CASE REPORT

Intravascular embolisation and surgical resection of a giant neurofibroma with intratumoural haemorrhage

¹R G JONES, MD, FRCR, ²P KIATISEVI, MD, ³D C MORRIS, MD, FRCPC, ³P L MUNK, MD, FRCPC,
⁴P W CLARKSON, MBChB, FRACS, FRCSC and ⁴B A MASRI, MD, FRCSC

¹Department of Radiology, Queen Elizabeth Medical Centre, University Hospital Birmingham, Birmingham, UK, ²Center of Excellence in Orthopaedics, Lerdsin General Hospital, Bangkok, Thailand, ³Department of Radiology, University of British Columbia and Vancouver General Hospital, Vancouver, BC, Canada, and ⁴Department of Orthopaedics, University of British Columbia, Vancouver, BC, Canada

ABSTRACT. The use of pre-operative embolisation has been described for small neurofibromas, but not for giant lesions. Advances in embolisation techniques are extending the indications for this procedure, in particular to assist with operative intervention on a range of lesions. This case report describes a 45-year-old male with a giant neurofibroma who underwent embolisation to stabilise intratumoural haemorrhage and to assist with haemostasis during the subsequent surgical resection. Minimal transfusion was required and the patient has made a good recovery. This case demonstrates that pre-operative embolisation of these large and challenging lesions is technically feasible and appears to be beneficial in reducing perioperative blood loss and morbidity.

Received 4 August 2009
Revised 29 October 2009
Accepted 10 November 2009

DOI: 10.1259/bjr/42783229

© 2010 The British Institute of
Radiology

Neurofibromas are common benign soft-tissue tumours arising within peripheral nerves. Solitary neurofibromas are more common, whereas multiple neurofibromas are often associated with neurofibromatosis type 1 (NF-1) [1]. Plexiform neurofibromas rarely grow to be larger than 5 cm; however, giant neurofibromas can occur and are commonly associated with NF-1. Neurofibromas are difficult to manage surgically as they are extensively infiltrative and highly vascular. Complications of neurofibromas are rare, but include malignant change and potentially life-threatening haemorrhage [2].

We report a case of haemorrhage into a giant plexiform neurofibroma of the lower back and buttock causing hypovolaemic shock. This was subsequently managed with intravascular embolisation followed by surgical resection, demonstrating that this technique is technically feasible and should be considered for these lesions.

Case report

An otherwise well 45-year-old Caucasian man presented with bleeding from a solitary large mass over the posterior aspect of his right buttock and lumbar region. Although he initially denied it, he later revealed the mass had been present for over 10 years and had gradually enlarged during this time. Recently, the lesion had

increased in size rather rapidly and suffered an overlying skin breakdown. He was bleeding from the surface of the lesion and became hypotensive with some renal impairment, which later recovered. He received a transfusion of 12 units of blood during stabilisation prior to transfer to our centre.

Upon arriving at our centre, examination revealed a large, tender, soft mass measuring 70 × 50 × 20 cm extending from the thoracolumbar region to the gluteal fold and overhanging the posterior aspect of his thigh. A large café-au-lait spot covered the lesion (Figure 1). He was haemodynamically stable and the lesion was no longer bleeding from the surface.

A CT scan revealed a large soft-tissue mass arising from the subcutaneous tissues of the lower back and extending outside of the available field of view. Loss of fat planes between the mass and the erector spinae muscle was suggestive of possible deep muscular invasion (Figure 2a).

MRI was technically difficult and limited by motion artefact; much of the gluteal mass was not included on the field of view. The mass exhibited intermediate signal on both T_1 and T_2 weighted images and several fluid collections were identified; one of these demonstrated a large fluid–fluid level compatible with haematoma. The gradient-echo images demonstrated multiple serpiginous flow voids in keeping with prominent vasculature. There was no convincing invasion of the underlying gluteal musculature (Figure 2b). Core needle biopsy was performed and the results were consistent with diffuse neurofibroma; there was no evidence of malignancy within the tissue obtained.

Address correspondence to: Dr Paul Clarkson, Department of Orthopaedics, University of British Columbia, 600 West 10th Avenue, Vancouver, BC, Canada, V5Z 4E6. E-mail: pclarkson@bccancer.bc.ca

Figure 1. Clinical photograph of patient's mass on arrival at our centre.

Following initial stabilisation the patient was reviewed at a multidisciplinary conference. The recommendation was for pre-operative embolisation, as this would potentially reduce the perioperative blood loss considering the vascularity and size of the lesion. Surgical resection of the bulk of the lesion would then be undertaken and closure achieved with local tissue advancement or free tissue transfer. It was not feasible to widely excise the lesion as it was infiltrating almost circumferentially around the patient.

Thorough diagnostic angiography was initially carried out under general anaesthesia via a left common femoral artery puncture. A single global diagnostic angiogram covering the entire lesion could not be obtained owing to its overall magnitude. Selective angiography demonstrated significant arterial supply to the lesion from a large right hypertrophied L3 lumbar artery (Figure 3a), as well as major branches of both internal iliac arteries. These were principally the right superior gluteal artery

and lateral sacral branches of the left internal iliac artery. The most abundant supply was shown to arise from the right superior gluteal artery and its branches (Figure 3b), which is the major posterior division of the internal iliac artery. No significant tumour blush was identified; however, all feeding vessels were tortuous and hypertrophied, indicating a rich blood supply.

The right superior gluteal artery, right L3 lumbar artery and a single left lateral sacral arterial branch were successfully embolised. This was carried out after safe positioning of a co-axial catheter/microcatheter system (Renegade™ HI FLO; Boston Scientific, Natick, MA) and multiple embolisation coils (Cook Nester and Micro-Nester embolisation coils; Cook Medical Inc., Bloomington, IN) were used (Figure 3c). Coil placement in the peripheral aspect of the feeding vessels within the lesion itself was avoided so that coil position did not interfere with surgical excision planes.

The patient was transferred to the operating room for surgical resection under the same anaesthetic. Positioning him prone on the operating table we were able to manipulate the lesion satisfactorily. Dissection through the superficial tissues necessitated some dissection through tumour tissue, which was highly vascular with large friable vessels; this stage was responsible for most of the blood loss. Once deep to the lesion it was possible to roll it and dissect it off the deep structures through a normal fat plane, tying off the main feeding vessels that were demonstrated by angiography. Local skin flaps were extensively mobilised and primary closure obtained, albeit under some tension. Four units of packed red cells were transfused intra-operatively.

The specimen weighed 15 kg and measured 65 × 51 × 10 cm. Grossly there was diffuse presence of a myxoid-appearing white tan tissue, infiltrating into the surrounding fat. There were two intratumoural haematomas, the larger measuring 20 × 13 × 5 cm. Histological

(a)

(b)

Figure 2. (a) Single representative axial CT image demonstrating a large soft-tissue mass arising from the subcutaneous tissues of the lower back, extending outside of the field of view. Note the loss of fat planes between the mass and the erector spinae musculature, suggestive of deep muscle invasion. (Intratumoural fluid collection not shown on this image.) (b) Single axial T_2 weighted MR image demonstrating the large soft tissue mass; multiple flow voids represent hypertrophied vessels (arrow head). Also shown is a focal fluid collection, in keeping with an area of intratumoural haemorrhage (arrow).

(a)

(b)

(c)

Figure 3. (a) Representative unsubtracted selective diagnostic angiogram of the right L3 artery demonstrating markedly abnormal and hypertrophied arterial supply to the lesion. A global image demonstrating the lesion's entire supply was unobtainable owing to its sheer magnitude. (b) Representative diagnostic digital subtraction angiogram of the right superior gluteal artery, which was a dominant feeding vessel. (c) Post-embolisation digital subtraction angiogram of superior gluteal artery, demonstrating satisfactory therapeutic occlusion with multiple embolisation coils.

examination revealed a diffuse neurofibroma with no evidence of malignancy.

Post-operatively the patient had central areas of skin breakdown that were managed with vacuum-assisted closure dressings (VAC; Kinetic Concepts Inc., San Antonio, TX) and eventually went on to heal. 6 months post-operatively the patient was mobilising independently with stable skin closure and considerable improvement in his appearance and mobility (Figure 4).

Discussion

To our knowledge this is the first report of intravascular embolisation in a giant neurofibroma. Although life-threatening intratumoural haemorrhage in neurofibromas is uncommon, it has been reported [3–9]. It is postulated that these haemorrhages are caused by rupture of friable vasculature secondary to arterial dysplasia or vascular invasion by the neurofibroma [9, 10].

Figure 4. Post-operative appearance (6 months).

Biopsy was performed owing to the recent size increase in the neurofibroma and because biopsy is recommended to differentiate a haemorrhagic complication from malignant transformation [11]. However, sampling issues are very problematic in such large tumours. Advanced imaging such as MRI and positron emission tomography (PET) CT can be helpful to localise areas of malignant degeneration.

An intralesional procedure was the only possible resection margin because of the almost circumferential nature of the tumour around the patient's body. Surgical experience with giant plexiform neurofibromas is limited to case reports [12–16]. After multidisciplinary discussion we felt that excision of the majority of the tumour followed by local advancement closure would obtain improved function and mobility and remove the friable and haemorrhagic areas of the tumour.

The most immediate challenge for surgical management is haemostasis, especially when the dissection is intralesional. In one report, the first operation was abandoned owing to life-threatening intra-operative haemorrhage prior to complete excision of the mass [15]. Diathermy is of limited use as the tissue is very friable [9]. A number of authors have reported significant blood loss during surgery requiring high-volume transfusion; several management strategies, including argon beam coagulation, have been suggested [9].

Technical advancements in embolisation techniques, such as microcatheter systems, have increased the potential for safe and effective treatment of life-threatening haemorrhage with an endovascular approach [17]. In addition to managing acute haemorrhage, the application of embolisation can be extended to nearly every organ system with a vascular abnormality or tumour [18], either as a stand-alone and definitive treatment or as an adjunct to surgery [19].

Pre-operative coil embolisation was used to reduce the blood flow to the tumour and to limit intra-operative blood loss [7, 20, 21]. The vessels were embolised proximally to reduce flow in the major vascular pedicles at the level of surgical dissection and to reduce skin necrosis associated with a distal embolisation technique. Tanaka et al [20] embolised a plexiform neurofibroma on the arm, but they used particulate embolic agents. We avoided particulates owing to the sheer size of the lesion and to limit skin necrosis. Littlewood et al [21] embolised a plexiform neurofibroma; however, there was a significant delay before surgery of 1 month and significant perioperative blood loss occurred. In our case, coils were placed proximally in major feeding vessels immediately prior to surgery and only four units of packed red cells were required intra-operatively.

Conclusion

This report of the successful management of a giant plexiform neurofibroma demonstrates that intravascular embolisation of such lesions is not only technically feasible, but can reduce blood loss and morbidity at the time of surgery. Giant plexiform neurofibromas are rare and surgical intervention on these hypervascular lesions can be complicated by life-threatening haemorrhage requiring high-volume blood transfusion; therefore, we recommend that embolisation be performed immediately prior to surgical intervention.

References

1. Skinner HB. Current diagnosis and treatment in orthopedics. 3rd edition. New York McGraw-Hill Professional, 2003.
2. Riccardi V. Neurofibromatosis: Phenotype, natural history and pathogenesis. 2nd edition. Baltimore, MD: Johns Hopkins University Press, 1992.

3. Baek SH, Kim JH, Kim JS, Han SB, Cho JS, Yoon YH, et al. Recurrent massive subcutaneous hemorrhage in neurofibromatosis type 1: a case report. *J Korean Med Sci* 2007; 22:728–30.
4. Morello F, Shah P, Dowling K, Siskin G. A hemorrhagic complication of neurofibromatosis. *J Vasc Interv Radiol* 2001;12:773–4.
5. Nopajaroonsri C, Lurie AA. Venous aneurysm, arterial dysplasia, and near-fatal hemorrhages in neurofibromatosis type 1. *Hum Pathol* 1996;27:982–5.
6. Rao V, Affifi RA, Ghazarian D. Massive subcutaneous hemorrhage in a chest-wall neurofibroma. *Can J Surg* 2000;43:459–60.
7. Tsutsumi M, Kazekawa K, Tanaka A, Ueno Y, Nomoto Y, Nii K, et al. Rapid expansion of benign scalp neurofibroma caused by massive intratumoral hemorrhage — case report. *Neurol Med Chir (Tokyo)* 2002;42:338–40.
8. Tung TC, Chen YR, Chen KT, Chen CT, Bendor-Samuel R. Massive intratumor hemorrhage in facial plexiform neurofibroma. *Head Neck* 1997;19:158–62.
9. White N, Gwanmesia I, Akhtar N, Withey SJ. Severe hemorrhage in neurofibromatoma: a lesson. *Br J Plast Surg* 2004;57:456–7.
10. Poston GJ, Grace PA, Venn G, Spencer J. Recurrent near-fatal hemorrhage in von Recklinghausen's disease. *Br J Clin Pract* 1990;44:755–6.
11. Ferner RE, Gutmann DH. International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis. *Cancer Res* 2002;62:1573–7.
12. Salazar R, Robotti EB, Chin DH, Grossman JA. Giant neurofibromatosis of the chest wall: two patient reports. *Ann Plast Surg* 1998;41:211–14.
13. Laky D, Petraru DM, Ancar V, Kelemen J. Giant neurofibroma of the pectoral region including the breast. Case report. *Rom J Morphol Embryol* 1990;36:213–15.
14. Wylock P CJ. An unusual case of neurofibromatosis. *Eur J Plast Surg* 2005;28:247–51.
15. Nahabedian MY, Rozen SM, Namnoum JD, Vander Kolk CA. Giant plexiform neurofibroma of the back. *Ann Plast Surg* 2000;45:442–5.
16. Topsakal C, Erol FS, Ozercan I, Murat A, Gurates B. Presacral solitary giant neurofibroma without neurofibromatosis type 1 presenting as pelvic mass — case report. *Neurol Med Chir (Tokyo)* 2001;41:620–5.
17. Nicholson AA. Vascular radiology in trauma. *Cardiovasc Intervent Radiol* 2004;27:105–20.
18. Pelage JP, Le Dref O, Mateo J, Soyer P, Jacob D, Kardache M, et al. Life-threatening primary postpartum hemorrhage: treatment with emergency selective arterial embolisation. *Radiology* 1998;208:359–62.
19. Lin PH, Terramani TT, Bush RL, Keane T, Moore R, Lumsden A. Concomitant intraoperative renal artery embolisation and resection of complex renal carcinoma. *J Vasc Surg* 2003;38:446–50.
20. Tanaka J, Kuramochi A, Nishi N, Yuasa M, Heshiki A. Preoperative transarterial embolization enhances the surgical management of diffuse plexiform neurofibroma: a case report. *Cardiovasc Intervent Radiol* 2005;28: 686–8.
21. Littlewood AH, Stilwell JH. The vascular features of plexiform neurofibroma with some observations on the importance of pre-operative angiography and the value of pre-operative intra-arterial embolization. *Br J Plast Surg* 1983;36:501–6.