CASE REPORT

Myxoma of the femur: an unusual site of origin

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Accepted 5 August 2015

SUMMARY

Skeletal myxomas are rare benign tumours. Their occurrence in long bones of the extremities is rarely reported. A 45-year-old man presented with pain in his left proximal thigh for a duration of 4 months. Movements of the hip were painful. Radiography revealed an expansile osteolytic lesion in the left proximal femur near the lesser trochanteric region. On MRI, the lesion showed a homogenous signal enhancement with no cortical disruption. Extended curettage and bone grafting was performed. On gross examination, the curetted specimen was a yellowish-white mucoid material. Histopathology showed a tumour consisting of spindle-shaped and stellate-shaped cells with widely separated myxoid mucoid stroma, suggestive of intraosseous myxoma. At 2 years follow-up, there were no signs of recurrence and the patient was doing well with excellent hip and knee function.

BACKGROUND

Myxomas of the heart and mandible have been well documented in the literature. Mazabraud's syndrome is a separate entity where soft tissue myxomas are found in association with fibrous dysplasia of bone.¹ But bone myxomas are rare benign tumours with very few occurrences quoted in the literature. McClure and Dahlin² reported a series of three cases of bone myxoma in 1977, from the Mayo Clinic. A case of myxoma of the phalanx of the toe was reported in 1978.³ We would like to add a case of myxoma of the femur in a middle-aged man to this short list, and discuss its clinical features and management.

CASE PRESENTATION

A healthy 45-year-old man presented to our outpatient department with pain over the left proximal thigh region for the past 4 months. He was otherwise in good health and his medical and surgical history was unremarkable. On examination, there was no swelling or any other signs of inflammation over the left thigh. Range of motion of the left hip and knee joint was full but painful.

INVESTIGATIONS

Plain radiography and CT of the left femur was performed, showing an expansile osteolytic lesion in the left proximal femur near the lesser trochanteric region (figures 1 and 2). A skeletal survey revealed no other lesion anywhere in the body. On MRI, the lesion showed a homogenous signal enhancement with no cortical disruption—hypointense on T1-weighted and hyperintense on T2-weighted images (figure 3).

Figure 1 Preoperative X-ray showing an osteolytic lesion in the left proximal femur.

DIFFERENTIAL DIAGNOSIS

Taking these features into account, we suspected a benign lesion. Our main differential diagnosis was simple bone cyst and aneurysmal bone cyst.

TREATMENT

Extended curettage of the lesion was performed. An intraoperative frozen section was sent and the histopathology report was in favour of myxoma. Curettage was extended using hydrogen peroxide, phenol and a high-speed burr, and the resulting

Figure 2 CT scan showing an expansile osteolytic lesion without cortical breach.

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To cite: Santhanam SS, Goni V, Saibaba B, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-211480

Figure 3 T2-weighted MRI showing a homogenous hyperintense lesion in the left proximal femur.

defect was filled with a corticocancellous allograft (three femoral heads) and fibular autograft.

The curetted specimen was a yellowish-white mucoid material, which was sent for histopathological examination (figure 4). The histopathological report indicated a tumour consisting of spindle-shaped and stellate-shaped cells with widely separated myxoid mucoid stroma. The tumour cells had bland hyperchromatic nuclei, inconspicuous nucleoli and moderate cytoplasm (figure 5). There was no evidence of mitosis. The tumour cells showed uniform and strong cytoplasmic positivity for vimentin. Smooth muscle actin was variably expressed by the tumour cells, indicating focal myofibroblastic differentiation. The tumour cells were negative for S-100 and myogenin (figure 6). Based on this, we arrived at a final diagnosis of intraosseous myxoma. During the post-operative period, which was uneventful, the patient underwent a thorough examination of the cardiovascular system to rule out the presence of an atrial myxoma and to rule out the remote possibility of the femoral lesion being metastasis to an atrial myxoma.

OUTCOME AND FOLLOW-UP

The patient was kept non-weight bearing for 8 weeks postoperatively, after which gradual weight bearing was started.

At 2 years follow-up, there were no signs of recurrence and the patient was doing well with excellent hip and knee function (figure 7).

DISCUSSION

So far, very few cases of myxoma of bone have been reported in the literature. McClure and Dahlin² reported three cases, and a case of myxoma of the phalanx of the toe was reported in 1978.³ A search of the English literature revealed a case of periosteal myxoma of the femur reported in 1972.⁴ A case of

Figure 4 Intra-operative picture showing a yellowish-white mucoid tumour material.

Figure 5 Histopathological photomicrograph showing tumour composed of many monomorphic spindle-shaped to stellate-shaped cells in a myxoid background (H&E x200).

myxoma of the femur was reported in a 12-year-old boy in 1954.⁵ Our case would be next in order in this extremely rare list of reports on bone myxomas. All the cases reported by McClure and the case of myxoma of the toe were in older female patients in the sixth or seventh decade of life, while our case was of a relatively young man in his fourth decade. Except for the case of myxoma of the toe, all the other cases, including ours, occurred in the femur. All the lesions described so far were osteolytic, as was ours, and two of the three lesions described by McClure were multilocular.

Stout,⁶ in 1948, devised criteria for the diagnosis of myxoma, stating that: (1) true neoplasms are composed of stellate-shaped cells in loose mucoid stroma; (2) they would have no recognisable cellular elements and specifically no chondroblasts, lipoblasts or rhabdomyoblasts; (3) they are poorly vascularised; (4) they are of unicentric origin and (5) they display benign behaviour. Our case fulfilled all of these criteria and hence was confirmed to be a case of bone myxoma.

Myxomas and fibromyxomas have very similar clinical, radiological and biological features. Both types are benign lesions without the potential to metastasise.²⁻⁷ Although it is not of major importance to differentiate between myxoma and fibromyxoma, since both have similar treatment protocol and recurrence rates, it is important to differentiate these entities from fibrous dysplasia because of the higher recurrence rate of fibrous dysplasia. Myxomas and fibromyxomas usually tend not to recur.⁸ In fibrous dysplasia, bone resorption after using autogenous bone graft is reported to be around 36–55%, which can be reduced to 18% by using allogenic bone graft.^{9,10}

Histologically, myxoma has a myxoid stroma with varying amounts of spindle-shaped or stellate-shaped cells. In contrast to chondromyxoid fibroma and enchondroma, a lobulated pattern or chondromatous elements are not seen.

Figure 6 (A) The tumour cells show strong vimentin positivity (immuno-peroxide (IP) x100). (B) Some of the tumour cells show faint positivity for smooth muscle actin, while blood vessels show strong positivity (IP x200).

Figure 7 Postoperative X-ray at 2 years follow-up.

Metastases from an osseous myxoma have not been reported and the prognosis depends on the primary site of the tumour. Moran *et al* reported a cardiac myxoma metastasising to the os pubis. Extragnathic myxoma of bone is distinct from myxoma of the jaw and has a better prognosis. Myxomas of the jaw present with aggressive proliferation, cortical destruction, infiltration of the surrounding tissues and recurrence after local removal. In our case, the lesion was isolated, with no other myxoma (osseous/muscular) being found elsewhere in the body.

The treatment of choice for bone myxomas is complete curettage and bone grafting. Kamiyoshihara *et al*⁸ and Caballes¹¹ recommend complete excision or curettage of the tumour, with regular oncological follow-up. In our case, we had performed extended curettage and bone grafting. The case was followed up

for 2 years, with the patient maintaining excellent range of motion with no signs of recurrence of the tumour.

The importance of recognition of myxomas of the bone lies in the fact that these are benign bone lesions that can be treated with simple curettage and bone grafting, whereas there are other malignant bony lesions with myxoid features requiring more radical procedures.

Learning points

- ▶ Intra-osseous myxomas are rare benign tumours.
- ▶ Pain is a common presenting symptom.
- ▶ The lesion is lytic and expansile on plain radiography and CT scan; MRI shows homogenous signal enhancement.
- ▶ Histopathology is diagnostic.
- ▶ Curettage and bone grafting gives excellent results.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed

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