Case report

Adult-onset Still’s disease: destructive distal interphalangeal arthritis associated with transient capsular calcification

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SUMMARY A case is described of adult-onset Still’s disease with severe destructive arthritis of the distal interphalangeal joints of the hand, associated with transient capsular calcification.

Adult-onset Still’s disease is an uncommon clinical entity characterised by an evanescent rash, arthritides, high and often spiking fever, and leucocytosis. Lymphadenopathy, splenomegaly, pleuritis, or pneumonitis and pericarditis are other abnormalities frequently found in this disease.1,2 Arthritis more commonly involves large than small joints of the hand and feet. Wrist involvement, with loss of cartilage in the intercarpal and carpometacarpal joints of the wrist progressing to bony ankylosis, has been described as highly suggestive of adult-onset Still’s disease.3

The present report discusses a peculiar type of arthritis found in a young woman with typical adult-onset Still’s disease. Severe destructive arthritis of distal interphalangeal joints of the hand was associated with transient capsular calcification. No evidence was found of the type of disease sometimes associated with capsular calcification in the hand, namely, gout, hyperparathyroidism, CRST syndrome (calcinosis, Raynaud’s phenomenon, sclerodactyly, telangiectasia), or tuberculous arthritis.

Case report

A 22-year old Caucasian woman was admitted to hospital in April 1977 with arthralgia of elbows, wrists, and shoulders, arthritis of some small finger joints, generalised morning stiffness, and muscular weakness. In 1972 she had developed transient arthritis of both knees, with several relapses in subsequent years.

Physical examination revealed anaemia and a generalised, nonitching skin rash. The heart outline was enlarged, without signs of heart failure. Auscultation disclosed no cardiac murmurs or friction rubs, and there was no paradoxical pulse. The second and third distal interphalangeal joints of the right hand were swollen and tender; no other signs of arthritis were detected.

Laboratory tests revealed a Westergren erythrocyte sedimentation rate (ESR) of 65 mm/h, haemoglobin 9.6 g/dl, haematocrit 30%, white blood count 13.5 x 10^9/l with 54% polymorphonuclear leucocytes, 32% bands, 12% lymphocytes, 2% monocytes, and 2% eosinophils; platelet count 578 x 10^9/l, serum creatinine 50 μmol/l, alkaline phosphatase 165 U/l (normal, 120 U/l), aspartate transaminase (SGOT) 17 U/l, alanine transaminase (SGPT) 12 U/l, serum lactic dehydrogenase (LDH) 256 U/l (normal 175 U/l). Rose-Waaler test, latex fixation test, and antinuclear antibodies (ANA) test were negative. A chest x-ray disclosed an enlarged heart but no signs of pulmonary oedema. Echocardiography revealed an echo-free space on the anterior and the posterior sides consistent with pericardial effusion. X-rays of the hands (Fig. 1), feet, ankles, knees, and pelvis were normal.

While in hospital the patient developed transient arthritis of the left wrist and spiking fever (up to 40.2°C) with negative blood cultures. On indomethacin she became afebrile and signs of pericarditis disappeared, whereupon she was discharged.
Adult-onset Still's disease

Readmission to hospital followed in September 1977 in view of unexplained weight loss, generalised lymphadenopathy, and signs of arthritis of the distal interphalangeal (DIP) joints of the fingers. Round the DIP joints a yellowish periarticular thickening was observed. There was no pitting of the nails nor any other sign of psoriasis. X-rays of these joints disclosed a narrowed joint space and soft tissue calcification (Fig. 2), especially around DIP 2 and 3 of the right hand and DIP 2 and 4 of the left hand. A surgical synovial biopsy specimen was taken from the third DIP joint of the right hand. At incision an effusion of greyish white viscous material occurred, apparently coming from the synovial cavity. Bacterial cultures of this material, including those for tuberculosis, remained sterile, and birefringent crystals were not detected by polarised light microscopy.

Histological examination of the synovial biopsy specimen revealed slight signs of chronic inflammation. On the irregularly formed synovial surface a fine to coarse granular material was present, staining dark brown to black with Von Kossa stain, indicating calcium deposits. No granulomas were found, nor were multinucleated giant cells loaded with foamy material as can be found in multicentric reticulohistiocytosis. A biopsy specimen from a right supraclavicular lymph node showed follicular and diffuse hyperplasia without signs of malignancy.

Laboratory studies: ESR 75 mm in the first hour, Hb 9.4 g/dl, leucocyte count 8.8×10^9/l with 22% bands. Serum calcium, phosphorus, creatinine, SGPT, SGOT, C3, C4, factor B, and urinary calcium excretion were normal. Serological studies included a negative Paul-Bunnell reaction and no antibodies against cytomegalovirus and *Toxoplasma gondii.*

![Fig. 1 Normal appearance of DIP, second and third finger of the right hand (4 April 1977).](image1)

![Fig. 2 Capsular calcification and severe narrowing of the joint space (9 September 1977).](image2)

![Fig. 3 Calcific deposits have almost disappeared (23 December 1977).](image3)
HLA typing revealed HLA B27 positivity. Indomethacin therapy was continued and the patient discharged. In the following months arthritis of the PIP joints, wrists, MTP joints, and ankles developed, and signs of pleuritis, pericarditis, hepatomegaly, and splenomegaly became overt, necessitating readmission in December 1977. At that time adult-onset Still’s disease was diagnosed in view of an evanescent rash, polyarthritis, episodes of high spiking fever, and leucocytosis. An x-ray of the hand showed that calcifications round the DIP joints (present in September 1977) had almost disappeared (Fig. 3). Treatment with larger doses of indomethacin (175 mg daily) resulted in gradual improvement of signs and symptoms.

In September 1979 no signs of disease activity were present except for an evanescent rash; the ESR was 5 mm in the first hour, haemoglobin 14·2 g/dl, and leucocyte count 7·9x10^9/l with a normal differential count. No relapse has occurred so far.

Discussion

Adult-onset Still’s disease was diagnosed in our patient in view of the presence of a typical evanescent rash, polyarthritis, high spiking fever, and leucocytosis.1 2 Other abnormalities found, including lymphadenopathy, hepatomegaly, splenomegaly, pleuritis, and pericarditis, were consistent with this diagnosis. Severe destructive arthritis, mainly of the DIP joints, associated with transient large capsular deposits containing calcium, has to our knowledge never been described in adult-onset Still’s disease, nor in any other form of polyarthritis of unknown origin. Capsular calcification in the joints of the hand may be found in a number of conditions, including degenerative joint disease, gout, hyperparathyroidism, CRST (calcinosis, Raynaud’s phenomenon, sclerodactyly, telangiectasia) syndrome and occasionally in tuberculous arthritis.4 Clinical and laboratory data, including normal serum calcium, phosphorus, uric acid, lipids, normal urinary calcium output, the absence of birefringent crystals in the calcium deposits, and negative cultures of the synovial membrane virtually exclude these diagnoses. The arthritis of the small finger joints, especially the DIP joints, was of a particular severe and destructive type, leading within a few months to an almost complete loss of articular cartilage and to an appreciable resorption of juxta-articular bone. It is conceivable that this aggressive inflammatory process produced a large amount of debris, which initially could not be adequately removed by phagocytic cells. Later on, when the arthritis had burned out, the deposits were gradually removed and disappeared. It is of interest that calcification in these deposits developed within a few months at the time of active arthritic disease and in the absence of factors that precipitate soft tissue calcification—for example, hypercalcaemia, hyperphosphataemia, and decreased renal function.

References

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Ann Rheum Dis 1982 41: 544-546
doi: 10.1136/ard.41.5.544

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