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Co-existence of Three Types of Fibromuscular Dysplasia associated with True Posterior Communicating Artery Aneurysm

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Abstract

Coexistence of three types of fibromuscular dysplasia (FMD) is extremely rare and it has never been reported associated with true posterior communicating artery (PCoA) aneurysm. Here we present a 54-year-old patient with bilateral carotid and left vertebral arteries FMD involvement and a ruptured "true" aneurysm of the left PCoA itself that was treated with guglielmi detachable coils (GDCs) that allowed exclusion of the aneurysm with parent vessel preservation. As our case shows, flow-related true PCoA aneurysm and the common angioarchitectural aspects of PCoA of the previously reported cases highlight the importance of hemodynamic stress, reflected in changed blood flow patterns, for the formation and/or rupture of this unusual subgroup of intracranial aneurysms.

Keywords: Fibromuscular dysplasia; True posterior communicating artery aneurysm; Hemodynamic factors

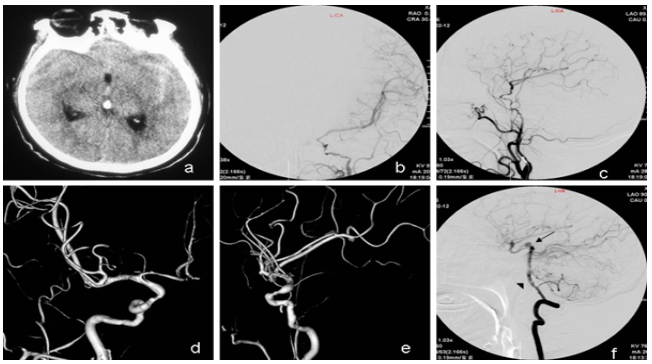
Introduction

Although FMD was first described by Lead better and Burkland in 1938, the first radiologically and histologically proven case of FMD of the internal carotid artery was reported by Connett and Lansche in 1965 [1,2]. Despite the fact that countless theories have been proposed, there have only been limited advancements in the knowledge of FMD during the recent 40 years, and the etiology and prevalence of FMD of cervical and intracranial arteries remains unknown [2,3]. We describe a patient who coexist three main types of FMD in the bilateral carotid and left vertebral arteries associated with a true aneurysm of the left PCoA. In this paper, we hypothesize that a changed blood flow pattern may be the main cause of formation and/or rupture of aneurysms in this location.

Case Report

A previously healthy 54-year-old woman experienced the sudden onset of a severe left-sided headache, as well as nausea and vomiting for about several times, was referred to our institute for suspected aneurysm due to the finding of diffuse subarachnoid hemorrhage noted in the left-sided Sylvain cistern and fourth ventricle on an axial unenhanced computed tomography (CT) scan (Figure 1a) performed at another hospital. There was no history of recent trauma to the cervical region. Examination at admission revealed a conscious, well oriented person with blood pressure of 160/100 mm Hg and pulse rate 72 bpm. She complained of severe pain in her neck and head. A neurological examination revealed somnolent, neck stiffness, and bilateral Babinski's sign. The rest of the physical examination was detected to be normal. There was no personal or family history of hypertension and cerebrovascular events. During the hospitalization the full blood count, serum chemistry and cholesterol were normal. Basic laboratory examinations were within normal range. An electrocardiogram disclosed sinus rhythm and no pathologic findings, and chest radiograph normal.

During the ensuing 2 days, we performed bilateral carotid and left vertebral angiograms under local anesthesia with sedation. In the both common carotid artery angiograms investigation, multisegmental stenoses of bilateral internal carotid arteries (ICA), suggestive of FMD, were found. At the area of stenosis in the left ICA, there was a marked decrease in the minimum luminal diameter. The lesion was fibro calcific and appeared to involve the adventitia (Figure 1b and Figure 1c). The left internal carotid angiogram demonstrated a complete filling defect of the A1 segment of the left anterior cerebral artery, and also showed a slight attenuated distal flow of the left middle cerebral artery. Frontal projection cerebral DSA with right internal carotid artery injections showed that the anterior cerebral artery on the left side is filled by the right ICA via the anterior communicating artery (Figure 1d and Figure 1e). A left vertebral angiogram demonstrated the classic



(a) Axial cranial unenhanced CT scan demonstrating SAH. (b) and (c) Angiography demonstrating a near-occlusive stenosis of the left ICA. (d) and (e) Three dimensional reconstructions of digital substraction angiography (3D– DSA) showing a focal, band-like, concentric stenosis of the right ICA. (f) A left vertebral angiogram demonstrated the classic "string of beads" appearance, with an aneurysm of the left PCoA itself. (arrow: "string of beads". arrow head: the true PCoA aneurysm).

Figure 1: Diffuse subarachnoid hemorrhage.

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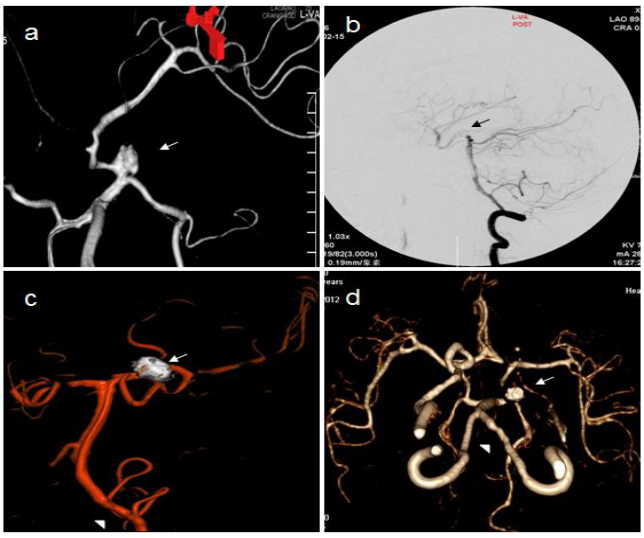
"string of beads" appearance of FMD in the left distal vertebral artery and revealed a small saccular aneurysm of the left PCoA itself located near to the proximal portion of the left posterior cerebral artery (PCA)-PCoA junction; the junction of the PCA and PCoA was not involved in the lesion. In addition, the angiogram indicated that the left middle cerebral artery was filled via the left PCoA (Figure 1f and Figure 2a). Due to the strictly rejected by the patient and her relatives, an initial attempt to perform endovascular therapy was aborted.

On day 4, the decision to proceed with endovascular embolization of the aneurysm was made after the relatives' careful consideration and assent. The procedure was undertaken under general anesthesia and full heparinization. The maximum and minimum diameters of the aneurysm dome were measured to select the size of detachable coils. The diameter of the dome was 6.81 mm×4.26 mm, and the diameter of the neck was 5 mm, so the neck of the aneurysm was judged to be wide at the PCoA. Four guglielmi detachable coils (GDCs, MicroVention, Inc. and Johnson & Johnson Family of Companies), with a total length of 41.2 cm, were used to totally exclude the aneurysm with the remodeling technique, without signs of embolic complications (Figure 2b and Figure 2c). Because the overall long-term prognosis of fibro muscular dysplasia of the cervico-cephalic arteries is quite good and this patient does not present any other downstream adverse vascular events, conservative treatment with periodical follow-up was given to her FMD lesions.

The final control angiography demonstrated complete exclusion of the aneurysm, with normal filling of the PCoA and PCA. The postoperative course was uneventful and she was discharged to home from our hospital 15 days later, with a normal neurological exam. Nine months post procedurally, CTA of the cranial arteries performed with a 320-row, multidetector computed tomographic (MDCT) scanners showed that complete obliteration of the aneurysm was achieved with the PCoA fully patent (Figure 2d).



(a) 3D –DSA showing the true PCoA aneurysm and its relation to adjacent vascular structures. (b) and (c) Angiography after occlusion of the aneurysm. (d) Nine months postprocedurally, 320-row 3D reconstructions of CTA showing complete obliteration of the aneurysm and preservation of the parent artery. (arrow: "string of beads". arrow head: the true PCoA aneurysm).

Figure 2: The angiogram indicated that the left middle cerebral artery was filled via the left PCoA.

Discussion

FMD is an idiopathic, segmental, nonatheromatous, non-inflammatory vascular disease of the musculature of arterial walls, leading to stenosis of small and medium-sized arteries, and has been described in almost every arterial bed in the body [1]. Cervical and intracranial FMD may cause critical arterial stenosis, spontaneous dissection, aneurysm, and embolism, but the majority of patients are likely asymptomatic and may remain undiagnosed. Despite the fact that countless theories have been proposed, there have only been limited advancements in the knowledge of FMD during the recent 40 years, and the etiology and prevalence of FMD of cervical and intracranial arteries remains unknown [1]. FMD is currently classified into three main types according to the histopathology based on the predominant arterial wall layer involved as follows [4]: intima, media, or adventitia (periarterial), which can be associated in a single patient. (1) The intimal type FMD (about 10% of FMD cases) is characterized by irregularly distributed mesenchymal cells within a loose matrix corresponding to fibrous circumferential intimal thickening with proliferation of sub endothelial connective tissue. The internal elastic lamina is always preserved whit fragment, and the media and adventitia are normal. It appears as focal truncal stenosis. (2) The medial type FMD is the most frequent (accounted for 60% to 70% of FMD in initial reports and >90% today) and consists of a rarefaction of smooth media muscle cells replaced by fibrosis. The intima, internal elastic lamina and adventitia are normal. It appears as a succession of dilatations and multifocal stenoses interspersed with aneurysmal segments, with a preserved, sometimes fragmented, internal elastic lamina corresponding to a characteristic string-of-beads aspect. (3) The adventitial or perimedial type (approximately <1% of FMD in adults) involves excessive elastic tissue in the external area of the media or hypertrophy of the connective tissue at the junction of the media and adventitia. Its aspect is close to the medial type with fewer dilatations that do not exceed the diameter of the normal artery. Stenosis often appears tubular. However, these categories are not mutually exclusive, as involvement of more than one layer in the same diseased artery is not uncommon. However, few dysplasias are currently examined histologically, so an angiographic diagnostic classification has been suggested based on differentiation between unifocal and multifocal appearance. Angiographically, FMD has also been recognized as three main types: (1) multifocal ('string-of-beads' appearance), (2) unifocal (solitary stenosis < 1 cm in length) and (3) tubular (stenosis at least 1 cm in length) [3,5]. Intimal fibroplasia is characterized by a focal, band-like, concentric stenosis (unifocal with solitary stenosis < 1 cm in length) (Figure 1d and Figure 1e), the lesion of medial fibroplasia resembles a "string of beads" appearance (multifocal) (Figure 1f and Figure 2c), adventitial fibroplasia appears as tubular stenosis (stenosis at least 1 cm in length) which is extremely rare type of all FMD lesions (Figure 1b and Figure 1c), seen in less than 1% of arterial stenosis [1]. Importantly, when stenosis occurs, occlusion is rarely complete. The diagnosis of FMD can be established by histopathology or angiography [5,6].

Our case is very uncommon, in which three main types of FMD coexist in the bilateral carotid and left vertebral arteries, with a true aneurysm of the left PCoA. To the best of our knowledge, we present the first report of three main types of FMD coexistence in a single patient, and there have been no literature reports of true PCoA aneurysm associated with FMD. An aneurysm originates from the PCoA itself that is known as a "true" PCoA aneurysm [7,8], with a few millimeters away from the junction of the ICA or PCA, which is rarely encountered, comprising 1.3% of all intracranial aneurysms and 6.8% of all PCoA aneurysms [8]. Although the mechanisms of aneurysm

growth and rupture remain controversial, it has been generally considered that hemodynamic factors play a significant role in the initial formation, the growth and the rupture of cerebral aneurysm. Kaspera et al. reported a"true" PCoA aneurysm coexisting with the internal carotid occlusion and demonstrated increased velocity and turbulent blood flow in both communicating arteries, which suggested that the blood flow disturbances might be conducive to the formation and development of the aneurysm [9]. Ogasawara et al. reported a case of ruptured true PCoA aneurysm 13 years after surgical occlusion of the ipsilateral cervical ICA with superficial temporal artery-middle cerebral artery anastomosis for ruptured right ICA aneurysm and suggested that hemodynamic factors were the reason for aneurysm formation [10]. These authors suggested that hemodynamic factors might play an important role in the development of "true" PCoA aneurysm associated with occlusion of major vessels. Our case suggests that the increased velocity and the blood flow disturbance resulting from the vertebrobasilar system through the PCoA to maintain nearly entire ICA system cerebral circulation might be a conductive factor causing the formation and development of the aneurysm. Although fibro muscular dysplasia of the carotid or vertebral arteries may result in a high prevalence of intracranial aneurysms and true PCoA aneurysms associated with increased blood flow through the PCoA have been reported [8], what we present here differs from those previously reported in that the current case suggested that hemodynamic factors related to critical stenosis of major arteries caused by cervico-cephalic FMD might play a vital role in the formation and development of "true" PCoA aneurysm.

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