Unusual association of diseases/symptoms

Pathological fracture of femur secondary to infected synovial sarcoma

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Summary

Pathological fracture is not uncommon in musculoskeletal oncological surgical practice. When complicated by infection, it is both limb-threatening and life-threatening problem. Pathological fractures require urgent investigation, not urgent treatment. We present the case of a 75-year-old man who presented with an infected sarcoma causing a pathological fracture of his right femur.

BACKGROUND

Pathological fracture of a bone is a challenging complication in orthopaedic practice, especially when the lesion is thought to be a primary lesion. This case describes an unreported combination of an infected primary sarcoma with an associated pathological fracture.

CASE PRESENTATION

A 75-year-old gentleman was referred by his family doctor to a local orthopaedic unit with a 7-month history of right thigh pain and swelling. He was well, with no systemic symptoms and on initial assessment was normocardic (80 bpm), normotensive (110/65 mm Hg) and no fever. Urgent imaging was obtained, with radiographs and an

MRI demonstrating a large soft tissue mass (12 cm×10 cm×8 cm) around the right distal shaft of femur, with a large zone of transition and associated cortical destruction (figures 1 and 2). The patient was referred to a tertiary referral orthopaedic musculoskeletal oncological service for a biopsy and further management.

Upon biopsy 5 days later, the patient was unwell; feeling weak, confused and anorexic with a 3-day history of fevers. Upon examination he was found to be tachycardic (110 bpm), hypotensive (85/50 mm Hg) and feverish (39.3°C). His urine was foul-smelling and macroscopically cloudy. Blood cultures and urine were sent for microbiological assessment. Haematological parameters on admission showed a raised white blood cell count (WCC) of

Figure 1 Radiograph of right femur on admission.

Figure 2 MRI of right femur.

$23 \times 10^9/l$ (normal range $4\text{--}11 \times 10^9/l$), with an erythrocyte sedimentation rate (ESR) of 100 and C reactive protein (CRP) of 343 mg/l. Renal functional parameters were markedly deranged with urea of 29 mmol/l (normal range 2.5–7.5 mmol/l) and creatinine of 490 $\mu\text{mol/l}$ (normal range 40–95 $\mu\text{mol/l}$).

The patient was admitted, aggressively hydrated and a percutaneous biopsy of the lateral right thigh was undertaken. Specimens were sent both for microbiological and histological assessments. The patient was started on a high-dose of broad-spectrum intravenous antibiotics (piperocillin/tazobacter). The following day the patient fractured his femur while turning in his bed (figure 3). A monoaxial external fixator was applied to the femur to achieve temporary stability, while the intravenous antibiotic therapy was continued. The urine, blood and soft tissue biopsy specimens taken on admission all cultured *Staphylococcus aureus*, and *Pseudomonas*. Antibiotics were adjusted to flucloxacillin and rifampicin following final sensitivities.

INVESTIGATIONS

A radionuclide bone scan did not show any other bony lesions, but CT-staging scans demonstrated subcentimeter

nodules in both lung bases suggestive of a metastatic disease. Histology of the biopsy specimen revealed a high-grade sarcoma-infiltrating skeletal muscle and connective tissue with abundant mitoses (mitotic index of 26 in 10 high powered field) and necrosis. Immunohistochemical studies demonstrated a strong staining with vimentin, and a weak staining with epithelial membrane antigen (EMA) with a focal BCL2 positivity. No staining was seen with S100, Desmin, Actins, CD99, CD45, CAM 5.2 or AE13. The histological appearances along with the immunohistochemical profile were the most in-keeping with a monophasic synovial sarcoma.

Five days after its application, the schanz pin sites of the distal femur began discharging a purulent material. Repeat cultures grew *S aureus* and *Pseudomonas*. Intravenous antibiotics were continued until the patient had no fever for a period of 5 days, at which time inflammatory markers were reducing (WCC 6.2, ESR 60 and CRP 45). A transoesophageal echocardiogram did not reveal any cardiac or valvular dysfunction.

TREATMENT

After a detailed consultation with the patient and his family, the patient underwent an above-knee amputation. A gross examination of the specimen revealed a grey-white tumour measuring $12.5 \times 10.5 \times 8.5$ cm. Extensive areas of haemorrhage and necrosis were present, centred on the medial portion of the thigh, 5 cm above the knee and 16 cm from the surgical margin. The histological and immunohistological assessments were similar to the biopsy specimen and the diagnosis of monophasic synovial sarcoma was confirmed.

OUTCOME AND FOLLOW-UP

The patient recovered well following a surgery, continuing intravenous antibiotics until his inflammatory markers returned to normal on the tenth postoperative day. He was assessed and fitted for an above-knee prosthesis, and was discharged home following a short period of inpatient rehabilitation. He was referred to the community palliative care service.

DISCUSSION

A pathological fracture of bone is a challenging complication in musculoskeletal oncological practice, especially when the lesion is thought to be a primary lesion. This case describes an unreported combination of an infected primary sarcoma with an associated pathological fracture. For a metastatic disease of bone, the primary intention of a therapy is not just to cure the disease or prolong a life but also to improve the quality of life remaining among such patients while keeping their stay in the hospital as short as possible and the rate of complications at an acceptable level.¹

Surgical options depend on the type and grade of lesion, and patient factors, including medical status, functional demands and life expectancy. For sarcomas, a better understanding of the biological behaviour of these tumours and advances in surgical techniques, surgical prostheses, radiographic imaging, radiotherapy and chemotherapy have led to the advent of limb-sparing surgeries where possible.²

Figure 3 Pathological fracture through the infected sarcoma.

Limb-sparing surgery is now the standard of care for a majority of extremity bones and soft tissue sarcomas.³ The primary indication for amputation now is the inability to obtain an adequate surgical margin while retaining a useful limb. Predisposing factors for amputation include widespread contamination of tissue by a previous surgery, fracture or infection, or in cases where significant neurological or vascular impairment is expected following a radical excision.

Although a recognised postoperative complication of a sarcoma surgery,⁴ preoperative infection of a sarcomatous lesion has not been described. Amputation is not an easy option, with a considerable impact on both the patient (including the psychological burden of losing a limb, phantom pain and a significant increase in the physiological burden of ambulation for lower-limb amputees especially in the elderly population) and on health provider (the expense of maintaining and replacing prosthetic limbs). At the above-knee level, amputation is associated with an increased risk of limp, walking-aid use, anxiety and an inability to drive.⁵ The decision to amputate should be based on local anatomic considerations, tumour grade and stage and consideration of the functional and psychological impact of the procedure.²

With a large high-grade primary sarcoma, evidence of septicaemia secondary to a urinary tract infection, polymicrobial infection of the lesion and pathological fracture with inevitable soft-tissue contamination, amputation was the inevitable option for this patient.

The result of this palliative amputation for this patient was an immediate clinical improvement in constitutional symptoms, elimination of pain, the ability to mobilise with crutches from day 1 postoperatively. This facilitated

an early-discharge home, usage of an above-knee prosthesis and an improvement in the patient's quality of life during the palliative phase of his treatment.

Learning points

- ▶ Infected malignant lesions of bone are a rare and challenging problem.
- ▶ Pathological fractures require urgent investigation, not urgent treatment.
- ▶ While limb salvage surgery is always the ideal, this is not always in the patients' best interests.
- ▶ Palliative amputation can be extremely effective at maximising a patient's quality of life.

Competing interests None.

Patient consent Obtained.

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