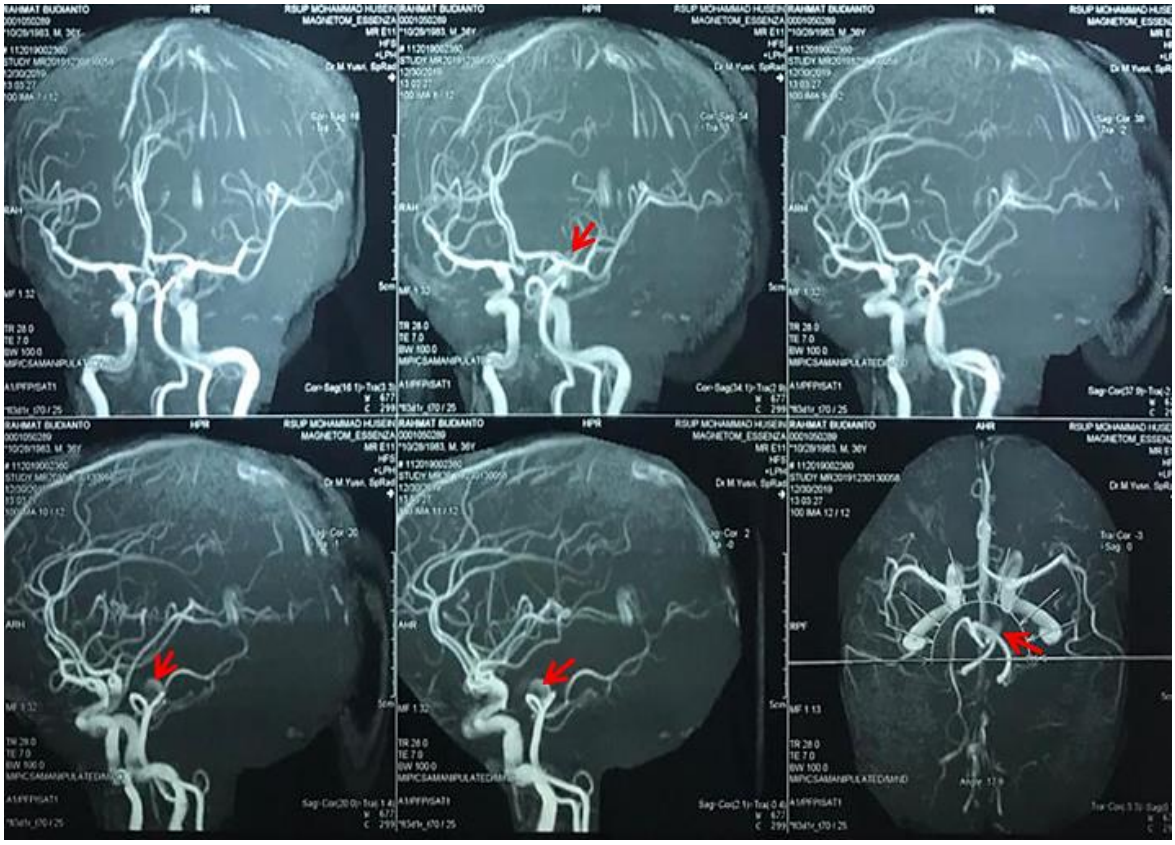

Figure 2b. Cerebral MRA showed a dilatation of the left PCA (red arrows)

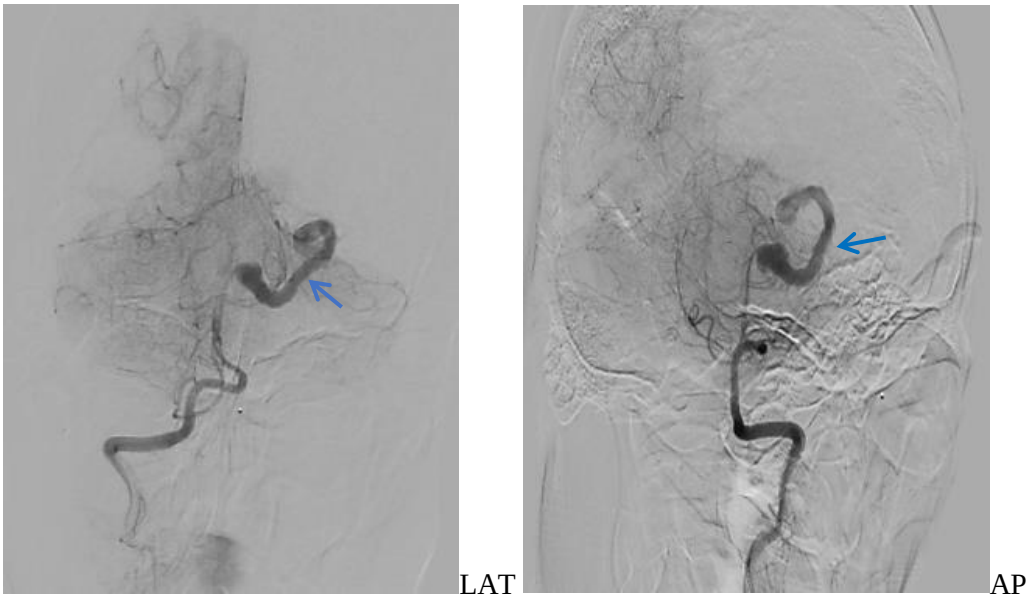

Figure 3. Cerebral DSA revealed dilated L PCA from P1 to P2 segment (blue arrows)

Figure 4. The face of patient revealed hyperpigmentation (red arrow), more elastic with dilated and prominent blood vessels (blue arrow)