Ruptured Thyrocervical Trunk Aneurysm in a Patient with Type I Neurofibromatosis

W. Al-Jundi*, S. Matheiken, S. Abdel-Rehim, P. Diwakar, R. Insall

East Kent Hospitals University NHS Foundation Trust, Ethelbert Road, Canterbury, Kent, CT1 3NG, UK

Submitted 8 September 2010; accepted 19 December 2010

KEYWORDS
Thyrocervical trunk aneurysm; Von Recklinghausen; Neurofibromatosis

Abstract
Vascular abnormalities are rare but recognised manifestation of type I neurofibromatosis. Only a few cases of thyrocervical trunk aneurysms have been reported in the literature in which rupture presented as neck swelling or haemothorax. We present an exceptional case of a life-threatening neck swelling in a patient with type I neurofibromatosis who previously had radical reconstructive neck surgery. An emergency angiogram revealed ruptured thyrocervical trunk aneurysm. Due to the complicated soft tissue condition, interventional management was indicated and the bleeding was treated successfully with endovascular coil embolisation. Subsequently, the neck haematoma was surgically evacuated.

Introduction
Neurofibromatosis type I (NF-I), or von Recklinghausen disease, is an autosomal dominant disorder affecting one in 3000 individuals. Cardinal features of NF-I include multiple café-au-lait macules, benign neurofibromas, and iris hamartomas. Arterial involvement in neurofibromatosis is a known but infrequent occurrence. Stenotic lesions predominate, while aneurysms are less common. Aneurysms involving the branches of subclavian artery have been rarely reported. Rupture of a thyrocervical aneurysm causing neck swelling has been reported only once, while another case reported presentation with haemothorax. We submit an unusual case of a ruptured thyrocervical trunk aneurysm in a patient with NF-I, treated successfully with coil embolisation and subsequent surgical exploration.

Case report
A 40-year-old woman presented to the Emergency Department via a 999 ambulance with painful, acute, spontaneous left sided neck swelling. She had difficulty in talking and swallowing, and had tracheal deviation with a tense fullness of the left side of her neck from the clavicle up to the jaw.

Fifteen years prior to this admission, the patient underwent excision of a large left mandibular neurofibroma, followed by musculocutaneous pedicle flap transfer onto the anterior left aspect of the neck.
To protect her compressed airway, the patient was intubated with rapid sequence induction; a bougie was required. While she was tachycardic at 120/min, her blood pressure was 115/65. A haemoglobin value of 5.8 g/dl suggested the cause of the acute swelling to be massive bleeding. Blood transfusion and inotropic support with noradrenaline were commenced. Computed tomography (CT) scan revealed a large haematoma in the left posterior triangle with active extravasation from a branch of the subclavian artery (Fig. 1).

Figure 1 CT scan of the neck demonstrating a large haemorrhage in the left posterior triangle with compression on the left common carotid artery (CCA) and bleeding from the left inferior thyroid artery aneurysm (ITA).

Transfemoral arch angiography was undertaken using a French introducer sheath, a French Pigtail catheter (Cook Medical) and a 3 mm J – wire (Cordis). Selective cannulation of the left subclavian artery and its branches localised the active bleeding to a ruptured aneurysm from the inferior thyroid branch of the thyrocervical trunk (Fig. 2a). A Progreat microcatheter (Terumo Medical Corp., Somerset, NJ) was advanced into the aneurysm, which, together with the inferior thyroid artery, was successfully embolised with coils (4 \times 4 \text{ mm} / 3.7 \text{ mm} and 2 \times 5 \text{ mm} / 5.5 \text{ mm} Vortx microcoils, Boston Scientific). These coils were all introduced via the Progreat catheter and a 177 cm \times 0.016\text{in} (0.41 \text{ mm}) Coil Pusher (Boston Scientific). Technical success was confirmed on completion angiography (Fig. 2b).

The following day the patient continued to be sedated and ventilated on Intensive Care, but her inotrope requirements remained high. Serial troponins returned as 0.67 and 1.49 at 12 hourly intervals each. Despite the suggestion of cardiac injury, concern regarding ongoing bleeding in the neck as a cause for her instability with the need to decompress her airway resulted in an exploration to evacuate the large haematoma (1400 ml) 48 hours after admission. Extensive abnormal venous plexuses in the depth of the flap tissue were found also to be bleeding, presumably torn by the rapidly expanding haematoma. Haemostatic measures, culminating in suture ligation and division of the left internal jugular vein and several of the sinusoidal abnormal veins were required.

The patient dramatically improved after the second procedure, and in quick succession was extubated next day and weaned off inotropes. She was discharged to the ward on the 6th day of admission, and allowed home on day 21.

**Discussion**

Neurofibromatosis was first described by von Recklinghausen in 1821.\(^4\) Types I and II neurofibromatosis are the most common varieties of this disease. Depending on the gene location, type I (chromosome 17q11.2) may be distinguished from type II (22q12). Patients with type I neurofibromatosis may have various cutaneous findings such as café-au-lait spots, benign neurofibromas, and iris hamartomas.\(^5\) Arterial involvement has also been noted, usually in the form of stenoses and most frequently of the renal artery. Arterial aneurysms have been less commonly described.

A review of the English-language literature from 1957 to 2005 yielded 237 patients with NF-I who had 320 vascular abnormalities.\(^6\) Renal artery lesions were most common (41%), were unilateral in 68%, and were more often stenotic than aneurysmal. The carotid, vertebral, or cerebral artery lesions seen in 19% of patients were commonly aneurysms, occurred in the third decade of life, and occurred more often in women (72%). Abdominal aortic coarctation or aneurysms, or without renal and mesenteric involvement, have been reported in 12%.

Our patient presented with life-threatening neck haematoma secondary to a ruptured thyrocervical aneurysm. Ishizu et al.\(^7\) reported the only similar case in which a patient with NF-I presented with neck swelling due to rupture of a thyrocervical branch. There have been reported cases of haemothorax in patients with neurofibromatosis due to ruptured intercostal artery aneurysm or aneurysmal branch of the subclavian artery.\(^8\) The fact that our patient had skin flaps on the same side could have caused the unusual presentation of neck haematoma rather than haemothorax. However, the previous reconstructive surgery made interpretation of the angiography challenging. Therefore, it was difficult to confirm whether the ruptured vascular abnormality was a true aneurysm due to NF-I or a pseudoaneurysm secondary to the previous reconstructive surgery.

This patient presented us with a number of quandaries. Our initial management by endovascular means obviated the need for arterial control in a difficult zone of a neck with abnormal anatomy and complex previous surgery. Endovascular embolisation was performed, however, it was not possible to pass the microcatheter completely beyond the aneurysm into the distal artery. Given the patient was unstable, time was of the essence. Fortunately, there was no retrograde flow to fill the aneurysm but every attempt was made to embolise as distally as possible.

The change in cardiac enzymes and ongoing haemodynamic instability posed a 'chicken and egg' challenge in determining whether ongoing bleeding was responsible for the inotrope requirements, or whether open drainage of the tense haematoma would further stress her overtly impaired cardiac function. The CT scan appearance (Fig. 1)
has also demonstrated compression of the left common carotid artery, which could also be a potential cause for inotrope dependence.

The surgical access for decompression was complicated by the neck flaps, and the procedure was eventually carried out by vascular and maxillofacial teams in conjunction though not without further difficulty in controlling bleeding from the abnormal veins. Therefore, our case has demonstrated that despite the increasing use of endovascular embolisation in the treatment of acute bleeding, in some instances a combination of open surgical and endovascular methods are indicated.

Conflict of interest/funding

None.

References