Bilateral demyelinating neuropathy in a solitary lytic and sclerotic myeloma of the proximal humerus: a case report

O. S. Schindler¹, T. W. R. Briggs¹, S. A. Gillies²

¹ Bone Tumour Unit, Royal National Orthopaedic Hospital, The London Bone Tumour Service, University of London

² Orthopaedic Department, Royal Infirmary of Edinburgh, University of Edinburgh

Accepted: 11 June 1996

Summary. *Mixed lytic and sclerotic myeloma are uncommon and occur at an earlier age than multiple myeloma in areas of persistent haemopoiesis. Their characteristic symptoms are pain, swelling and pathological fractures and they are usually detected at an early stage. We report a case of a mixed sclerotic/lytic myeloma of the humerus presenting at an unusually advanced stage and associated with a bilateral progressive demyelinating peripheral sensory and motor polyneuropathy resembling polyradiculitis. The neurological changes partly disappeared after the tumour had been resected which suggests that nerve reconstitution may occur due to reversal of the demyelination.*

Résumé. *Les myélomes sclérolitiques mixtes et isolés sont des lésions relativement peu communes et se révèlent à un âge plus jeune que les myélomes multiples débutant dans des zones d'hématopoïèse persistante. Ils sont généralement accompagnés de symptômes caractéristiques comme la douleur, des gonflements, des fractures pathologiques et sont par conséquent détectés au stade précoce de la maladie. La majorité des lésions myélomateuses isolées décrites dans la littérature sont purement ostéoscléreuses. Bien qu'elles soient en partie comparables au type sclérolitique, et que les deux apparaissent favorables en tant que lésions isolées, aucune donnée suffisante n'existe à ce jour, définissant l'aspect clinique et les traits caractéristiques des myélomes mixtes sclérolitiques. Nous rapportons un cas exceptionnel d'un myélome*

mixte isolé de l'humérus associé à une poly-neuropathie motosensorielle démyélinisante progressive, bilatérale et périphérique ressemblant à une polynévrite et apparaissant à une étape inhabituellement avancée de la tumeur. Les troubles neurologiques disparurent en partie après la résection, suggérant la reconstitution nerveuse due probablement à l'aspect réversible de la démyélinisation.

Introduction

Multiple myeloma is the commonest primary malignant bone tumour, comprising 17% of all malignant tumours [16]. About 3% of myeloma show osteosclerotic changes and only a very small number of these are solitary lesions [1, 15]. Solitary myeloma arise in areas of persistent haemopoiesis, such as the spine, pelvis, skull and ribs, and are rare in the humerus and femur [7]. They present at an earlier age than multiple myeloma and occur mainly between the fourth and sixth decades. Solitary myeloma do not usually have the typical features of the multiple lesions such as anaemia, hypercalcaemia, infiltration of the bone marrow by plasma cells, increase in M-protein or Bence-Jones proteinuria [6, 15]. Provided they are treated by local excision and adjuvant irradiation patients with solitary myeloma also have a much better prognosis compared to those with multiple myeloma, 50% of whom die within 2 years. The effectiveness of chemotherapy remains controversial [6, 11].

Fig. 1 a, b. Anteroposterior and lateral radiographs of the proximal part of the left humerus showing the expansive growth of a solitary mixed lytic/sclerotic myeloma

Fig. 2. Anteroposterior radiograph after the insertion of a custom-made humeral replacement

Demyelinating peripheral sensory and motor neuropathy is recognised as a complication of solitary myeloma with a reported incidence of between 12% and 50%, but has only been seen in purely sclerotic lesions [5, 6, 10, 15]. There is not sufficient data available at present to state the probability of developing neurological defects in mixed lytic and sclerotic myeloma [2].

We report the case history of a patient who presented with clinical signs of a bilateral demyelinating peripheral sensory and motor neuropathy of the upper extremities caused by an advanced mixed lytic and sclerotic myeloma in the upper humerus.

Case report

A nurse, who was 50 years of age, reported to her general practitioner in May 1994 with pain in her left shoulder after her arm had been pulled and twisted by a patient. She was referred for physiotherapy and improved, but she continued to have pain at the extremes of movement. In May 1995 she developed weakness of both arms; EMG and nerve conduction

tests suggested a progressive demyelinating peripheral sensory and motor neuropathy of unknown origin. The pain in her shoulder became worse and sensation in her left hand diminished, but it was another 2 months before radiographs were taken. These showed a pathological fracture of the surgical neck of the humerus with an osteolytic soap-bubble lesion occupying the whole of the proximal part of the left humerus (Fig. 1 a, b). No other skeletal lesions were found. Laboratory investigations showed anaemia [(9.6 g/dl), leucocytosis ($22.9 \times 10^9/l$)] and a sedimentation rate of 20 mm/h. The serum calcium was normal. The distribution of proteins on electrophoresis showed an increase in the α_2 fractions, while immunoelectrophoresis showed monoclonal IgG- λ in low concentration. There was no Bence-Jones protein in the urine or depletion of amyloid. The histological appearance was of a combined lytic and sclerotic myeloma.

At this stage she had gross weakness of her forearms and was subsequently unable to dorsiflex her wrists actively, with a strength of MRC grade 1 on the right and grade 2 on the left [9]. Elbow flexion and extension were grade 2 on the right and 4 on the left. Active abduction of the shoulder was normal on the right and limited to 30° on the left. Sensation was severely impaired in the areas supplied by the superficial radial nerve in both hands, by the upper lateral cutaneous branch of the axillary nerve in the left upper arm and the lower lateral cutaneous branch of the radial nerve.

Bone scintigraphy showed increased uptake in the proximal humerus. MRI confirmed that all the major vessels were clear of the tumour and this was confirmed by angiography. A bone marrow trephine excluded dissemination of the disease.

Operation was carried out in August 1995. The radial and axillary nerve were involved in the tumour and had to be sacrificed during resection. A custom-made proximal humeral replacement was inserted (Fig. 2).

The sensory and motor deficits in her left arm did not change, but the symptoms in her right arm were improved. The haemoglobin, sedimentation rate and serum proteins returned to normal.

Discussion

Solitary myeloma are rare and do not usually occur in long bones [17]. Symptoms arise early, but pain is often precipitated by an injury resulting in a delay in diagnosis, which was as long as 12 months in our case.

A mixed lytic and sclerotic myeloma is a rare manifestation of a solitary myeloma and has an uncertain relationship to multiple myeloma [2]. Reports do not dwell on the clinical features [2, 4, 14]. Radiographs show the combined features of solitary and multiple myeloma with expansion and thinning of the cortex and the conglomeration of individual expansive osteolytic lesions with sclerotic margins [4, 13, 19]. Although the scintigram in our case showed increased uptake, false negative results have been reported and the value of the technique is questioned [3].

The neurological defects present in both arms of our patient were proved to be due to a demyelinating peripheral sensory and motor neuropathy which had previously only been described as a complication of osteosclerotic myelomatous lesions [5, 6, 12, 15]. The cause of this progressive neurological disorder is unknown, although an immunological reaction of monoclonal proteins with peripheral nerve structures or the presence of selectively acting neurotoxic substances produced by the tumour [6] have been suggested. The deposition of amyloid has also been considered as a possible cause, but was discarded since amyloidosis is not always present [15]. In our case the neurological symptoms in the opposite arm improved after resection of the tumour and her haemoglobin and serum proteins returned to normal, suggesting that the neurological and blood changes are reversible.

Early diagnosis and treatment of mixed lytic and sclerotic myeloma is essential to prevent the

occurrence of a demyelinating peripheral sensory and motor neuropathy.

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