CASE REPORT

Tumour in a tumour: a well-differentiated spindle cell sarcoma within a forearm lipoma

James C. R. Wadkin,¹ Sebastian Senes,² Scott Evans,³ Jonathan Stevenson³

¹Kingston Hospital NHS Foundation Trust, Kingston upon Thames, UK

²Department of Orthopaedics, Hospital Británico de Buenos Aires, Buenos Aires, Argentina

³Department of Orthopaedic Oncology and Arthroplasty, Royal Orthopaedic Hospital, Birmingham, UK

Correspondence to

Dr James C. R. Wadkin,
james.wadkin1@nhs.net

Accepted 24 December 2018

SUMMARY

Lipomas are common, benign, soft tissue masses in adults. Sarcomatous change within a giant lipoma is a very rare event. We present a case of a woman presenting with a forearm lipoma that subsequently developed a central, well-differentiated, spindle-cell sarcoma within the lipoma. This case underlines the importance of identifying red flags for referral of soft tissue tumours.

BACKGROUND

National and international guidelines for the management of soft tissue sarcomas state that any soft tissue lump exhibiting any of the following three clinical features should be considered malignant until proven otherwise^{1,2}:

1. Increasing in size.
2. Size >5 cm.
3. Painful.

Malignant change within a pre-existing benign lipoma is a very rare event, however, patients who re-present should always be reassessed based on their current clinical status, regardless of a previous diagnosis. An increase in size or the appearance of new symptoms, such as pain or compression, may be an indication of the transformation of a previously benign lipoma to a sarcoma.

A soft tissue mass that is evolving and exhibits any red flag signs should always be referred urgently to a regional sarcoma service. Size matters in sarcomas; the smaller the tumour at diagnosis, the better the prognosis.³ As a useful aide-memoire; if a lump is bigger than a golf ball and growing, then think sarcoma.⁴

CASE PRESENTATION

A 58-year-old right-hand dominant woman had presented to her local hospital in 2012 with a 4-year history of a painless mass arising from the volar

Figure 2 Image of the patient's forearm with soft tissue mass in the volar aspect.

aspect of her right forearm. She had no history of trauma, no skin changes over the mass, no signs of inflammation and no systemic upset. She worked as a cleaner and had no significant medical history.

On examination, there was large painless soft tissue mass in her right forearm that had gradually enlarged over the last 4 years, with no evidence of any distal neurovascular deficit. An MRI scan was performed and revealed a 17 cm x 6 cm homogeneous lipomatous tumour arising deep within the flexor compartment of the forearm (*figure 1*). She was reassured that the mass was not malignant and discharged to be managed conservatively as the lesion underwent full-fat suppression on fluid-sensitive MR sequences.

In 2015, 3 years following her initial presentation, the patient re-presented to her local orthopaedic unit with enlargement of the mass and pain in her right forearm. She was referred to our tertiary sarcoma unit for further investigation, diagnosis and treatment.

Clinical examination revealed similar findings to her previous examination with a soft-tissue swelling in the flexor compartment of the right forearm. There was no regional lymphadenopathy or distal neurovascular deficit, however, the patient now reported pain from the mass (*figure 2*).

INVESTIGATIONS

A repeat MRI demonstrated a lipomatous tumour had increased in size; it now measured 21 x 9 x 6 cm in maximum diameter (an increase in size by approximately 4 x 2 x 3 cm from the previous scan). Most significantly, there was also a new, maximally 5 cm, low signal, non-lipomatous, oval focus in the more distal portion of the high signal lipoma on T1 sequences not previously evident (*figure 3*).

An ultrasound-guided biopsy of the new central lesion revealed a grade II (high-grade) sarcoma within the giant lipoma (*figure 4*). A staging chest CT excluded metastases. A diagnosis of a

Figure 1 MRI scan illustrating the presence of a soft tissue lipoma in the right forearm in (A) T1-weighted sagittal plane, (B) T1-weighted axial and (C) the lesion undergoing full suppression on fat suppression sequence.

© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Wadkin JCR., Senes S, Evans S, et al. *BMJ Case Rep* 2019;**12**:e227533. doi:10.1136/bcr-2018-227533

Figure 3 MRI scan illustrating the presence of a new oval focus of non-fatty tissue within the pre-existing soft tissue mass in the right forearm in (A) sagittal plane and (B) axial.

well-differentiated spindle cell sarcoma within a giant lipoma was made.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included necrosis within a lipoma; acute haemorrhage within a lipoma or dedifferentiation into another lesion within the pre-existing lipoma, that is, a second tumour within the lipoma.

TREATMENT

The patient was discussed in the sarcoma multidisciplinary team meeting. In line with recognised guidelines for deep, high-grade soft tissue sarcomas, the patient underwent surgical excision of the mass with a plan to marginally excise the lipoma, accepting the benign lipoma as the surgical margin from the sarcomatous component. Following representation, investigations including MRI and biopsy were performed within 2 weeks and excision of the mass within the next month.

Henry's volar approach to the forearm was used at surgery. A well-defined pseudo capsule was found and the lipomatous lesion and biopsy tract were excised en-bloc with minimal blood loss (figure 5). The ulnar nerve was visualised during the procedure and was not involved in the tumour mass. Excising the whole of the lipoma was considered to be a safe surgical margin for the malignant sarcoma's central nidus. Due to the high grade of the spindle cell sarcoma, the patient received adjuvant radiotherapy (60 Gy in 30 fractions of intensity-modulated radiotherapy).

OUTCOME AND FOLLOW-UP

In line with international sarcoma surveillance guidelines, post-operative surveillance consists of clinical review for local recurrence and chest radiography at 3 monthly intervals for the first

Figure 4 Histology of (A) the border between the non-fatty oval focus and the lipomatous tissue and (B) the centre of the non-fatty oval focus, illustrating the morphology of a well-differentiated spindle-cell sarcoma within the lipoma on H&E staining.

Figure 5 Image of (A) the excised mass within its capsule and (B) following transection of the mass.

2 years, 6 monthly intervals until year 5 and annually until 10 years. If any clinical or radiographical abnormality is detected, then supplementary MRI or chest CT are indicated.

The patient's postoperative radiotherapy was completed uneventfully. At 2 years following excision of the forearm mass, the patient has full function of the right forearm and hand, with no pain or neurovascular deficit and no evidence of local recurrence or metastasis.

DISCUSSION

Although sarcomatous change within a benign soft tissue tumour is a rare event, any soft tissue mass that changes should be reassessed and investigated. This is highlighted by this case. Sarcoma guidelines state that any soft tissue mass that is increasing in size, >5 cm in size or painful should be considered malignant until proven otherwise.^{1 2} The importance of investigating any new mass in patients with previous sarcoma diagnoses has previously been described.⁵ This patient represented because of new symptoms of pain and more rapid increase in the size of a lesion previously described as painless and enlarging very slowly.

Lipomas are typically slow-growing benign tumours. Although they are usually subcutaneous, they can occur in any tissue plane including intramuscular, however, tumours deep within the fascia account for <1% of all lipomas.⁶ They were historically considered rare in the upper extremity, particularly the hand, however, this concept has changed over time.⁷⁻¹⁰

There are many different types of lipomas that are classified according to their histological characteristics including chondroid, cellular angiolipoma, spindle cell variant or pleomorphic lipomata. Their origin is believed to be from mesenchymal primordial adipocytes, increasing in size as the patient accumulates fat but unaffected by weight loss.¹¹

Transformation of a lipoma into a liposarcoma is a rare event, though liposarcomas are the most common adult soft-tissue sarcoma.¹² They generally arise from subcutaneous tissues or from a pre-existing lipoma and tend to present in the fourth to the sixth decade of life and have a female predilection. The incidence of malignant change in lipomas is unknown but felt to be greater in intramuscular and atypical lipomatous tumours.¹³

MRI remains the imaging modality of choice for the investigation of soft tissue masses, due to high specificity, sensitivity and assessment for features suggestive of malignant transformation.¹¹ These features were not apparent on initial imaging in our case. The definitive diagnosis of a malignant tumour, however, can only be made following the histopathological examination.^{14 15}

The surgical indications for excision include local compression of the surrounding tissues (causing pain, muscle dystrophy and/or neuropathies), aesthetic complaints, giant lipomas (>5 cm in diameter) or atypical radiological imaging suggestive of malignant transformation with malignant histological degeneration after a biopsy.⁸ The size of the original lipoma in our case would

be an indication for referral to a specialist sarcoma centre and removal of the mass.

Although the transformation of an area or of the whole lipoma has been broadly described, we could find no case describing a completely different tumour (spindle cell sarcoma) encased within a lipoma. The guidelines for suspecting a soft tissue sarcoma remain pertinent even in rare cases such as this, as a large mass, which is increasing in size and is painful, should always be referred urgently to a specialist sarcoma service.

Learning points

- ▶ Patients should always be reassessed based on their current clinical status regardless of previous diagnoses.
- ▶ Sarcomatous change within a lipoma is a rare but significant event.
- ▶ If a patient presents with a soft tissue mass that is increasing in size, >5 cm in size or painful, then they should be referred urgently to a regional sarcoma service.
- ▶ As a useful aide-memoire; if a lump is bigger than a golf ball and growing, then think sarcoma.

Contributors JS, SE and SS: conceived and designed research; JCW: drafted the manuscript and prepared figures; JS and SE: edited and revised manuscript and approved final version of manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Dangoor A, Seddon B, Gerrand C, *et al.* UK guidelines for the management of soft tissue sarcomas. *Clin Sarcoma Res* 2016;6:20.
- 2 Casali PG, Abecassis N, Bauer S, *et al.* ESMO Guidelines Committee and EURACAN; Soft tissue and visceral sarcomas: ESMO–EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of Oncology* 2018;29:iv51–67.
- 3 Grimer RJ. Size matters for sarcomas!. *Ann R Coll Surg Engl* 2006;88:519–24.
- 4 Nandra R, Forsberg J, Grimer R. If your lump is bigger than a golf ball and growing, think Sarcoma. *Eur J Surg Oncol* 2015;41:1400–5.
- 5 Lex JR, Aoude A, Stevenson JD, *et al.* Multiple soft tissue sarcomas in a single patient: an international multicentre review. *Sarcoma* 2018;2018:1–7.
- 6 Murphey MD, Carroll JF, Flemming DJ, *et al.* From the archives of the AFIP: benign musculoskeletal lipomatous lesions. *Radiographics* 2004;24:1433–66.
- 7 Butler ED, Hamill JP, Seipel RS, *et al.* Tumors of the hand. A ten-year survey and report of 437 cases. *Am J Surg* 1960;100:293–302.
- 8 Bogumill GP, Sullivan DJ, Baker GI. Tumors of the hand. *Clin Orthop Relat Res* 1975;108:214–22.
- 9 Leclère FM, Casoli V, Pelissier P, *et al.* Suspected adipose tumours of the hand and the potential risk for malignant transformation to sarcoma: a series of 14 patients. *Arch Orthop Trauma Surg* 2015;135:731–6.
- 10 Mentzel T, Toennissen J, Rütten A, *et al.* Palmar atypical lipomatous tumour with spindle cell features (well-differentiated spindle cell liposarcoma): a rare neoplasm arising in an unusual anatomical location. *Virchows Arch* 2005;446:300–4.
- 11 Allen B, Rader C, Babigian A. Giant lipomas of the upper extremity. *Can J Plast Surg* 2007;15:141–4.
- 12 Peterson JJ, Kransdorf MJ, Bancroft LW, *et al.* Malignant fatty tumors: classification, clinical course, imaging appearance and treatment. *Skeletal Radiol* 2003;32:493–503.
- 13 Somerville SMM, Patton JT, Luscombe JC, *et al.* Clinical outcomes of deep atypical lipomas (well-differentiated lipoma-like liposarcomas) of the extremities. *ANZ J Surg* 2005;75:803–6.
- 14 Gaskin CM, Helms CA. Lipomas, lipoma variants, and well-differentiated liposarcomas (atypical lipomas): results of MRI evaluations of 126 consecutive fatty masses. *AJR Am J Roentgenol* 2004;182:733–9.
- 15 Kransdorf MJ, Bancroft LW, Peterson JJ, *et al.* Imaging of fatty tumors: distinction of lipoma and well-differentiated liposarcoma. *Radiology* 2002;224:99–104.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow