CASE REPORT

Radiotherapy for a benign cause of cauda equina compression in a known case of breast carcinoma

Neeraj Bhalla,¹ David J Husband,¹ Robin Pillay,² Nicki Thorp¹¹Clatterbridge Cancer Centre NHS Foundation Trust, Wirral, UK²The Walton Centre for Neurology and Neurosurgery NHS Trust, Liverpool, UK

Correspondence to

Dr Neeraj Bhalla,
neerajgowda@gmail.com

SUMMARY

A woman underwent breast conservation surgery and axillary clearance for T1N1M0 breast carcinoma, followed by adjuvant chemotherapy, radiotherapy and hormone therapy. At 3-year follow-up she presented with lumbar back pain and developed bilateral lower limb weakness. MRI spine demonstrated an expansile lesion at L1 causing cauda equina compression. The mass, unusually, was centred on the spinous process; metastases typically involve pedicles. The patient underwent surgical decompression with complete resolution of neurological signs. Histology revealed Masson tumour (intravascular papillary endothelial hyperplasia), a benign vascular lesion. Pain recurrence 9 months later prompted imaging demonstrating recurrent mass. Preoperative embolisation and re-excision was undertaken for recurrent Masson tumour. Recurrence of these lesions is rare and it was felt residual disease was likely. Radiotherapy has been used in isolated cases; therefore, she was treated with adjuvant radiotherapy, the first reported case of radiation in management of extracranial Masson tumour, and remains well 3 years later.

BACKGROUND

This case highlights the need to be aware of alternative, benign diagnoses in a patient with previous malignancy and to be especially wary of solitary lesions. It also highlights the importance of keeping a broad range of possible diagnoses in mind when faced with unusual radiological appearances. In this instance, vascularity of the mass lesion on MRI and its involvement in the spinous process was atypical of breast carcinoma metastasis. Involvement of the multidisciplinary team including radiology, spinal surgeons and pathologists enabled correct diagnosis and management of this patient who may otherwise have been assumed to have metastatic breast cancer.

Intravascular papillary endothelial hyperplasia (IPEH) is rare and complete surgical excision is the mainstay of management. Recurrences are unusual but there are reports of re-excision in the literature. However, there are only two case reports of radiotherapy being used in this setting, in both instances for intracranial disease. This case demonstrates the potential benefit of radiation following recurrence of benign IPEH in extracranial disease also.

CASE PRESENTATION

A 51-year-old postmenopausal woman was diagnosed with left breast carcinoma following

screening mammography within the NHS Breast Screening Programme. The patient underwent breast conservation surgery and axillary node sampling. Two out of four lymph nodes excised contained metastatic carcinoma; therefore, the patient underwent a left level II axillary node clearance. Final histology showed a grade 1 11 mm invasive ductal carcinoma, with 2 of the total 11 nodes positive for carcinoma. This T1N1M0 left breast carcinoma was strongly positive for oestrogen receptor (90%) and positive for progesterone receptor (50%). Routine Human Epidermal Growth Factor Receptor 2 (HER-2) receptor status testing was not performed when the patient originally presented to the Breast Unit, in 2005.

There were no comorbidities and the patient was not on any regular medication. The patient had one son, was a non-smoker and had no family history of breast or ovarian malignancy. Following surgery the patient received eight cycles of adjuvant chemotherapy with epirubicin followed by CMF (cyclophosphamide, methotrexate, 5-fluorouracil). This was followed by adjuvant radiotherapy to the left breast to a dose of 45 Gy in 20 fractions followed by a boost of 15 Gy in five fractions to the tumour bed. The patient tolerated the treatment reasonably well and on completion was started on adjuvant hormone therapy with anastrozole. Baseline bone densitometry scan showed osteopenia within the lumbar spine and normal bone density in the femoral neck; therefore, adcal-D3 was recommended.

The woman remained well following completion of her adjuvant chemotherapy and radiotherapy. Three years later, at routine outpatient follow-up, the patient described a 4-week history of sudden onset low back pain. The patient had attended the accident and emergency department for this and undergone plain x-ray of the lumbar spine that had not shown an obvious lesion. Further investigations were arranged.

When the patient was reviewed with the results 4 weeks later, the back pain was more severe and the patient now reported a 2-week history of right leg weakness and some difficulty walking. The patient denied any paraesthesia or sphincter disturbance. The back pain was eased but not relieved with co-codamol and ibuprofen.

On examination the patient had marked tenderness on palpation of the lumbar spine; power was reduced in the lower limbs bilaterally to four out of five, knee and ankle reflexes were brisk and there was sensory level at L2.

To cite: Bhalla N, Husband DJ, Pillay R, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009549

INVESTIGATIONS

At onset of pain:

Plain x-ray lumbar spine (figure 1): Mild osteopenia and degenerative changes. No evidence of sclerotic or lytic metastatic deposit. No vertebral collapse or fracture.

6 weeks later: DEXA bone densitometry scan–

L2–L4 osteoporosis (significant decrease in bone mineral density compared with baseline scan 3 years prior to this): T-score -3.6 .

Normal bone density in femoral neck: T-score -0.6 .

Isotope bone scan: Patchy, moderately intense abnormal uptake within T12 and L1 vertebral bodies, appearances suspicious of neoplastic disease but not diagnostic.

4 weeks later (arranged urgently on day of clinic follow-up):

MRI whole spine (figures 2 and 3): A large, expansile mass lesion centred on the spinous process of L1 with expansion anteriorly into the epidural space and laterally into the transverse processes producing a 2.8 cm segment of cauda equina compression with the canal diameter measuring just 2 mm at its narrowest point. Mass size $4.6 \times 4.3 \times 5.5$ cm. Signal change noted in both pedicles of L1 and within the pars intra-articular of L2.

No other metastatic foci seen in the rest of the spine.

Mild left-sided hydronephrosis noted, no convincing soft tissue lesion seen in left renal pelvis but further assessment advised with CT imaging.

CT scan chest/abdomen: Mild left hydronephrosis, no renal mass lesion; no visceral metastases.

DIFFERENTIAL DIAGNOSIS

A woman previously treated for node-positive breast carcinoma (with treatment including an aromatase inhibitor) who presented with back pain may have osteoporotic vertebral collapse,

Figure 1 Plain x-ray of lumbar spine demonstrating mild osteopaenia and degenerative changes but no evidence of metastases or vertebral collapse.

Figure 2 Sagittal MRI spine image (STIR sequence) showing mass lesion centred on spinous process of L1 vertebra.

bone metastases, benign degenerative disc disease or acute muscle strain. It would be important to elicit if there was any history of trauma. As the patient had been treated with an aromatase inhibitor, hormones which can cause a degree of bone thinning, it would be useful to know the results of any bone densitometry scans and whether she had been taking the commonly prescribed vitamin D and calcium supplements.

If there are also neurological symptoms or signs, neural axis compression becomes an important and urgent consideration and is more suggestive of metastatic disease with malignant cord compression, though if there was a history of trauma, acute disc protrusion could also be a cause of pain and leg weakness if severe. There are some rare benign causes of neural axis compression such as vertebral haemangiomas and myxolipomas. However, with a history of malignancy, it would be more likely in many cases for the cause to be malignant disease.

It is therefore important to take into account the stage, grade and natural history of the previous malignancy when weighing the probability of the cause being malignant cord compression. In this case, the history was of node-positive breast cancer, node-positivity being the strongest negative prognostic marker

Figure 3 Axial T1 MRI image through L1 vertebra showing mass lesion encroaching upon spinal canal.

in breast cancer, but her tumour was small (11 mm) and only Grade 1. Another consideration is whether this could be the first manifestation of a second malignancy. In this case, the patient had been diagnosed with breast cancer only 3 years earlier, but as more long-term survivors of breast, prostate and bowel cancer are now seen, consideration should be given to second malignant diagnoses, especially if there has been a long disease-free interval in a relatively good prognosis tumour.

In this case, MRI scan of the spine revealed a mass centred on the spinous process of L1 vertebra. This is an unusual appearance for a breast metastasis that typically involves the pedicles of the affected vertebra. It also had a vascular appearance on contrast images and raised the possibility of a renal metastasis, as there had been abnormal appearances of the left kidney also on imaging. Unusual imaging appearances can therefore act as an alert to clinicians to broaden the possible differential diagnoses.

Diagnosing the first relapse of a previous malignancy is a serious and life-changing diagnosis for the patient. If there is at all doubt, further investigations, imaging and biopsies should be sought to ensure correct diagnosis as all further management depends on this. Incorrectly diagnosing relapse of a cancer means a patient may be subjected to unnecessary and potentially harmful chemotherapy and radiotherapy treatments.

TREATMENT

The patient was started on dexamethasone. MRI appearances suggested vascularity of the mass lesion and therefore preoperative embolisation was considered. The patient underwent urgent L1 decompression and posterior stabilisation surgery with complete resolution of neurological symptoms and signs. Histology showed skeletal muscle, fibrofatty tissue and pieces of reactive bone and fibrocartilage. The bone and fibrofatty tissue contained many vessels including clusters of capillaries and larger cavernous and thick walled vascular channels. There was extensive thrombosis with organisation and IPEH or 'Masson tumour'.

Therefore, appearances were those of a vascular malformation with extensive secondary changes. There was no evidence of malignancy.

Pain recurrence 9 months later prompted a repeat MRI that demonstrated a recurrent soft tissue mass at L1. The patient underwent preoperative embolisation, in view of the vascularity of the lesion, and re-excision. Histology revealed recurrent Masson tumour.

Following re-excision it was felt highly likely that there was residual Masson tumour at the site of recurrence but further surgery was not thought to be feasible. Therefore, the potential role of radiotherapy was discussed with her. The patient was subsequently treated with 40 Gy in 20 fractions over 28 days with 10 MV photons using rapid arc technique and cone beam CT for image guidance. The treatment was tolerated well with no acute toxicity.

OUTCOME AND FOLLOW-UP

The patient remains clinically well and mobile with aids. There is no evidence of progression of the spinal IPEH 3 years after preoperative embolisation, re-excision and adjuvant radiotherapy, and no evidence of recurrent breast carcinoma 9 years after initial treatment.

DISCUSSION

This case illustrates well the importance of multidisciplinary team management in Oncology. Compression of the spinal neuraxis may be managed by surgery and radiotherapy or radiotherapy alone. Therefore, involvement of spinal surgical colleagues

following such a diagnosis is essential in assessing the most appropriate management in individual cases.

There is also evidence of improved functional outcome with regard to mobility following cord compression managed by surgical decompression and radiotherapy, rather than radiotherapy alone.¹ However, in many cases, surgery is not appropriate due to factors such as multiple metastases in adjacent vertebrae, comorbidities, the patient is of poor performance status and a long history of established neurological deficit.

In the case described here, surgical intervention enabled a benign tissue diagnosis to be made. The need for histology is especially relevant in the setting of a solitary metastasis and even following biopsy confirmation of metastatic disease, there can be a role for metastasectomy in certain situations. In the setting of vertebral lesions histology is also important when masses involve the spinous process only, as metastatic deposits are most commonly centred on the pedicles. Discussion between oncologist, radiologist, pathologist and the appropriate surgical speciality is required.

IPEH is a benign lesion of vascular origin.² It was first described by pathologist, Pierre Masson, in 1923, and hence carries the eponymous name 'Masson tumour'.² It is caused by excessive proliferation of endothelial cells in normal blood vessels or vascular malformations and is associated with organising thrombus (figure 4).³ It presents as an abnormal mass and, depending on the location, may be confused with benign or malignant neoplasms.³ Histologically and clinically it can be mistaken for a low grade angiosarcoma.⁴ It is a rare lesion but has been shown to have a predilection for the head and neck region.⁵ Contrast-enhanced MRI evaluation can show variable non-homogeneous enhancement, probably related to the fact that lesions are likely to have varying amounts of both solid areas of papillary structure and thrombus and anastomotic low-flow vascular channels.⁶ Conservative surgical excision is the treatment of choice with an excellent prognosis, except in intracranial lesions. Recurrence is rare.⁶

There are only two previous case reports in the literature of Masson tumours causing compression of the spinal neuraxis.^{7 8} The first case⁷ is described in a boy of 16 years who developed progressive bilateral lower limb weakness and mid-thoracic back pain over 1 week, with urinary hesitancy in the final 24 h. Examination revealed upper motor neuron signs and a CT

Figure 4 Masson tumour (intravascular papillary endothelial hyperplasia). Haematoxylin and eosin staining demonstrates fibrous tissue surrounding dilated vascular spaces. The central space shows endothelial proliferation. (Original magnification $\times 200$).

myelogram demonstrated marked cord compression at T6 level due to an extradural mass. He underwent resection of the lesion whose histological appearance was typical of Masson's vegetant haemangioendothelioma. Symptomatic improvement was seen within 24 h postoperatively with completely normal neurological function by 6 months.

The second case⁸ again occurred in a young boy, 17 years of age, who presented with pain, lower limb weakness, paraesthesia and bladder dysfunction. Following imaging confirmation of cauda equina compression he underwent a T12-L1 laminectomy. He had complete resolution of his pain and neurological symptoms improved significantly following excision of the Masson tumour.

There are two reported cases of radiotherapy for IPEH, both involving cavernous sinus lesions incompletely resected.^{9 10} In the first case, the lesion originated in the cavernous sinus but extended into the sellar region and was incompletely resected. MRI 3 months postoperatively demonstrated further growth of the lesion hence treatment with radiation was given. Subsequent shrinkage of the lesion was seen over 3.5 years follow-up imaging.⁹ The second case involved a 41-year old woman with a left middle fossa Masson tumour involving the cavernous sinus.¹⁰ Approximately half of the histologically confirmed IPEH was resected and the residual disease was treated with γ knife radiosurgery postoperatively with no recurrence 2 years after completion of treatment.¹⁰ A total dose of 40 Gy, delivered as 2 Gy per fraction, was chosen in our case based on the recommended radiation dose for treatment of symptomatic vertebral haemangiomas.¹¹

Radiotherapy in non-malignancy is justified in invasive or aggressive disease when other treatment modalities are unavailable or have failed. It can play a role in the treatment of benign conditions such as acoustic neuromas, arteriovenous malformations, thyroid eye disease and craniopharyngiomas.

We describe the first reported case of extracranial IPEH receiving radiotherapy, in the setting of recurrent disease with incomplete excision, in a woman with a history of breast carcinoma. It serves as a useful reminder of the principles of irradiation in benign disease including failure of conventional therapy, consideration of consequences of non-treatment, consideration of late effects and careful patient discussion to ensure informed consent.

Contributors All authors participated in discussions regarding the outline of the case report, revision of the article and gave final approval of the submitted version.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Patchell RA, Tibbs PA, Regine WF, *et al.* Direct decompressive surgical resection in the treatment of spinal cord compression caused by metastatic cancer: a randomised trial. *Lancet* 2005;366:643–8.
- 2 Hooda S, Humphreys MR, Wong SW, *et al.* Masson's pseudotumour of the ethmoid sinus—a case report. *J Laryngol Otol* 2008;122:990–2.
- 3 Akhtar M, Aslam M, Al-Mana H, *et al.* Intravascular papillary endothelial hyperplasia of renal vein: a report of two cases. *Arch Pathol Lab Med* 2005;129:516–19.
- 4 Barr RJ, Graham JH, Sherwin LA. Intravascular papillary endothelial hyperplasia. A benign lesion mimicking angiosarcoma. *Arch Dermatol* 1978;114:723–6.
- 5 Lancaster JL, Alderson DJ, Sherman IW, *et al.* Papillary endothelial hyperplasia (Masson's tumour) of the maxillary sinus. *J Laryngol Otol* 1998;112:500–2.
- 6 Pantanowitz L, Muc R, Spanger M, *et al.* Intravascular papillary endothelial hyperplasia (Masson's tumour) manifesting as a lateral neck mass. *Ear Nose Throat J* 2000;79:806, 809–10.
- 7 Porter DG, Martin AJ, Mallucci CL, *et al.* Spinal cord compression due to Masson's vegetant intravascular haemangioendothelioma. Case report. *J Neurosurg* 1995;82:125–7.
- 8 Taricco MA, Vieira JO Jr, Machado AG, *et al.* Intravascular papillary endothelial hyperplasia causing cauda equina compression: case report. *Neurosurg* 1999;45:1478–80.
- 9 Kristof RA, Van Roost D, Wolf HK, *et al.* Intravascular papillary endothelial hyperplasia of the sellar region. Report of three cases and review of the literature. *J Neurosurg* 1997;86:558–63.
- 10 Ohshima T, Ogura K, Nakayashiki N, *et al.* Intravascular papillary endothelial hyperplasia at the superior orbital fissure: report of a case successfully treated with gamma knife radiosurgery. *Surg Neurol* 2005;64:266–9.
- 11 Rades D, Bajrovic A, Alberti W, *et al.* Is there a dose-effect relationship for the treatment of symptomatic vertebral haemangioma? *Int J Radiat Oncol Biol Phys* 2003;55:178.

Learning points

- ▶ In the setting of a solitary metastasis, surgery should be considered to give histological diagnosis as well as being a treatment option.
- ▶ Surgical decompression followed by radiotherapy gives improved mobility following malignant spinal cord compression, compared with radiotherapy alone.¹
- ▶ Intravascular endothelial papillary proliferation or 'Masson tumour' is a benign, vascular lesion caused by excessive proliferation of endothelial cells with associated organising thrombus.³
- ▶ Complete surgical excision is the treatment of choice for Masson tumours but in rare cases of recurrence or progression following incomplete excision, adjuvant radiotherapy may be considered.
- ▶ The principles of irradiation in benign disease include failure of conventional therapy, consideration of consequences of non-treatment, consideration of late effects and careful patient discussion to ensure informed consent.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-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