

Primary leiomyosarcoma of the femur and bony metastasis from the breast in the same patient

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Abstract

A 78-year-old lady, with no history of trauma, was admitted with gradually increasing pain in the right groin and thigh; she had been unable to weight-bear for 24 h. The patient also complained of intermittent left shin pain. She had been treated for carcinoma of the breast 22 years earlier by total mastectomy with axillary clearance followed by local radiotherapy; she showed no signs of recurrence. The plain X-ray revealed a pathological fracture of the right proximal femur with a large osteolytic lesion. The left tibial radiograph showed extensive osteolytic lesions. The patient underwent palliative internal fixation of both the femur and the tibia. The histopathological report from the right femoral biopsy revealed a primary leiomyosarcoma and from the left tibia revealed a bony metastasis from a breast primary. Further investigation excluded any extraskeletal leiomyosarcoma. Primary leiomyosarcoma is a very rare tumour involving long bones in adults; only a few cases have been reported in world literature. The breast tumour gave rise to a bony metastasis 22 years after 'cure' and without local recurrence, which is again very uncommon. Such a combination is previously unreported.

Keywords

Primary leiomyosarcoma; femur; bony; metastasis; breast carcinoma; tibia; orthopaedics.

Case history

A 78-year-old lady was admitted with gradually increasing pain in the right groin and thigh; she had been unable to weight-bear for 24 h. There was no history of trauma. The patient also complained of intermittent left shin pain, which she related to trivial trauma. The patient had been treated for carcinoma of the breast 22 years earlier by total mastectomy and axillary clearance followed by local radiotherapy. She showed no clinical sign of recurrence and had been discharged from follow-up some years previously.

Plain radiographs showed pathological fracture of the right femur with a large osteolytic lesion (Fig. 1), and extensive osteolytic lesions of the left tibia (Fig. 2). A bone scan was performed which showed extensive bony metastases involving the right proximal femur, the left tibia, the right hemipelvis and the skull. The primary could not be found despite extensive investigations.



Fig. 1. Radiograph showing pathological fracture of the right femur with a large osteolytic lesion.

In view of the general health of the patient, it was decided to proceed with palliative internal fixation of both the femur and the tibia.

The histopathology from the right femoral biopsy revealed a primary leiomyosarcoma of the femur and from the left tibial biopsy revealed a metastasis from breast carcinoma. Further investigation excluded an extraskeletal leiomyosarcoma as a primary lesion. Unfortunately, the patient died 4 weeks after the operation from a pulmonary embolism.

Diagnosis

Any patient with a suspected pathological lesion of the bone should undergo both radiological and blood investigations to determine the underlying pathology. Sometimes this is not possible without a biopsy, which in this case was taken during internal fixation. Only a few malignancies can be diagnosed by blood tests alone; these include prostate-specific antigen (PSA) for prostatic malignancy and monoclonal antibodies for myeloma. There is no specific blood marker for either leiomyosarcoma or breast metastasis. Primary leiomyosarcoma of the bone and bony metastasis from the breast presents identical radiological features. MRI cannot differentiate between primary bony tumours.^[1]

In this case the histology of the sarcomatous bony lesion was the same as soft tissue leiomyosarcoma, and consisted of broad, interlacing fascicles of spindle cells with eosinophilic cytoplasm, pleomorphic nuclei, mitosis and necrosis. Immunohistochemical and/or electron-microscopic studies have proved helpful in differentiating the tumour from other spindle-cell neoplasms.^[2] In our study, the immunohistochemistry on the right femoral lesion revealed strong tumour-cell labelling for vimentin and weak tumour cell labelling for Alpha-Smooth-Muscle-Actin (ASMA). The tumour cells were negative for epithelial markers (CAM 5.2, AE1 and AE3). The CD34 and S-100 were positive overall.

The histological sections from fragments of bone from the left tibia revealed a malignant epithelial tumour. The phenotypes of the tumour cells were in keeping with metastatic breast carcinoma. The epithelial nature of the tumour was confirmed by immunohistochemistry. The tumour cells showed strong labelling with CAM 5.2, AE1 and AE3, and were negative for ASMA, vimentin, desmin, CD34 and S-100.



Fig. 2. Radiograph showing extensive osteolytic lesions of the left tibia.

Unusual features

Secondary bony deposits are common in breast cancer but most unusual in the absence of local recurrence 22 years after treatment of the primary.^[3] Primary leiomyosarcoma of the proximal femur is again a very rare tumour. Fewer than 50 extragnathic cases have been reported in world literature.^[4] The clinical presentation is non-specific and it is a tumour found in adults with slight male predilection. The metaphyses of long bones are most commonly affected. The radiological features are non-specific, with aggressive osteolysis and no surrounding sclerosis. Neither MRI nor isotope bone scanning is specifically diagnostic. As mentioned previously histology, electron microscopy and immunohistochemistry are the investigations of choice for diagnosis and differentiation from other spindle-cell neoplasms. The surgical treatment of isolated compartmental primary leiomyosarcoma without metastasis is compartmental resection and amputation if the compartment is breached. Chemotherapy is an option for those with cutaneous

or pulmonary metastasis. The 5-year survival rate following a diagnosis of primary leiomyosarcoma is approximately 50%.

The unusual feature of this case is the occurrence of two extremely rare tumours in the same patient in different bones at the same time. In a younger, fitter patient this would have presented therapeutic difficulties.

Lesson

Primary leiomyosarcoma of the proximal femur is a rare tumour and the diagnosis is confirmed only by histopathology. Bony metastasis from breast cancer is possible even after 22 years without local recurrence. Tissue from the different sites must be sent separately for diagnosis because the same radiological appearances can have different underlying pathologies, as in rare cases the cause can be unexpected.

References

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