CASE REPORT

Internal Carotid Artery Aneurysm in Neurofibromatosis: An Emergency Presentation

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Introduction

Neurofibromatosis type 1 (NF1) is associated with vascular involvement in up to 1% of cases but only rarely is aneurysmal dilatation of the extracranial internal carotid artery (ICA) seen. This is the first report of such an aneurysm in the U.K. and the first in the literature to present as an Ear Nose and Throat (ENT) emergency.

Case Report

A 53-year-old man with known NF1 presented to the ENT department with a 2-week history of headaches, malaise and a sore throat which had worsened over the preceding 3 days, producing difficulty in swallowing and speaking. Physical examination revealed the obvious cutaneous features of NF1. There was a large left-sided non-pulsatile neck swelling. Oral examination showed that the uvula was deviated to the right and there was a left parapharyngeal swelling. A clinical diagnosis of quinsy was made and under local anaesthetic, 2 ml of blood were aspirated, after which the patient underwent intubation and ventilation due to a compromised airway.

A computerised tomography (CT) scan of the neck showed a large mixed attenuation mass within which was a large enhancing vascular structure. This was thought to represent an aneurysm of the ICA which was subsequently confirmed on angiography. The intracranial circulation and the rest of the systemic vasculature were normal.

The patient was transferred to a London teaching hospital for surgery, at which the aneurysm was found to be surrounded by infected haematoma. Attempted repair of the aneurysm failed. The ICA was ligated with consequent development of a right hemiparesis. The patient made a good recovery with rehabilitation and was discharged with only mild residual disability 6 weeks later.

Discussion

NF1 associated with vascular abnormalities is generally known as “vascular neurofibromatosis”, with the most commonly affected vessels being the renal vessels, followed by the intracerebral arteries, in which the abnormalities are, in order of frequency: stenosis, arteriovenous fistulae and aneurysm.1 There are five previous instances in the literature2–4 of an extracranial ICA aneurysm being discovered in the context of neurofibromatosis, and one of bilateral common carotid aneurysms.1 These cases presented either during investigation of unrelated symptoms or as pulsatile swellings in the neck. The histology1,2 suggests a different pathogenesis to that of atherosclerotic aneurysms, with weakening of the vessel wall secondary to smooth muscle proliferation and disruption of the elastic network. Malecha et al.1 suggest that this may be the common histopathological abnormality in vascular neurofibromatosis.

In our case, the high origin of the aneurysm above the bifurcation and its friability secondary to the surrounding haematoma made surgical reconstruction...
impossible. At presentation there was no history of trauma and no pulsatility to suggest the true diagnosis. Aneurysm of the ICA is a known rare differential for parapharyngeal swellings in the ENT literature. Infection and rupture of extra-cranial carotid aneurysms after ill-advised needle aspiration has been previously described, although not in the context of neurofibromatosis.

The authors feel that an awareness of the possible existence of associated vascular lesions in patients with neurofibromatosis might prevent misdiagnosis and its potentially life-threatening consequences in future.

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References