Amyloid shoulder pads

Two cases of multiple myeloma

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Case 1

A 70-year-old woman presented with a 2-year history of swelling in the shoulders, diffuse arthralgias, low back pain, weakness, loss of appetite and weight loss.

The patient was pale and thin with a distinctive swelling in both shoulders, characterized by the admitting physician as appearing like shoulder pads worn by American football players (Figure 1). The shoulder mass was more prominent on the right than on the left. On both sides, the shoulder masses were firm, immobile, non-fluctuant, non-pulsatile and were minimally painful to palpation. We found a bilateral mild loss of shoulder motion. There was no substantial muscle wasting, but there was weakness at the extremes of motion against resistance.

Serum protein electrophoresis was normal, but urine protein electrophoresis showed a characteristic myeloma pattern and Bence Jones protein in the urine was grossly positive. Shoulder radiographs showed the soft tissue masses in the areas of clinically obvious swelling and diffuse osteoporosis of the humerus.

Open biopsy of the soft tissue mass in the subacromial area of the right shoulder revealed amorphous pink material on Congo Red staining, characteristic of amyloid. Thioflavine staining of an incidentially biopsied vessel in the area of the distribution of the thoracoacromial artery revealed yellow-green birefringency around the vascular channel, typical of amyloid infiltrate.

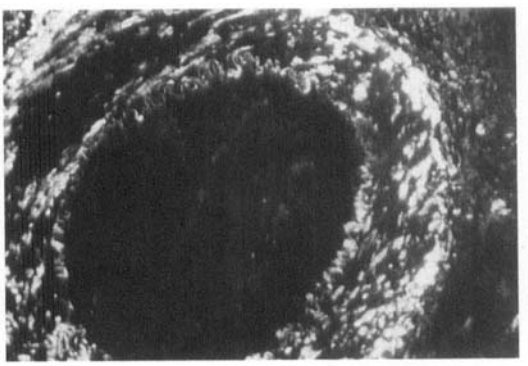
A diagnosis of multiple myeloma with secondary amyloidois was made. 2 years later she succumbed to her illness, at which time bone biopsies at multiple sites confirmed the presence of plasma-cell myelomata.

Case 2

A woman, aged 52, was admitted to hospital with swelling of the shoulders, headache, and pains in the neck and back. 4 years previously one of her ribs had fractured spontaneously and for the past 3 years she had been treated for nephritis because of albuminuria.

Figure 1. Case 1.

70-year-old woman with a shoulder pad sign of multiple myeloma as a result of amyloid deposition in the soft tissues around the joint.



Cross-section of artery in biopsy specimen, stained with thioflavine. Perivascular infiltrates which, in polarized light, have a yellow-green birefringency are characteristic of amyloid.

Examination showed pure and abundant Bence Jones protein in the urine and multiple areas of rarefaction in the skeleton. The disease progressed for another year, when she died of bronchopneumonia.

Postmortem examination showed numerous plasma-cell tumors throughout the skeleton. In addition, there were large yellowish translucent masses distending the shoulder and adjacent bursae (Figure 2). The articular surfaces were normal, but the synovial membranes were grossly thickened by accumulations of amyloid material, which also lined the joint cavities.

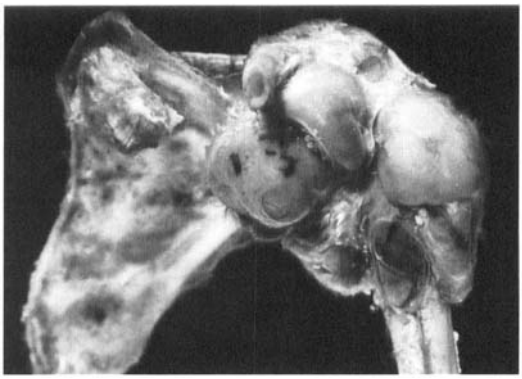


Figure 2. Case 2. Autopsy specimen shows distention of shoulder joint bursae and capsule by amyloid deposits.

Discussion

Amyloid deposition in various body parts is most commonly seen in patients on chronic renal hemodialysis (Ogawa et al. 1987). It is also associated with advanced ageing and with a variety of systemic maladies, especially those accompanied by chronic infection, such as tuberculosis and osteomyelitis, and with disturbances of autoimmune processes, such as rheumatoid arthritis (Scarpelli and Chiga 1985). Moreover, it is found in certain neoplasms, e.g., Hodgkin's disease, medullary carcinoma of the thyroid and, most commonly, in multiple myeloma (Bayrd and Bennet 1950). In renal dialysis patients, the shoulder is most often affected by symptoms associated with amyloid deposition (Chattopadhay et al. 1987, Kurer et al. 1991). However, in the other diseases associated with amyloidosis, involvement around the shoulder is less well known.

Amyloid is an amorphous, insoluble product of protein breakdown which, when present in excess, is pathologically deposited preferentially in bursae, tendons, joints and bones (Noel et al. 1987, Ogawa et al. 1987). It is rarely seen as a primary disease but is more common, as in our cases, as a result of excessive production of immunoproteins or an inability to clear these compounds from the circulation. The excess accumulation in hemodialysis patients is the result of the inability of the cellulose dialysis membranes to remove a particular protein, microglobulin, found on the surface of nearly all cells as the light chain of the major immune histocompatibility complex (Kurer et al. 1991).

The abnormal accumulation of amyloid can affect up to 25 percent of persons with multiple myeloma (Huvos 1979). In our 2 patients, the most striking joint manifestations of their underlying disease were amyloid deposition in the soft tissues and bursae of the shoulder. The shoulder pads mnemonic may be helpful in calling to mind this unusual differential diagnostic possibility. We have subsequently discovered that this analogy to football players' shoulder pads has been made before (Glenner 1980). One must consider other diagnoses which may lead to swelling of the shoulder, such as infection (Leslie et al. 1989), primary bone tumors (Craig and Thompson 1987), rheumatological diseases (Steinfeld et al. 1994) or the bursal effusion arising from extensive underlying rotator cuff disease (Neer 1990). The presentation in each of these circumstances, however, both clinically and in terms of imaging and laboratory studies, will usually demonstrate pathognomonic features different from those in our 2 cases.

Biopsy may be thought necessary, as was performed in case 1, to distinguish between various soft tissue tumors. However, this procedure might have been avoided had the shoulder pad sign of amyloid involvement in multiple myeloma been more judiciously considered.

Acknowledgements

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