Cerebral Hyperperfusion Syndrome following Staged Bilateral Internal Carotid Artery Stenting

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Abstract

Cerebral Hyperperfusion Syndrome is a relatively rare event following carotid revascularization. It can occur after both carotid endarterectomy and carotid artery stenting. It is characterized by focal neurodeficit, seizures and headache in the absence of ischemia. It occurs due to ipsilateral cerebral edema secondary to hyperperfusion. CT and MRI of the brain are the main modalities used for diagnosis and to rule out infarct. Prompt recognition and treatment can prevent permanent injury to the brain. We present a case of cerebral hyperperfusion syndrome in an elderly gentleman after a staged bilateral internal carotid artery stenting.

Introduction

Although carotid endarterectomy (CEA) has remained a standard therapy for stroke prevention in patients with significant carotid artery stenosis, carotid artery stenting (CAS) has emerged as an alternative.

Neurological complications following carotid revascularisation with either technique are well known, of which ischemic injury to the brain, due to embolisation or carotid occlusion are the most common. Cerebral hyperperfusion syndrome (CHS) is a relatively rare, but potentially devastating event known to complicate carotid revascularisation. It is defined as a clinical triad of ipsilateral headache, seizures and focal neurological deficits occurring in the absence cerebral ischemia. Awareness of this entity enables early detection and initiation of appropriate therapy to limit brain injury.

Case Report

A 72 year old gentleman, diabetic and hypertensive presented with bilateral critical internal carotid artery (ICA) stenosis of more than 70%. He had recurrent episodes of transient ischemic attacks (TIA) involving the left upper and lower limbs in the preceding 2 months. Stenting of the symptomatic artery was planned, to be followed by the contralateral vessel after four weeks. He underwent angioplasty and stenting to right ICA on 19/2/2016 using 9-7 x 40 mm X-ACT stent with a distal protection filter, which showed a significant amount of debris as shown in figures 1 and 2. The post procedure period was uneventful and he was discharged 2 days later. He remained asymptomatic, with no further episodes of TIA. He was readmitted one month later and underwent left ICA stenting on 18/3/2016 with 9-7 x 40 mm X-ACT stent, the retrieved filter showing no debris (Figure 3). His blood pressure (BP) in the immediate post procedure period was normal. 2 hours later patient became delirious and developed sensory aphasia, there was a decline in his BP, with systolic BP measuring 70 mmHg. Electrolytes were normal. CT brain and angiogram of neck and intracranial vessels was done which showed evidence of cerebral oedema involving the left hemisphere, with patent carotid stents and normal intracranial vessels (Figure 4). Patient was started on IV steroids. Over the next 12 hours patient developed right hemiplegia with right sided facial palsy and global aphasia. MRI brain revealed tiny multiple embolic infarcts, which did not correlate with the clinical signs (Figure 5). Cerebral hyperperfusion syndrome was diagnosed. He was started on IV mannitol. On Day 2 following the procedure, patient had two episodes of seizures, managed with anticonvulsants. Throughout the
period patient remained in a state of confusion and irritability. From day 3 onwards he started showing signs of recovery, conscious levels returned to normal and he had complete recovery with no residual neurological deficits.

**Discussion**

Hyperperfusion is defined as a 100% increase in cerebral blood flow (CBF) compared to pre-operative baseline. CHS was first described by Sundt et al as a clinical syndrome complicating CEA. The first report on CHS after CAS was published by Schoser et al. The incidence of this syndrome following CEA, as reported by numerous publications ranges from 0.4 to 14%, whereas following CAS varies between 0.96 to 11.7%.

Impaired cerebral autoregulation seems to play a significant role in the pathogenesis of CHS. The severity is proportional to the duration of carotid occlusion, severity of cerebral hypoperfusion, presence of contralateral carotid occlusion and poor collaterals. Elevated BP in the post-operative period is another factor in the pathogenesis of CHS. It occurs due to baroreceptor reflex failure following CEA. During CAS transient hypotension and bradycardia can occasionally be observed due to stimulation of the Carotid body nerve that can be followed by rebound hypertension. It is important to note that mechanisms that lead to post operative elevation in BP are not completely elucidated and could be multifactorial. However its absence does not confer protective value, as was observed in our patient who developed CHS inspite of normal BP. Intraoperative ischemia, ischemia-reperfusion injury with oxidant production, complement activation and microvascular permeability are other factors known play a role in the pathogenesis.

CHS can develop at any time from immediately after the procedure to up to a month later, but most patients develop symptoms within the first few days. Ogasawa et al reported occurrence of CHS peaking on 6th post op day after CEA and 12 hrs after CAS.

Confusion, deterioration of conscious level and headache are the most common presentations. Focal neurodeficit is secondary to cerebral oedema and in most cases is transient and reversible. The neurodeficit is usually cortical with hemiplegia, neglect, hemianopia, aphasia, as happened in our case. Seizures may be focal or generalized. The most severe complication secondary to hyperperfusion is intracranial haemorrhage (ICH). Its incidence is reported as 0.37% after CEA and 0.74% following CAS.

Transcranial Doppler(TCD) has been used to measure CBF velocity and helps to predict occurrence of CHS, but has its own limitations. It is very crucial to identify CHS early to avoid irreversible brain damage. Clinical suspicion is important. TCD, SPECT, PET, CT and MRI aid in diagnosis. CT reveals ipsilateral sulcal effacement and cerebral oedema, immediately following symptom onset as in our patient. T2W and FLAIR MRI is more precise in demonstrating areas of cerebral oedema and DW-MRI helps to rule out embolic events. Angiography is almost universally normal.

Management of CHS involves aggressive BP control with target BP being 20-30% below baseline in patients with impaired CVR. There is no randomised control trial addressing the optimised treatment protocol for patients with CHS. Measures to treat cerebral oedema like mannitol, hypertonic saline, steroids and hyperventilation have been shown to be effective, anticonvulsants to treat seizures. One should also be aware about possibility of a delayed CHS and instruct patients accordingly at the time of discharge.

Nicolas et al have described that staged bilateral carotid stenting 30 days apart is an effective strategy to avoid CHS in high risk patients. They found that there was no statistically
significant differences with regard to the 30-day and 12-month clinical outcomes in patients undergoing unilateral compared to those receiving staged bilateral CAS.\textsuperscript{12} In our patient, despite taking measures to minimize complications, like staged carotid stenting, using distal protection filter and appropriate BP control, CHS still occurred. However with timely recognition and treatment, patient had a complete neurologic recovery.

**Conclusion**

CHS although rare, is a potentially devastating complication following carotid revascularisation. The occurrence is not always predictable, despite taking precautions. With prompt recognition and treatment of the disorder, most patients make complete recovery. ICH is a serious and at times fatal complication.

**Abbreviations**

BP: Blood pressure; CAS: Carotid artery stenting; CEA: Carotid endarterectomy; CHS: Cerebral hyperperfusion syndrome; CT: Computed tomography; CVR: Cerebrovascular reactivity; DWI: Diffusion-weighted imaging; ICA: Internal carotid artery; ICH: Intracerebral haemorrhage; MRI: Magnetic resonance imaging; SPECT: Single photon emission computed tomography; TCD: Transcranial Doppler.

**References**

7. Adhiyaman V, Alexander S. Cerebral hyperperfusion syndrome following carotid endarterectomy. QJM 2007; 100:239-244.