Idiopathic haemorrhagic rupture of the shoulder in destructive disease of the elderly

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SUMMARY The cases of two patients with hydroxyapatite crystal associated destructive disease of the shoulder are described who have developed gross haemorrhagic effusions with spontaneous joint rupture and extensive soft tissue damage. No collagenase activity was found in the synovial fluid. Other possible mechanisms of the destructive process are discussed.

Key words: hydroxyapatite, joint rupture, haemorrhage, collagenase.

There have been several reports describing a destructive arthritis of the elderly affecting the shoulders.^{1–7} More recently, reports have documented similar damage to additional joints, most commonly the knees.^{1,8} The radiological changes include severe cartilage and bone attrition with joint instability. A relative lack of regenerative changes in subchondral and periarticular bone has been noted. Rupture of the rotator cuff is common when the shoulder is involved, and areas of articular or periarticular calcification have often been seen.^{1,6}

The synovial fluid is present in often large volume^{1,6} with a low number of white blood cells, predominantly mononuclear cells, but frequently contains red blood cells and is often lightly blood stained.^{1,2,6–8} Collagenase and neutral protease activity has been found to be high in the synovial fluid.^{8,9} Hydroxyapatite crystals have also been detected in the synovial fluid from some of these joints.^{1,8,9}

We describe the cases of two patients with apatite associated destructive arthropathy with shoulder involvement who have developed huge grossly haemorrhagic effusions that have ruptured spontaneously in the absence of collagenase activity in their synovial fluids.

Case reports

CASE 1

The patient was a 78 year old woman. In 1971 at the

age of 64 years she developed a painful left shoulder with no history of trauma. In 1977 she developed painful knees, and in 1980 her right shoulder became symptomatic. There was rapid progression of damage in all four joints from 1980 onwards, and by 1983 there was gross instability of the knees, bilateral ruptured rotator cuffs, large, cool effusions in each joint, and severe pain and disability. Radiographs showed attrition of articular cartilage and bone on both sides of the joint line; there was considerable loss of bone but little sclerosis or osteophyte formation (Fig. 1), as described previously.¹ Large numbers of hydroxyapatite containing particles were detected by alizarin red staining in synovial fluid from both shoulders and both knees,¹⁰ and apatite associated destructive arthropathy was diagnosed.

Synovial biopsy of the left knee showed a papillary synovium with surface fibrin, hyperplastic synovial lining cells, and a patchy mononuclear cell infiltrate including plasma cells. Numerous hydroxyapatite crystals were identified in the superficial synovial tissue by analytical electron microscopy.

In January 1985 she spontaneously developed a red 'raw meat' like area over the anterior aspect of the right shoulder, and within two days there was extensive bruising down the right upper arm (Fig. 2). Right shoulder movement had become very painful. 250 ml of heavily bloodstained fluid was aspirated from the right shoulder. Plain radiographs showed no change from previous films, but arthrography demonstrated rupture of the shoulder joint (Fig. 3). Nineteen days later a tense effusion of the right shoulder developed, from which a further

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150 ml of bloodstained fluid was aspirated, suggesting resealing of the shoulder joint. No collagenase activity was detected in the synovial fluid either before or after gel filtration,¹¹ and β glucuronidase and lactate levels were low. Synovial fluid findings are summarised in Table 1.

Coagulation tests were normal, buffy coat vitamin C was $71.5 \mu\text{g}/10^8$ white blood cells (WBC) (normal range 15–60). Routine haematological and biochemical screening tests were negative, and there was no evidence of intercurrent neurological or other disease.

Fig. 1 x Ray of the right shoulder of case 1.

Fig. 2 Aspiration of heavily blood stained fluid and extensive bruising of the right shoulder of case 1.

CASE 2

The patient was a 75 year old women. In 1978 at the age of 68 she fell, injuring her shoulders, which subsequently became painful. She also developed painful knees. In 1979 she spontaneously ruptured

Fig. 3 Arthrogram of the right shoulder of case 1 demonstrating rupture of the shoulder joint.

Table 1 Synovial fluid findings: characteristics of synovial fluid at the time of shoulder rupture

	Case No 1	Case No 2
Volume (ml)	150	70
Appearance	Heavily bloodstained	Heavily bloodstained
Viscosity	High	High
Clot formation	Negative	Negative
Total white cell count	$<1 \times 10^5/\text{ml}$	$1 \times 10^5/\text{ml}$
Differential	Mainly mononuclear	90% mononuclear
Cartilage fragments (0–3)	1	2
Birefringent crystals	0	0
Alizarin red staining (0–3)	2 (large ovoids)	3 (large ovoids)
β Glucuronidase (OD)	0.20	0.38
Lactic acid (mg/100 ml)*	5.9	2.1
Active collagenase	0	0
Total collagenase	0	0

*SI conversion: $\text{mg}/100 \text{ ml} \times 0.111 = \text{mmol/l}$.

her right shoulder cuff and subsequently the left in 1980. Radiology showed grossly damaged shoulders with large effusions. A Stanmore arthroplasty was performed on the right shoulder, which subsequently dislocated. By 1982 there was gross instability of both knees, with destruction of the lateral compartments and large effusions. Hydroxyapatite containing particles were detected by alizarin red staining in synovial fluid from both shoulders and both knees. Synovial biopsy of the right knee showed some densely cellular synovium, in part due to proliferation of fibroblasts, large multinucleate giant cells, and numerous deposits of hydroxyapatite. The clinical, radiological, and pathological features are therefore very similar to those of the first patient.

In August 1984 she spontaneously developed a large area of soft tissue bruising around the left shoulder with increasing pain on movement, and a large volume (70 ml) of heavily bloodstained fluid was aspirated from the left shoulder. The synovial fluid findings are summarised in Table 1; they include a low total white cell count, numerous alizarin red staining particles, but no collagenase activity, and low levels of β glucuronidase and lactate. Routine haematological and biochemical screening tests were normal, coagulation tests were normal, buffy coat vitamin C was $38.6 \mu\text{g}/10^8 \text{ WBC}$.

The patient was otherwise well. The bruising on the arm became more extensive during the first week and extended laterally to the elbow. The shoulder was reaspirated twice, but after two weeks the pain and bruising slowly subsided.

Discussion

The destructive arthropathy with rotator cuff rupture of the shoulders in these two cases is similar to that previously described and variously labelled 'l'arthropathie destructrice rapide de l'épaule' (AD RE),² 'Milwaukee shoulder',⁶ 'cuff-tear arthropathy',⁷ 'basic calcium phosphate crystal deposition disease',⁸ or 'apatite associated destructive arthritis'.¹ Both patients also had destructive changes of the knees, as has been described elsewhere.^{1,8}

Hydroxyapatite crystals were detected in the involved joints in both patients, as in some of the previously described cases.^{1,8,9} Hydroxyapatite crystals have also been found commonly in synovial fluid from patients with osteoarthritis¹² and pyrophosphate arthropathy,¹³ and in small quantities in normals.¹⁴ The quantity of hydroxyapatite determined by alizarin red staining may be associated with the severity of the radiological changes of osteoarthritis,¹⁰ and heavy staining has been reported in destructive arthropathy.¹ It is unclear,

however, whether this is a crystal induced or associated disease.

Spontaneous haemarthrosis of the shoulder joint has been previously described,^{3,5,7,15} and on review some of these cases may have been apatite associated destructive arthropathy, but synovial fluids were not examined for hydroxyapatite crystals. Gross haemorrhagic effusion with spontaneous rupture of the shoulder has not been previously noted in described cases of apatite associated destructive arthritis, though fluids are often lightly bloodstained.^{1,6,8} Haemorrhagic synovial effusions can also occur in scurvy, but this was excluded in these patients. Similarly, there was no evidence of coagulation defect, neurological disorder, or other intercurrent systemic or articular disease predisposing to the haemorrhagic joint rupture.

A striking feature of these and other described cases is the rapid progression of severe destructive changes in bone, cartilage, and periarticular tissues. The cause of this is unknown. It has been postulated that hydroxyapatite crystals are phagocytosed by synovial lining cells stimulating the secretion of collagenase and neutral protease.^{6,8,16} We therefore assayed these fluids for collagenase activity both before and after gel filtration with a previously described method that detects activity in some rheumatoid synovial effusions.¹¹ No collagenase activity was found in either patient, though three separate samples were assayed from the first patient and two from the second. It therefore seems unlikely that generation of free collagenase was responsible for the joint attrition and rupture. The levels of β glucuronidase and lactate (Table 1) were also low in comparison with the levels of inflammatory fluids.¹⁴ No mechanism is yet apparent on examination of the patients, their synovial fluids, and biopsy material.

Rapidly destructive disease of shoulder and knee joints, in particular, is an important phenomenon in elderly, usually female, patients. These cases show that spontaneous haemorrhagic rupture of the shoulder can occur in this condition. Although rupture is associated with apatite crystals in the effusions, the role of the crystals is unclear. No evidence for collagenase induced destruction was found.

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