CASE REPORT

Carotid Artery Aneurysm in a Child with Tuberous Sclerosis

M. E. D. Jarrett1, G. Libertiny1, S. J. Gould2 and P. Morris1

Nuffield Department of 1Surgery and 2Cellular Pathology, John Radcliffe Hospital, Oxford, U.K.

Introduction

Tuberous sclerosis complex (TSC) is an autosomal dominant disease of variable penetrance characterised by the presence and growth of hamartomas in almost every organ. Dysplasia and hamartomatous proliferation of small blood vessels including haemangiomas are commonplace in TSC but intrinsic disease of medium-sized and large vessels is rare and often fatal.1

To our knowledge this is the first reported case of carotid artery aneurysm in a patient with tuberous sclerosis.

Case Report

An 8-year-old Caucasian boy with a history of mental retardation, seizures and facial adenoma sebaceum was admitted with a right-sided neck lump that had grown in size and become pulsatile over 4 years. TSC had been diagnosed at 3 months of age. MRI suggested a carotid artery aneurysm. This was confirmed on bilateral carotid angiography which showed a 2.5 by 4.2 cm saccular aneurysm at the bifurcation of the right common carotid artery (Fig. 1).

At operation the aneurysm was exposed and after vascular control was obtained, the external carotid artery was ligated and the aneurysm resected (Fig. 2). Mobilisation of the common and internal carotid arteries allowed end-to-end spatulated anastomosis. Intraoperative transcranial Doppler monitoring of the right middle cerebral artery showed that no shunt was required.

The postoperative course was uneventful. Histology showed an aneurysm wall with little normal vascular structure. It was composed almost entirely of dense hyalinised fibrous tissue with focal

Fig. 1. Intra-arterial digital subtraction angiogram of right carotid system, showing saccular aneurysm at carotid bifurcation.
calcification. Only a few residual elastic fibres were present. Biopsies of carotid vessels close to the anastomotic site showed some intimal proliferation with medial fibrosis and marked fragmentation of elastic fibres.

**Discussion**

Large blood vessel aneurysms in TSC are rare. There are eight reported cases of aortic aneurysms associated with the disease.\(^1\,^2\) Seven of the eight cases were in children under the age of 5 years. Fewer than 10 cases of intracranial aneurysms have been reported and most were under 30 years of age. One affected patient has been described as having a vertebral artery aneurysm.\(^1\)

Pathological descriptions of TSC associated aneurysms are scant.\(^1\,^3\) In abdominal aneurysms the similarity to cystic medial degeneration seen in Marfan’s syndrome has been emphasised, suggesting intrinsic collagen weakness as a cause. In this case, the smaller vessel involvement and the replacement of the media by collagen precludes a ready comparison with Marfan’s, although the elastic fragmentation in the adjacent more normal vessels does suggest a similarity.

Few aneurysms of the extracranial carotid artery occur, but an increasing number are being recognised as more cases of carotid vascular disease are studied angiographically. True aneurysms are usually atherosclerotic, although syphilitic and mycotic aneurysms do occur. A smaller proportion are associated with degenerative arterial disease as in this case.\(^4\) The natural history, although infrequently documented, is associated with progressive enlargement and rupture, or cerebral embolisation. Surgery carries the risk of iatrogenically precipitated stroke, however, Winslow noted conservative treatment resulted in a long-term mortality of 71% and suggested that surgical intervention was therefore indicated.\(^5\)

**References**


Accepted 12 September 1997