

## Medical Memorandum

### A Fatal Case of Still's Disease Complicated by Amyloidosis

The following case is that of a child who died from Still's disease and in whom evidence of amyloidosis was found at the post-mortem examination. This adds to the number of cases described by various writers recently, including Hill (1948), Jennings (1950), and Skelton (1951).

#### Case Report

A girl aged 12 years was admitted to hospital in September, 1948, suffering from rheumatoid arthritis, the sequel to an attack of acute rheumatism in January, when both wrists and both knee-joints were involved. A previous attack of acute rheumatoid arthritis had occurred in 1940, but between the two illnesses her health had been normal. In early childhood she had suffered from whooping-cough and measles, and tonsillectomy had been performed at the age of 2.

The patient was an undersized thin pale girl, and weighed 4 st. 3 lb. (26.8 kg.). The chest was poorly developed. There was no cardiac enlargement and the heart sounds were normal. The pulse rate was 100 and the blood pressure 136/86. The lower edge of the liver was palpable. There were stiffness and swelling of the right elbow-joint with limitation of extension, and swelling and reduction of movements were present in both wrist-joints. The knee-joints were enlarged and globular in outline and very stiff. An x-ray examination of the hands showed osteoporosis of the bones and diminution of the carpo-metacarpal joint spaces. The E.S.R. was 66 mm. in one hour (Westergren). The red cells numbered 4,300,000 per c.mm., and the white cells 7,800; the haemoglobin was 75% (Sahli). The urine was normal. In December, 1948, the E.S.R. was 70 mm., the liver was enlarged one fingerbreadth below the costal margin, and the spleen was palpable. There was no fever, and less pain was complained of in the affected joints.

In March, 1949, the joints became stiffer and more painful, and the E.S.R. was 110 mm. During the summer there was a gain of 4½ lb. (2 kg.) in weight, but in the autumn an exacerbation of joint symptoms took place, the liver and spleen showed further enlargement, and there was a sharp epistaxis in October. In November, 1949, generalized enlargement of the lymph nodes was noticed, and the blood count showed: red cells, 3,200,000 per c.mm.; white cells, 11,900; and haemoglobin, 7.7 g.% (Sahli). No albumin was noted in the urine. Several attacks of vomiting and a further epistaxis occurred in December, 1949, when her weight was 3 st. 1 lb. (19.5 kg.).

A lymph node was removed for histological examination and reported upon as follows: "There is follicular hyperplasia with amyloid infiltration in many of the germinal centres. The sinuses are filled with catarrhal cells and show also small haemorrhages and haemosiderin. In the medulla there are numerous clumps of polymorphs and plasma cells, but no organisms were seen. The appearances are considered to be not incompatible with findings reported in Still's disease."

During January, 1950, there were attacks of vomiting and diarrhoea. The plasma proteins were 7 g.% and the icteric index 7. The Takata-Ara test was weakly positive, and the thymol turbidity test gave 5 units. During February the patient became more feverish and suffered from attacks of epigastric pain and further diarrhoea, due to Sonne dysentery infection. Her condition steadily deteriorated, and death occurred on February 9, 1950.

*Post Mortem.*—The body was that of an undersized and extremely emaciated child. Movement of the limbs and head was limited by arthritic changes. Some enlarged

lymph nodes could be felt, and there was a firm homogeneous mass in the upper part of the abdomen. The tongue, larynx, oesophagus, and trachea were extremely dry; the lungs were pale, and bound to the chest by numerous adhesions. No fluid was found in the pleural cavities. The hilar nodes were enlarged and elastic. The pericardial sac contained a small quantity of clear fluid. The heart was small and pale. No valvular defects were noticed, and the heart muscle did not give any reaction with iodine. The liver was very large and pale, and weighed 2,030 g. The cut surface had the appearance of amyloid after staining with iodine. The stomach was small, the mucous membrane pale. The small intestines were distended and purple, and contained blood and mucus. The large bowel presented a similar appearance. The kidneys were small and gave the reaction for amyloid. The suprarenals were large and pale, and were considered to contain amyloid. The spleen weighed 230 g. and gave a doubtful amyloid reaction. The para-aortic glands were enlarged and of an indiarubber consistence, and the mesenteric glands were dark in colour.

#### Discussion

The most pronounced amyloid changes were found in the liver, kidneys, suprarenals, and lymph nodes. The persistent vomiting, diarrhoea, and haemorrhage were important factors in bringing about the fatal termination. The urine was free from albumin throughout the illness. A certain degree of anaemia was present in November, 1949, and, since the blood picture suggested malabsorption as the cause, the patient was treated with liver. The value of liver therapy in amyloidosis has been shown by Grayzel and Jacobi (1938), but benefit is not invariable, and in this case the treatment was not effective. Reimann and Eklund (1935) state that amyloidosis appears only after a protracted illness, and that all the cases described have been emaciated and anaemic. Death in this particular case was due to terminal dysentery, and the amyloidosis was not by itself responsible. It was evident that the rheumatic process was never properly arrested—a fact which no doubt prevented regression of the amyloid changes.

Recovery from this condition has been noted by Trasoff *et al.* (1944), who have found albuminuria present in nearly all the described cases. Carroll and Nelson (1927) emphasize the gravity of its occurrence and say that it is always preceded by severe or moderately severe arthritis of some duration. Hill (1948) states that most of the published cases refer to children. It is suggested that in the present case the amyloid changes found in the liver, kidneys, and lymph nodes were due to the rheumatic infection and that the numerous pleural adhesions were also probably of rheumatic origin, since no evidence of tuberculosis could be found in the body at necropsy.

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