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Combined Open and Endovascular Stent Grafting of Internal Carotid Artery Fibromuscular Dysplasia: Long Term Results

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Background. *Symptomatic fibromuscular dysplasia (FMD) of the internal carotid artery (ICA) can present as thromboembolic ischemic events, spontaneous or post-traumatic dissection, aneurysmal degeneration or intracranial haemorrhage and needs definitive surgical treatment.*
Patients and methods. *Six patients and nine ICA with FMD were revascularised using a carotid approach with minimal exposure of the common, external and internal carotid arteries for covered stent repair. All patients were female, the age ranged from 30 to 65 years (mean 44).*
Results. *One patient suffered from a perioperative transient neurological deficit. Duplex revealed a patent stent. The patient fully recovered after 5 h, not showing any changes on repeat CT scans. One patient developed a recurrent laryngeal nerve palsy. The symptoms gradually resolved within 1 month. No perioperative strokes or deaths occurred. During a mean follow up of 48 months (range 13–63) no thromboembolic neurological events, graft occlusions or haemodynamically significant stenoses occurred.*
Conclusion. *ICA FMD stent grafting is an alternative to open surgery or percutaneous endovascular intervention with excellent long-term results.*

Keywords: Fibromuscular dysplasia; Internal carotid artery; Hybrid revascularization; Covered stent.

Introduction

Fibromuscular dysplasia (FMD) is one of the most common causes of non-traumatic, non-atherosclerotic, non-inflammatory lesions of the internal carotid artery (ICA) causing embolic events in young patients. The prevalence of FMD is 0.5–0.7% by angiography.[1,2] However, in most cases the angiographic finding of FMD is incidental without associated symptoms, as demonstrated in an autopsy study, where the incidence of ICA FMD was only 0.02%.[3] As the natural history of ICA FMD is generally benign, initial therapy consists of antiplatelet therapy and clinical follow up.[4] Symptomatic FMD of the ICA can present as thromboembolic ischemic events, spontaneous or post-traumatic dissection, aneurysmal degeneration or intracranial haemorrhage.[5] Symptomatic patients need definitive surgical treatment. The most widely used surgical technique for treating critical stenosis is gradual internal dilation (GID) with a reported stroke and death rate of up to 4.2%[6] and a 16.6% cranial nerve injuries rate.[7] During the past two decades, percutaneous[8] or open operative balloon angioplasty[9] have been increasingly used to dilate critical stenoses. Aneurysmal disease is most commonly treated surgically with resection and vein graft interposition.[7] The use of covered stents has been reported with good short-term results[10] in stenotic lesions. Additionally, this technique has the advantage of universal application in stenotic lesions, aneurysms and dissections. Yet, long-term results have not been published so far. Herein, we report our experience of a combined conventional and endovascular covered stent treatment of patients with ICA FMD under reversed flow and their respective long term results.

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Patients and Methods

Patient selection

Symptomatic and asymptomatic patients referred to our department with FMD of the ICA were initially screened with duplex sonography of the carotid and vertebral arteries accompanied by a magnetic resonance angiography (MRA). Digital subtraction angiography (DSA) was performed selectively. In cases of aneurysm formation, a CT scan was obtained to evaluate maximum diameter and length of the aneurysm. After assessing all patients neurologically as well as the morphology of the lesions, antiplatelet therapy was commenced or continued. The indication for operation was continued neurological symptoms despite medical treatment in the presence of high grade stenosis or aneurysm.

Ultrasound follow up

Carotid arteries were assessed according to a routine scheme. All patients had an initial assessment of the CCA, ECA and ICA with 6 and 16 kHz probes. Peak systolic velocities of more than 120 cm/s and end diastolic velocities of more than 40 cm/s and an ICA/CCA index of more than 1.5 were considered haemodynamically significant stenoses. High resolution images of the proximal landing zone of the stent graft as well as the graft itself were recorded.

Patients

From February 1999 to July 2001, six patients and nine ICA with FMD were revascularised using a hybrid conventional/endovascular technique (Table 1). All patients with FMD were female, the age ranged from 30 to 65 years (mean 43.7 years). All patients had neurological symptoms. Three patients were revascularised bilaterally and three had unilateral stent grafting. One patient with FMD had a spontaneous dissection and high-grade stenosis with fresh thrombi with a contra-lateral occlusion of the ICA, four patients had ICA aneurysms and four patients had high-grade stenosis. One patient with bilateral disease (patient #1) underwent repair of the contra-lateral asymptomatic side. The intervention was carried out due to explicit patient wish, even though she had a TIA after repair of the symptomatic side. Of the other eight revascularised ICA, four had recurrent episodes of amaurosis fugax and four had at least one ischemic hemispheric event prior to intervention. The mean length of the diseased vessel (including ICA dissection, stenosis and aneurysms) was 3.1 cm (range 2.1–4.1 cm), the mean diameter of the largest aneurysm within a diseased ICA was 1.9 cm (range 1.6–2.3 cm).

Operative procedure

Briefly, after transverse skin incision of the neck, the common carotid artery (CCA), external carotid artery (ECA) and the ICA were dissected free with minimal manipulation. Subsequently, a reverse blood flow in the ICA was induced by clamping the CCA and ECA. After arteriotomy, an introducer sheath was inserted into the CCA. Under fluoroscopy, a guide wire was advanced through the pathologically altered ICA up to the siphon of the ICA and a 5 cm$\times$6 mm PTFE-HEMOBAHN (WL Gore and Ass, Flagstaff, AZ, USA) endoprosthesis was deployed into the ICA, overlapping the diseased portion of the artery. Subsequently, a 6 mm dilatation balloon was inserted and the graft was moulded to the ICA and blood was drawn from the side port of the introducer in order to aspirate intravascular debris. At the end of the procedure, a digital subtraction angiography with the guide wire in place was performed. Before removing the introducer system, it was flushed retrogradely and the arteriotomy of the CCA was closed with interrupted sutures (Fig. 1). Five patients and eight ICA were operated under general anaesthesia, one patient was operated under loco-regional anaesthesia.

Results

Perioperative results

One patient suffered from a perioperative transient neurological deficit, lasting 5 h. After skin closure, weakness of the upper extremity corresponding with the operated ICA was noted. Duplex revealed an ideally patent prosthesis. The patient fully recovered after 5 h, not showing any changes on repeat CT scans. One patient developed a recurrent laryngeal nerve weakness. The symptoms gradually resolved within 1 month of follow up. No perioperative strokes or deaths were observed.

Follow up

Patients were followed up 1, 3, 6 and 12 months postoperatively. Thereafter, routine follow up with neurological evaluation and duplex sonography was performed once every 12 months. The follow up ranged from 13 to 63 months (mean 48 months). No thromboembolic neurological events, graft occlusions

Table 1. Patient characteristics

Patient	Pathology	Age	Symptoms	Operative morbidity	Clamping time (min)	Follow up (months)
# 1	R: high grade stenosis	37	R: amaurosis fugax, TIA	Right TIA	R: 9	63
	L: high grade stenosis		L: asymptomatic		L: 13	61
# 2	ICA aneurysm	30	Aphasia	RLN palsy (transient)	L: 11	63
# 3	R: high grade stenosis	42	R: amaurosis fugax	None	R: 9	60
	L: ICA aneurysm		L: TIA (aphasia)		L: 12	59
# 4	High grade stenosis/dissection	39	Amaurosis fugax	None	L: 11	52
# 5	ICA aneurysm	49	R: minor stroke L: TIA (aphasia)	None	L: 13	40
# 6	R: ICA aneurysm	65	R: recurrent TIA	None	R: 13	17
	L: high grade stenosis		L: amaurosis fugax		L: 10	13

ICA, internal carotid artery; TIA, transient ischemic attack; RLN, recurrent laryngeal nerve; R, right ICA; L, left ICA.

or haemodynamically significant stenoses were seen during the observation period (Fig. 2).

Discussion

The prevention of stroke in young patients is the aim of surgical therapy of non-atherosclerotic ICA pathologies. This is a challenging task as experience with patients with these pathologies is limited and surgical techniques are more demanding compared to endarterectomy of atherosclerotic ICA lesions.[6,11] Surgical treatment options of symptomatic patients include resection and interposition of the diseased artery, endarterectomy with patch grafting, gradual internal dilatation and open angioplasty.[6–9]

As ICA FMD usually affects the middle and distal portion of the ICA at the level of the first and second cervical vertebrae,[12] graduated internal dilation needs mobilisation of the ICA at least to the distal level of the styloid process. The artery is then coaxially dilated by passing dilators through an arteriotomy. This method was the preferred technique in the past as endarterectomy is rarely the appropriate technique in stenotic and aneurysmal ICA FMD. Sheer forces are applied to the arterial wall, potentially damaging the endothelium severely. However, good long-term results are reported,[6,7] suggesting that FMD artery walls are rather forgiving to mechanical trauma.

Surgical and percutaneous angioplasty appear to be better alternatives with radial forces dilating the stenotic area and less surgical exposure of the carotids.[8,9] However, both methods are only suitable for stenotic lesions, not for dissections, especially with fresh thrombi, and aneurysms.

Revascularisation with covered stents is an appealing option, as all morphological presentations of ICA FMD can be treated with one surgical technique. This seems to be an important consideration in a rare condition, as frequently performing an operation significantly reduces major complications.[13] In our series, we chose a surgical carotid approach to implant a covered stent into the diseased portion of the ICA. For ICA FMD, a polytetrafluorethylene (PTFE) covered stent seems to be an ideal therapeutic option, since the diseased portion of the vessel is in the transition zone of the ICA, where the elastic type of the artery changes into the muscular type.[14] This is usually a few centimetres distal to the carotid bulb. Therefore, a cylindrically shaped graft can be used as no calibre changes are noted in this particular area. Therefore, aneurysm exclusion can be performed equally safe as stenting dissections and stenosis.

A carotid approach was chosen as we believe that the device has to be implanted with retrograde flow of the ICA in order to avoid periprocedural embolisation. This strategy is especially important since all lesions were more than 2 cm long, and five lesions had mixed thrombi adherent. It has been described that during all steps of carotid artery stenting microembolisations occur,[15,16] potentially causing stroke. Also, at the time of treating our first patients, we did not have access to a Parodi balloon-occluding sheath or similar devices. Moreover, micro-catheterisation of the lumen in FMD with or without dissections from the femoral approach is technically demanding. By accessing a healthy portion of the CCA, technical success is easily achievable.

One patient suffered from an intraoperative neurological deficit, affecting the upper extremity corresponding to the operated carotid artery. The symptoms only

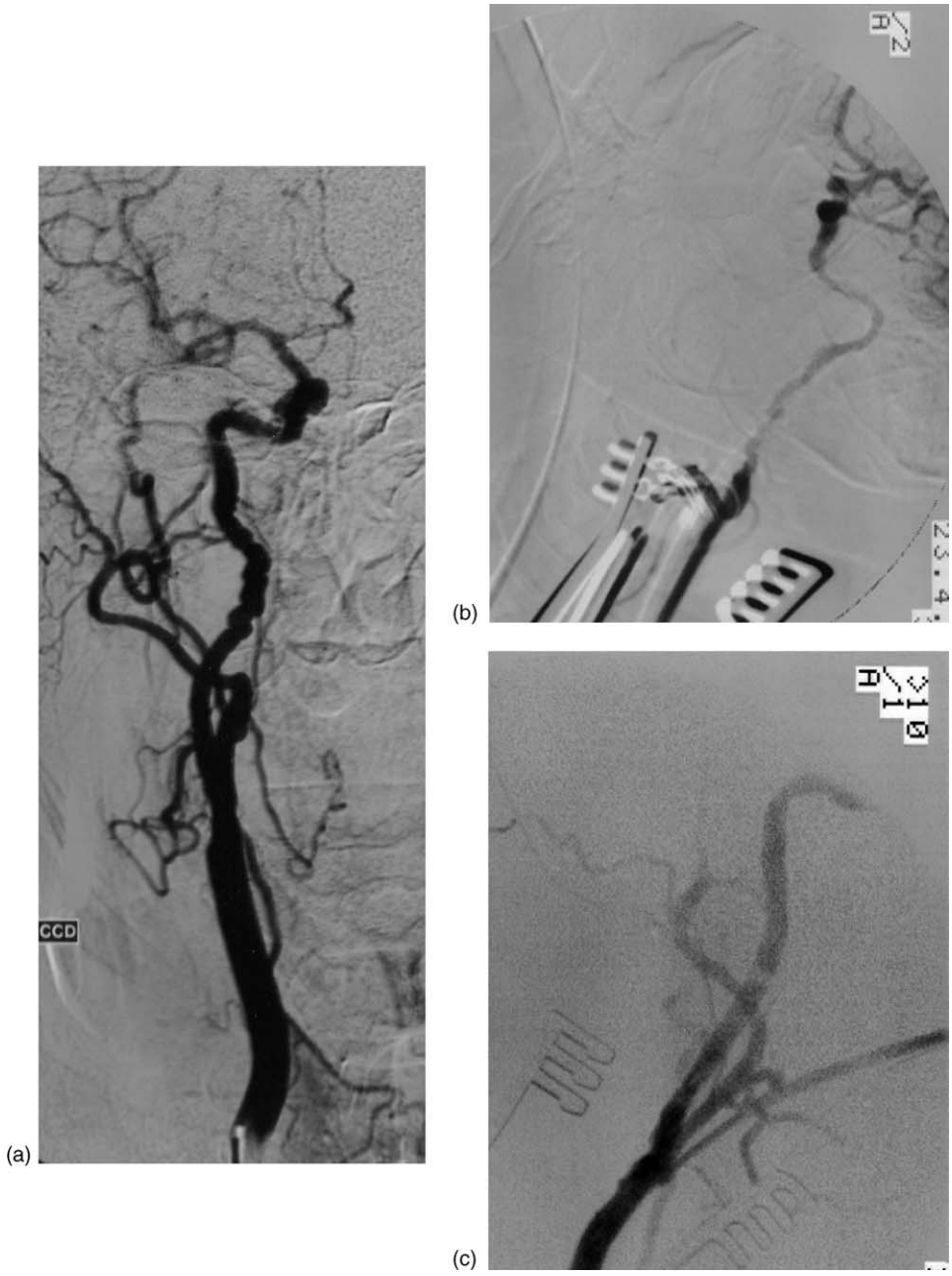

Fig. 1. Angiography. (a) Preoperative angiography. (b) Intraoperative angiography prior to stent deployment. (c) Completion angiography after stent deployment.

lasted a few hours and the patient did not have any CT changes. This patient was the first one operated on with this technique. Also, the contra-lateral artery had a high-grade stenosis also due to FMD. Up to 10% of patients do not tolerate flow reversal,[17] and this may be accentuated in patients with contra-lateral haemodynamically significant stenoses. One patient suffered from transient recurrent laryngeal nerve palsy. One month post-operatively the patient did not have any residual symptoms. This is a rare complication and in our own series of carotid endarterectomy, applying a similar skin incision and much more surgical exposure of the carotid arteries, this morbidity occurred only once in 180 operated arteries.[18]

Long term durability of ICA FMD reconstructions is imperative, especially as the mean age of patients with cerebrovascular FMD is 50 years.[19] The long-term results in our patients treated for ICA FMD with a covered stent are excellent with no symptomatic recurrences or haemodynamically relevant stenoses during follow up.

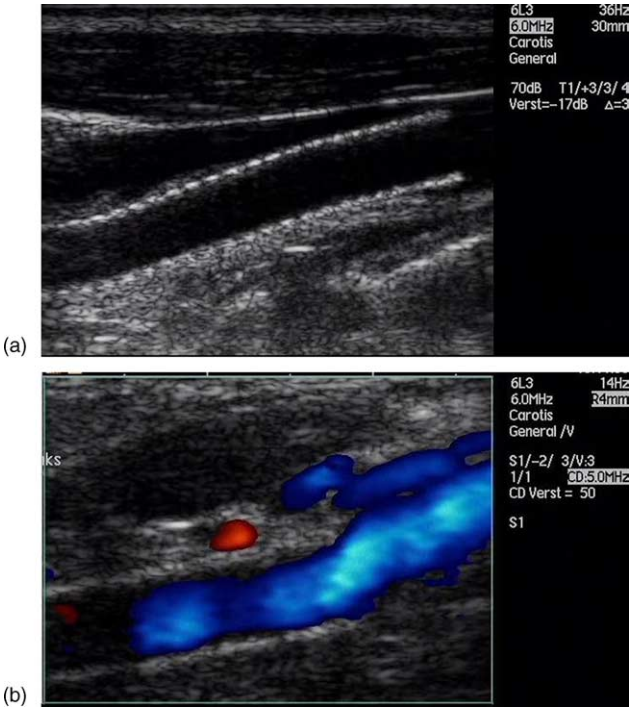

Fig. 2. Five year follow up after stent grafting of the internal carotid artery. (a) b-Mode sonography of the internal carotid artery and the proximal portion of the stent graft. (b) Color coded duplex of the healthy proximal portion of the internal carotid artery and the stented portion.

These results are similar to patients with spontaneous and traumatic dissections of the ICA treated similarly.[20] It appears that non-atherosclerotic, non-inflammatory carotid artery lesions do behave more benignly when stented compared to atherosclerotic ICA lesions.[21]

Our series of six patients and nine operated ICA with an open endovascular procedure with a covered stent does demonstrate an alternative to open surgery and percutaneous endovascular intervention for ICA FMD, with good long term results.

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