

Late recurrence of pelvic osteosarcoma: a case report and review of the literature

B.G.I. Spiegelberg, K. Gokaraju, M.T. Parratt, A.M. Flanagan,
S.R. Cannon and T.W.R. Briggs

The Royal National Orthopaedic Hospital NHS Trust, Stanmore, Middlesex, UK

*Corresponding address: Ben Spiegelberg, MBBS BSc MRCS (Eng), London Bone and
Soft Tissue Tumour Service, The Royal National Orthopaedic Hospital,
Brockley Hill, Stanmore, Middlesex, HA7 4LP, UK.
E-mail: benspiegelberg@doctors.net.uk*

Date accepted for publication 27 November 2009

Abstract

A rare case of local recurrence of a high-grade osteosarcoma of the pelvis is described, 19 years after initial presentation, highlighting the need for continued long-term follow-up of patients with osteosarcoma. We review the literature concerning osteosarcoma of the pelvis, local recurrence and treatment with custom-made hemipelvic replacements.

Keywords

Pelvic bone tumour; osteosarcoma; recurrence; hemipelvic replacement.

Case report

A 42-year-old patient, initially presented in 1989 having been referred to The London Sarcoma Service at Royal National Orthopaedic Hospital by her GP. She had been complaining of intermittent pain in her right hip that occasionally radiated to her foot. A plain radiograph revealed a lytic lesion in the right ilium involving the superior acetabulum (Fig. 1) and preoperative magnetic resonance imaging (MRI) confirmed the mass measuring 20 × 14 cm. Subsequent biopsy confirmed a high-grade intramedullary osteosarcoma with both chondroblastic and osteoblastic differentiation. Staging scans, including computed tomography (CT) chest and bone scans, were negative for metastatic disease. The patient was treated with neoadjuvant chemotherapy (cisplatin and doxorubicin) 1 cycle preoperatively and 5 postoperatively. The patient underwent a two-stage hemipelvic replacement; tumour excision was undertaken initially and a temporary spacer inserted after which a custom-made prosthesis was manufactured and inserted (Fig. 2). The 2nd stage was delayed as the patient developed a deep vein thrombosis at the sapheno-femoral junction which was treated by heparin anticoagulation and insertion of an inferior vena cava filter. Following the 2nd stage operation the heparin infusion was restarted prior to the patient being given warfarin. Subsequently the wound required debridement in theatre and a large haematoma was evacuated. This went on to form a discharging sinus that required further debridement 4 months postoperatively. Ultimately the patient developed a fistula communicating with the hip joint, however, her symptoms were controlled with oral metronidazole and cephalexin therefore further revision surgery was not undertaken.



Fig. 1. Initial plain antero-posterior pelvic radiograph displaying lytic lesion in right ilium.



Fig. 2. Post-operative antero-posterior radiograph of right hip with hemipelvic replacement in situ.

Four years after tumour excision the patient developed a fluctuant mass over the posterior aspect of the proximal femur and an ultrasound scan showed a multiloculated abscess around the prosthesis. Both femoral and pelvic prosthetic components were removed and the wound washed out; subsequent microbiology grew *Staphylococcus epidermidis*. The patient was also noted to have an upper third femoral fracture which was treated with postoperative traction for 8 weeks

via a Stienmann pin in the distal femur. The femoral fracture healed in a non-union and the patient continued to mobilise with two crutches and was unwilling to have further surgery in view of the fact the wound had healed. The patient remained well for the next 15 years and was reviewed at regular intervals in outpatient clinics.

In 2008, while under investigation for a possible renal clear cell carcinoma she was noted to have a mass in the right iliac fossa involving psoas muscle adjacent to the previous surgical area (Fig. 3). Biopsy of the mass revealed a high-grade osteosarcoma similar to the initial histology. Staging investigations revealed no evidence of metastatic disease. The mass was excised with L4 and L5 nerve roots as they were involved in the tumour; despite this intralesional margins were identified. She had an uneventful perioperative course and subsequently had the renal clear cell carcinoma excised by assisted laparoscopic technique. The patient was due to undergo postoperative radiotherapy to the right hip but before this was initiated she was noted to have further recurrence on MRI. Re-excision of the recurrent tumour was performed, histology confirmed a recurrent high-grade osteosarcoma showing a mixed fibroblastic and telangiectatic phenotype with inadequate margins. She subsequently developed pulmonary metastatic disease that was not amenable to treatment; the patient died shortly afterwards.

Discussion

Pelvic osteosarcomas are rare; they account for less than 10% of all osteosarcomas^[1]. The overall survival of patients with pelvic osteosarcoma is reported to be between 20 and 47%. One of the reasons for the poor prognosis is that pelvic osteosarcomas are often diagnosed late when the tumour has reached a large size^[2]. In a series by Fahey *et al.*^[3] in which patients with pelvic osteosarcoma were observed, the duration of their symptoms ranged from 1 to 15 months before first biopsy. Intrapelvic growth causes few early symptoms accounting for the large size of some tumours at presentation. There are few major fascial or anatomical barriers within the pelvis to prevent extension into or along adjacent tissues.

There are no previous documented cases of high-grade osteosarcomas specifically of the pelvis, giving rise to local recurrence after such a long interval. There are previous reports of lower grade parosteal osteosarcomas of the femur that have recurred at the site of previous operation after 17 and 20 years, respectively^[4,5].

Grimer *et al.*^[6] followed a series of 96 patients with local recurrence of osteosarcoma; the median time to develop local recurrence was 11 months from initial excision (range 1-66 months). This study also highlighted that the risk of developing local recurrence decreased with time; 60% of local recurrence arose within 12 months of initial surgery and 82% within 24 months. In contrast, data on osteosarcoma of the pelvis show that local recurrence is far more common, with studies by the Cooperative Osteosarcoma Study Group reporting a 62% risk of local recurrence within 3 years of surgical excision^[7].

There are several reasons for the poor prognosis associated with pelvic osteosarcoma. Preoperative chemotherapy is less effective at reducing tumour volume generally because of the bulk of the primary lesions^[8]. This correlates with large tumour size (greater than 15 cm) being a poor prognostic factor^[9] and complete tumour excision conferring an improved prognosis. In a study of 28 surviving patients with pelvic osteosarcoma 32% developed local recurrence; the incidence of local recurrence was 13% in wide excisions, 38% in marginal excisions and 80% in intralesional excisions^[10]. There is no reported difference in survival after 5 years between those patients having hindquarter amputation (33%) and those with limb salvage surgery (37%)^[8]. This is corroborated in other studies^[11,12]. When comparing the margins of excision between hemipelvectomy and limb salvage procedures there was no statistical difference between the two surgical interventions^[10].

The Rizzoli Institute looked at predictive factors for local recurrence in osteosarcoma. They found that with adequate surgical margins, there was 97% 7-year local recurrence free survival, whereas with inadequate margins this was reduced to 71%^[13]. This group also analysed the effectiveness of chemotherapy and found 4% of patients with more than 90% tumour necrosis developed local recurrence in comparison with 10% with less than 90% tumour necrosis. The Institute also performed a study specifically to identify predictive factors of late relapse^[14]. They analysed the effect of sex, site, tumour volume, histological subtype, serum alkaline phosphatase, serum lactate dehydrogenase, type of surgery and histological response to chemotherapy. They found that none of these parameters were predictive for late relapse. In their series, patients who

had intralesional excision and relapsed did so with distant metastases as opposed to local recurrence.

The responsiveness of pelvic osteosarcomas to chemotherapy is particularly poor. A study comparing the proportion of tumour necrosis in pelvic osteosarcomas found a mean amount of tumour necrosis of 40%^[8]. This coupled with the difficulty in gaining adequate surgical margins results in the poor prognosis seen with pelvic osteosarcomas.

When local recurrence of osteosarcoma occurs it tends to be extraosseous; in 96 patients followed in one series, 80% of local recurrence occurred within the soft tissue^[6]. Furthermore, at the time of development of local recurrence 18% of patients were already known to have metastases; 23% were found to have metastases synchronously and 38% developed them at a later date. Once local recurrence has occurred survival at 1 year is 68%, 51% at 2 years, 41% at 5 years and 36% at 10 years. In patients who have metastases at the time of local recurrence the survival statistics are overwhelmingly poor; 30% at 1 year and 14% at 2 years^[6].

The case described is particularly rare in that there was such a long interval between excision of tumour and development of recurrence especially as many of the characteristics in the history are predictive of an earlier recurrence. The lesion was large (20 × 14 cm), was of high-grade intramedullary subtype and responded poorly to chemotherapy with <90% necrosis. Once the local recurrence occurred and complete excision was not achieved the disease inevitably progressed resulting in metastatic spread.

Long-term follow-up of patients who have had hemipelvic replacements reveal the commonest complication to be infection. Jaiswal *et al.*^[15] identified 98 patients who had reconstruction of the hemipelvis following tumour excision. Fifty-eight percent developed surgical complications, of which 32% required one or more further operations. Infection was the most frequent complication in 31%, followed by local recurrence in 30% and recurrent dislocation of the hip in 20%. An earlier study^[16] also stated that preservation of the sciatic nerve is essential for adequate function and that if removal of the gluteal vessels and nerves is necessary for tumour clearance, then limb preservation is contraindicated due to lack of abductor function.

This case highlights the importance of long-term follow-up in patients with osteosarcoma. Routine follow-up after 5 years should include annual clinical assessment, examination and radiography of the chest and primary site of disease^[17]. Patients with osteosarcoma are generally discharged after 10 years at our centre if they have remained disease free. However, it is important to advise them that there is still a risk of recurrence so that they can remain aware of any symptoms that may be suggestive of recurrence.

References

1. Unni KK. Dahlin's bone tumours: general aspects and data on 11,087 cases. Philadelphia, PA: Lippincott-Raven; 1996.
2. Wurtz LD, Peabody TD, Simon MA. Delay in the diagnosis and treatment of primary bone sarcoma of the pelvis. *J Bone Joint Surg Am* 1999; 81: 317-25.
3. Fahey M, Spanier S, Vander Griend RA. Osteosarcoma of the pelvis. A clinical and histopathological study of twenty-five patients. *J Bone Joint Surg Am* 1992; 74: 321-30.
4. Koksall Y, Akyuz C, Varan A. Late recurrence in primary region of parosteal osteosarcoma: a case report. *Pediatr Hematol Oncol* 2008; 25: 83-8. doi:10.1080/08880010701621010.
5. Lau TW, Shek TWH, Wong LLS. Local recurrence of parosteal osteosarcoma adjacent to a prosthesis after 20 years: a case report. *J Orthop Surg (Hong Kong)* 2004; 12: 263-6.
6. Grimer RJ, Sommerville S, Warnock D, *et al.* Management and outcome after local recurrence of osteosarcoma. *Eur J Cancer* 2005; 41: 578-83. doi:10.1016/j.ejca.2004.11.012. PMID:15737563.
7. Ozaki T, Flege S, Kevric M, *et al.* Osteosarcoma of the pelvis: experience of the cooperative osteosarcoma study group. *J Clin Oncol* 2003; 21: 334-41. doi:10.1200/JCO.2003.01.142. PMID:12525527.
8. Grimer RJ, Carter SR, Tillman RM. Osteosarcoma of the pelvis. *J Bone Joint Surg (Br)* 1999; 81: 796-802. doi:10.1302/0301-620X.81B5.9241.
9. Taylor WF, Ivins JC, Dahlin DC. Trends and variability in survival from osteosarcoma. *Mayo Clin Proc* 1978; 53: 695-700.
10. Kawai A, Huvos AG, Meyers PA, Healey JH. Osteosarcoma of the pelvis oncological results for 40 patients. *Clin Orthop Relat Res* 1998; 348: 196-207.

11. Carter SR, Eastwood DM, Grimer RJ, Sneath RS. Hindquarter amputation for tumours of the musculoskeletal system. *J Bone Joint Surg (Br)* 1990; 72: 490-3.
12. Skin K-H, Rougraff BT, Simon MA. Oncologic outcome of primary bone sarcoma of the pelvis. *Clin Orthop* 1994; 304: 207-17.
13. Bacci G, Ferrari S, Mercuri M, *et al.* Predictive factors for local recurrence in osteosarcoma. *Acta Orthop Scand* 1998; 69: 230-6.
14. Ferrari S, Briccoli A, Mercuri M, *et al.* Late relapse in osteosarcoma. *J Pediatr Hematol Oncol* 2006; 28: 418-22. doi:10.1097/01.mph.0000212944.82361.1d. PMID:16825986.
15. Jaiswal PK, Aston WJS, Grimer RJ, *et al.* Peri-acetabular resection and endoprosthetic reconstruction for tumours of the acetabulum. *J Bone Joint Surg (Br)* 2008; 90: 1222-7.
16. Abudu A, Grimer RJ, Cannon SR, Carter SR, Sneath RS. Reconstruction of the hemipelvis after the excision of malignant tumours. Complications and functional outcome of prostheses. *J Bone Joint Surg (Br)* 1997; 79: 773-9. doi:10.1302/0301-620X.79B5.6749.
17. Strauss S, McTiernan A, Whelan J. Late relapse of osteosarcoma: implications for follow-up and screening. *Pediatr Blood Cancer* 2004; 43: 692-7. doi:10.1002/pbc.20154.