Endovascular treatment of basilar artery stenosis due to cerebral vasculopathy related to neurofibromatosis (NF1)

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Abstract

Background—Cerebrovascular lesions are uncommon in neurofibromatosis type 1 (NF1).

Case Description—We report a case of 34-year-old man with NF1 who developed posterior circulation stroke. Diffusion-weighted imaging showed acute infarcts in the right vertebra basilar artery territory. Digital subtraction angiography demonstrated significant stenosis of the basilar artery in the mid segment that was identified as the etiology of the symptoms. The vertebral arteries were tortuous and the basilar artery was ectatic. Subsequently, endeavor resolute stent was placed across the lesion and post-procedure angiogram showed resolution of stenosis.

Conclusion—Selective stenotic involvement of the basilar artery with ectatic vertebrobasilar circulation associated with NF1, which was successfully treated with endovascular method, was not been reported previously to our knowledge.

Keywords
basilar artery; neurofibromatosis; stent; stroke

Introduction

Neurofibromatosis type 1 (NF1) is a genetic disorder that produces a broad spectrum of clinical manifestations as a result of abnormal growth of neuroectodermal tumors throughout the body. It is a genetic disorder, caused by a mutation in the NF1 gene on chromosome 17 [1]. The most common manifestation of NF1 vasculopathy is renal artery dysplasia and hypertension. Cerebrovascular abnormalities in NF1 are uncommon manifestations. Neurofibromin, the protein product of the NF1 gene, expressed in endothelial and smooth muscle cells of blood vessels [2], is lost in NF1 which increases the proliferation of vascular smooth muscle cells with subsequent intimal proliferation and arterial stenosis. We report a case of posterior circulation ischemic stroke caused by basilar artery stenosis in an Asian man with NF1, treated successfully with percutaneous transluminal angioplasty and stenting. This is an unusual clinical presentation in NF1 with only involvement of the basilar artery and no other vascular involvement.

Case report

A 34-year-old man with no previous history of any medical illness presented with recurrent symptoms of ataxia, dizziness, and imbalance from past 3 months. The frequency of these symptoms was increasing despite the continuous medical treatment (Plavix 75 mg and Ecosprin 75 mg). On clinical examination, he has multiple neurofibromas (Figure 1) predominantly involving the joints. Magnetic resonance imaging (MRI) of the brain showed foci of restricted diffusion in the right vertebrobasilar artery territory suggestive of acute infarcts. Three-dimensional (3-D) time-of-flight MR angiography showed ectatic basilar with stenosis in the mid basilar segment (Figure 2). Digital subtraction angiography done subsequently showed tortuous vertebral arteries and the ectatic basilar artery with significant flow limit-
ing stenosis in the mid segment (Figure 3A). In view of the increasing frequency of the symptoms despite the optimum medical management, endovascular treatment was planned.

Under general anesthesia, right common femoral was accessed. Standard heparinization was given to maintain the activated clotting time of ~250 s. Guide catheter (5F Envoy, cordis) was placed in the left vertebral artery (with Nimodipine infusion). The stenotic segment of the ectatic basilar artery was crossed using a 0.014 micro-wire. Subsequently, drug eluting stent (Endeavour) was placed across the lesion (Figure 3B). Post-stent angiogram showed resolution of the stenosis with good ante grade flow across the treated segment (Figure 3C and D).

The intra- and periprocedural periods were uneventful. The patient did not have any further episodes of transient ischemic attacks and was discharged on dual antiplatelet (Ecospirin 75 mg and Plavix 75 mg). On 1-month clinical followup, neurological examination was normal.

**Discussion**

NF1 present frequently with “cafe au lait” spots and neurofibromas. Vasculopathy in NF1 commonly involves the renal arteries but cerebrovascular involvement can occur.

Neurovascular abnormalities in NF1 are rare. The various abnormalities in NF1 include steno-occlusive disease, arteriovenous fistula, and aneurysm formation [3]. Mitsui et al reported a case of the basilar artery fusiform aneurysm manifesting as Wallenberg’s syndrome in a patient with NF1 [4]. In our present case, the vertebrobasilar circulation was ectatic and the patient did not have any other risk factors for atherosclerosis. Ectasia of the vertebrobasilar artery system was defined as arterial diameter 4.5 mm in any location along its course [5]. The cerebrovascular alterations in NF1 are usually asymptomatic, with few cases of ischemic stroke being reported [6]. The most common form of cerebral vasculopathy in NF1 patients is occlusive disease, usually occurring in childhood or adolescence and often associ-
Table 1. Review of previously reported cases of NF1 with cerebrovascular disease

<table>
<thead>
<tr>
<th>No.</th>
<th>Study</th>
<th>Demographics (M: male; F: female)</th>
<th>Presentation</th>
<th>Cerebrovascular Manifestations</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>Partha et al [14]</td>
<td>7 F and 8 M in pediatric age group</td>
<td>No neurological deficit</td>
<td>Moyamoya pattern in seven cases and occlusion of major arteries in eight cases.</td>
</tr>
<tr>
<td>5</td>
<td>Pereira et al [11]</td>
<td>45 M</td>
<td>Cervical bruit, retroauricular pain, and progressive paraparesis</td>
<td>Vertebrovertebral arteriovenous fistula</td>
</tr>
<tr>
<td>7</td>
<td>Pereira et al [11]</td>
<td>14 F</td>
<td>Radiculopathy</td>
<td>Right vertebral aneurysm at the C5–C6 level</td>
</tr>
<tr>
<td>8</td>
<td>Miyazaki et al [12]</td>
<td>52 F</td>
<td>Left vertebral artery aneurysm</td>
<td>Left vertebral artery aneurysm</td>
</tr>
<tr>
<td>9</td>
<td>Myung Won You et al [13]</td>
<td>16 M</td>
<td>Visual disturbance for 1 year</td>
<td>Fusiform aneurysms from bilateral extracranial internal carotid arteries.</td>
</tr>
<tr>
<td>10</td>
<td>Present case</td>
<td>35 M</td>
<td>Posterior circulation stroke</td>
<td>Fusiform aneurysms from bilateral extracranial internal carotid arteries.</td>
</tr>
</tbody>
</table>

We did not find any reports of treatment of intracranial arterial stenotic disease associated with NF1. The present case report is unique because of the unusual clinical manifestation in middle-aged man with NF1, the ectatic basilar artery with segmental flow limiting stenosis which was successfully treated with endovascular treatment.

References


