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## Juvenile Rheumatoid Arthritis (Case Report)

by

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### Introduction

Juvenile Rheumatoid Arthritis (JRA) or usually also called Still's disease, is a chronic disease occurring in children with the clinical signs and symptoms of joint involvement and systemic manifestation. It has many similarities with adult rheumatoid type, but also distinct differences.

There are some criteria to diagnose JRA clinically, e.g. polyarthritis affecting more than 4 joints and a chronic course of at least 3 months. If less than 4 joints are involved, a synovial membrane biopsy must be done revealing changes of JRA whereas other articular or connective tissue diseases should be excluded (Ansell, 1966).

Practically all cases of adult rheumatoid arthritis and most cases of JRA have a predominance of joint manifestations over systemic involvement. However, in about 20% of the

cases, JRA has severe systemic manifestations with only very mild or even absence of joint complaints (Calabro, 1971).

JRA has variable signs and symptoms depending on how the disease begins. There are 3 modes of onset with its own clinical picture, namely:

1. polyarticular onset,
2. monoarticular onset,
3. acute febrile onset.

About 50% of JRA cases have a polyarticular onset (used to be called adult type) and 30% have a monoarticular onset; both have only mild or moderate systemic manifestations but the joint involvements predominate. The joint involvements vary from stiffness, mild pain, limp in walking to various enlargements with severe pain, redness, warm feeling and limited motion.

Among the joint lesions that are only found in the JRA and usually

absent in the adult type, are the involvements of cervical vertebrae and the fusiform swelling of the interphalangeal joints replacing the usual larger joint involvement. The head is usually fixed in a rather stiff position and is projecting forwardly.

In the acute febrile onset, the systemic manifestation predominates and the joint involvement is only very slight such as mild arthralgia without apparent swelling. The systemic manifestations consist of restlessness, loss of appetite, fever, tachycardia not proportional to the fever, skin rash (usually appearing and disappearing quickly in several hours), lymphadenopathy, splenomegaly, heart involvement (usually in the form of pericarditis), renal involvement and eye complication.

In the beginning of the disease, the fever is usually rather characteristic due to its quotidian type (fever increases to its peak once or twice daily followed by sudden drop to normal temperature).

The iridocyclitis of the eye is mostly found in the monoarticular form. The laboratory examinations are not specific, except for the detection of the rheumatoid factor (also called anti gamma globulin) by "sheep cell agglutination test" or "latex agglutination test", and the detection of "anti nuclear antibodies". Unfortunately in JRA only about 20% are positive to those rheumatoid factor tests as compared with 80% in the adult type, and only 19% are positive

to the antinuclear antibodies test (Rothfield, 1971). But both of these factors are not only positive in JRA, but also in some other diseases. There is a newer and most sensitive method for this rheumatoid factor test (with up to 94% positiveness), e.g. the Torrigiani's method (Torrighiani, 1967).

Rontgenological pictures of the joints are not characteristic, except in the advanced stage which may become helpful. The articular surface erosion and irregularities, the obliteration of the joint space and fibrosis or bony ankylosis may be detected. Steinbrocker et al. (1949) divided the JRA into 4 stages consistent with the radiological abnormalities. In the early stage (first stage) there are no destructive changes, only osteoporosis may be present and in the fourth terminal stage fibrosis or bony ankylosis may be obvious.

Concerning the prognosis, Laaksonen (1966) stated that 70% had a complete recovery, 10% became bedridden and 20% had considerable limited motion. JRA with an earlier onset has usually a better prognosis than with a later onset.

### Case report

#### Case I :

N, an Indonesian boy of 2½ years, was admitted for the first time to the Department of Child Health of the Dr. Tjipto Mangunkusumo General Hospital on August 8, 1972, with the following history.

Since 8 months he has been suffering from fever with cough; the fever lasted for about 2 weeks during which he became limp. Two months later the right wrist became swollen and the proximal interphalangeal joint of the third digit of the right hand enlarged fusiformly; the swelling disappeared but not completely within 2 weeks. Since then the fever and joint swelling appeared periodically even on treatment by the general practitioner. Both knees were involved 1 month prior to admission, and a week before admission both wrists, both ankles and the cervical vertebrae were attacked.

He was the fifth child of a family of 6 children and nobody else had the same disease. Contact with tuberculosis was denied.

Physical examination on admission revealed a 2½-year-old boy with a body weight of 10 kg, a body length of 80 cm, a temperature of 38.9 centigrades, a pulse rate of 108/min. and a respiration rate of 20/min. He was conscious, and did not look seriously ill. The head was held slightly forward and was painful on movement. Both wrists, knees and ankles were swollen, tender, not warm, and of normal color; there was no limitation of movement.

The proximal interphalangeal joint of the third digit of the right hand was swollen fusiformly. Nose and pharynx were hyperemic; tonsils were slightly hyperemic but not enlarged. Heart and lungs were normal.

Liver and spleen were not palpable. Edema was not present. Both palms showed palmar erythema.

Laboratory: Mantoux tests of 1:2000 A.T. and 5 T.U. PPD were negative but later on a repeated Mt 1:100 test gave a positive result (15 mm in diameter); BCG scar was not detected. Three times A.S.T.O. measurements were within normal limits. Hemoglobin content was 8.6 gm%, wbc was 7000/cubic mm with a differential count of eos. 0/stabs. 5/segm. 35/lymph. 56/mono. 4. Blood sedimentation rate was 55 mm in the first hour and 95 mm the second hour.

Thorax photo revealed a normal heart, small perihilar and retrocardial infiltrates were visible. Electrocardiogram showed only sinus tachycardia (QRS rate of 144/min). Blood culture was sterile. Throat swab culture revealed Pneumococcus and Staphylococcus anhemolyticus. No acid-fast bacilli were found in the gastric juice. Serum Iron was 72.72 gamma%, I.B.C. 150 gamma%. Bone marrow showed a hyperactive erythropoietic system. Repeated blood LE. cell examinations were negative. Uric acid (4.1 mg%), albumin (3.15 gm%), and globulin (2.94 gm%) concentrations in the blood were within normal limits. No abnormalities of the urine and stool were detected.

X-Ray of the joints revealed only periarticular swelling. Based on all above mentioned data, the di-

agnosis of JRA with Pulmonary tuberculosis was established.

The treatment consisted of aspirin, sodium bicarbonate (to neutralize the aspirin acidity), triple tuberculostatics, sulfas ferrosus, roborantia and adequate feeding. Follow-up revealed that the fever and joint swelling subsided gradually after about 2 weeks of treatment, but the fusiform painless swelling persisted. After the clinical symptoms had subsided the dose of aspirin was decreased rather quickly, and in one week the joint swelling reappeared, but could be overcome promptly by increasing the dose again to the initial amount. Ophthalmic, E.N.T. and dental examinations did not reveal any focal infection or complication.

The boy was discharged after 2½ months of hospitalization in a good condition except for the persistent painless interphalangeal fusiform swelling.

#### Case II :

T.J.N., a Chinese girl of 12 years, was admitted for the first time to our hospital on June 30, 1972, with the following history.

The girl got the first attack of joint affection 5 years ago. There was swelling of both knees and wrists with fever followed by both ankles' enlargement. During remission those joints remained enlarged and restricted in motion. The present attack was the third and occurred a week before admission. She got moderate fever, both knees and wrists became

acutely swollen and she had difficulty in breathing. Slight edema in both legs occurred, since 3 days before admission associated with oliguria. She was the fourth child of a family of 4 children and nobody else suffered from the same disease.

On admission physical examination revealed a 12-year-old girl with a body weight of 24 kg, a body length of 135 cm, a temperature of 37.2 degrees Celsius. Pulse rate = heart rate = 124/min., the pulse was equal and regular. Blood pressure was 100/60, respiration rate 24/min. She looked ill, was conscious and not dyspnoeic. Nose and pharynx were hyperemic. Tonsils were normal. Jugular venous pressure (JVP) was 5 + 0, hepatojugular reflux (HJR) was positive. The lungs were normal, the heart was slightly enlarged, ictus cordis was in the 5th intercostal space and left midclavicular line. Heart sounds were normal, but a pansystolic murmur, grade II, was heard with the punctum maximum at the apex and radiating over the whole heart and back. Ascites was absent.

The liver was enlarged 1/2 — 2/3 with a blunt edge and was tender on palpation. The spleen was palpable and enlarged to Schuffner I. Pretibial edema was present. Both wrists and ankles were swollen, rather bluish in color, painful on moving and very limited in flexion and extension. Ulnar deviation of the fingers of both hands was very obvious.

Laboratory examinations revealed negative Mantoux tests (Mt 1/2000 A.T., 1/100 A.T. and PPD 5 TU). Hemoglobin content was 7.4 gm%, reticulocytes 1.6‰, WBC 15,800/cubic mm, diff. count: eos. —/stabs 2/segm. 75/lymph. 21/mono. 2. E.S.R. was 79 mm/96 mm, ASTO 500 TU. Peripheral blood smear showed anisocytosis, poikilo- and fragmentocytosis, but target cells were not detected.

Urine and stool were normal. Blood culture was sterile, throat swab culture revealed *Streptococcus anhemolyticus* and *Hemophilus* group. The electrocardiogram showed enlargement of the left ventricle (voltage criteria) with intraventricular conduction disturbances. Thorax photo showed slight enlargement of the heart with slight hilar and retrocardial infiltrates. Malaria was negative, uric acid 3.1 mg%, albumin 3.37 gm%, globulin 3.05 gm%. L.E. cells were not found, Hb. F. 3.28%. Bone marrow showed a slight hyperactive erythropoietic system. Examination of the mother's blood: Hb. F. 2.94%.

X-ray photos of the joints showed periarticular swelling and osteoporosis. The diagnosis was JRA, heart failure e.c. Rheumatic heart disease, and *Thalassemia minor*. The treatment consisted of Penicilline for 10 days followed by long acting Penicilline monthly, aspirin, low salt diet, diuretics (Lasix) and bedrest. Edema disappeared within one week, the liver decreased in size

(1/4 — 2/3), whereas the HJR and increased JVP were not seen any more. The fever (quotidian type) subsided within a weeks. The heart murmur changed from pansystolic to systolic murmur, which later on disappeared completely after about 2 months of treatment.

On the 8th week of hospitalization the ESR returned to normal, and the ECG changes disappeared. The wrist and ankle joints were still rather swollen and limited in movement despite of regular and intensive physiotherapy.

The patient was discharged after 2½ months of hospitalization, and maintained on the same dose of aspirin and monthly long acting Penadur. E.N.T. examination did not reveal any focal infection, but the Dental Department had to extract several decayed teeth. No eye complication was detected.

### Discussion

JRA is not commonly encountered in the Department of Child Health, Dr. Tjipto Mangunkusumo General Hospital. To the authors' knowledge no single report is found in the *Pædiatrica Indonesiana* previously.

In this paper 2 cases of JRA are reported and the diagnosis was based on the clinical signs and symptoms and by excluding other possible causes of arthritis (especially rheumatic fever, other systemic collagen diseases, gout, infectious arthritis and leukemia). Unfortunately,

rheumatoid factor and "anti-nuclear antibodies" tests could not be done due to lack of facilities, but these tests are not absolutely needed in establishing the diagnosis.

In the first case the onset of the disease was at the age of 2 years, when JRA has the greatest incidence (the peak is - 4 years and 9 - 14 years, Laaksonen 1966). Rheumatic fever starts very seldom before the age of 5 years, although some cases may be found starting before the age of 3 years. Dieterlen (1971) found 3 cases of Rheumatic fever below 3 years of age but with obvious signs of carditis, some with heart failure and increased titre of A.S.T.O.

Next to age onset, the symmetrical joint involvements of long duration (more than 3 months), fusiform swelling of the proximal interphalangeal joints, the cervical vertebrae involvement, the normal value of A.S.T.O., the normal heart and electrocardiograms are all supporting the diagnosis of JRA instead of Rheumatic fever.

Other important examinations to exclude other possibilities of etiology have been performed with negative results (leukemia, systemic lupus erythematosus, gout, tuberculosis and infectious arthritis).

The JRA in the first case had the monoarticular type of onset. The active pulmonary tuberculosis was based on the positive Mantoux test without previous BCG vaccination,

and the presence of lung abnormalities in the X-ray photo.

By proper treatment this patient improved clinically, except for the slight painless interphalangeal fusiform swelling, and was discharged after 2½ months of hospitalization.

After one month of ambulatory treatment, the interphalangeal swelling disappeared completely. Until now, about 5 months after discharge, there has been no exacerbation of fever or joint attack, ophthalmic complication, heart involvement or leukemic changes. These leukemic changes must not be overlooked since Schaller (1972) published a series of 13 cases consisting of 11 cases suspected as JRA based on clinical and laboratory examinations, one case of Acute Rheumatic fever, and one case of S.L.E. which later on showed leukemic changes in the blood pictures.

In the second case, the diagnosis was more complicated. However, JRA is most likely justified here due to the presence of long duration and repeated attacks of symmetrical joint swellings (polyarticular type of onset) with sequelae, ulnar deviation of digits of the hand which was in-correctable even after serial physiotherapy. The possibility of Rheumatic fever coincidentally occurring simultaneously is very great in this patient due to several reasons e.g. the presence of right heart failure, the long persisting murmur, ECG changes and the increased A.S.T.O.

The right heart failure was based on the presence of murmur, enlarged heart, increased venous pressure and the positive hepato-jugular reflux, liver enlargement with blunt edge, edema and dyspnea.

The murmur lasted for about two months and this is very suggestive of Rheumatic fever, though in JRA a murmur can sometimes also be heard but only transiently (functional murmur).

Moreover in JRA the heart complication is mostly in the form of pericarditis. Calabro (1971) stated that if a heart murmur lasted for more than 2 months, then JRA as the cause could be excluded. The X-ray pictures of the joints of this patient did not show advanced abnormalities of JRA, except osteoporosis and periarticular tissue swelling (stage I). The osteoporotic process may be due to JRA or Thalassemia or both.

The diagnosis of Thalassemia was due to the presence of anemia, reticulocytosis, signs of increased destruction of red blood cells in the peripheral blood (aniso-, poikilo-, fragmentocytosis), hyperactive erythropoietic system in the bone marrow and splenomegaly. The HbF measurement confirmed this diagnosis (HbF of the child was 3.28% and that of the mother 2.94%, whereas normal values in children and adults are below 0.5%; Nelson, 1969). In this patient, the Thalassemia minor does not need any treatment yet.

There is a joint abnormality in Rheumatic fever that clinically resembles that which is found in Rheumatoid arthritis, and which used to be called Jaccoud's arthritis, i.e. repetitive attacks of migratory polyarthritis, the presence of Rheumatic fever, recovery from the attack with minimal residual metacarpophalangeal joint lesion as an insidious painless appearance of correctable deformity of the joint (usually flexion and ulnar deviation) and the absence of Rheumatoid factor.

So, the persistent wrist joint residual abnormality in this case which was uncorrectable, seemed not to be a Jaccoud's type arthritis. Until now (for about 6 months) follow up of this second case does not show any sign of exacerbation, but the wrist joint swelling and limited motion still persist.

### Summary

Juvenile Rheumatoid Arthritis (JRA) is not commonly found in our pediatric clinic, and this is supposed to be the first report in the Indonesian pediatric literature.

In this report 2 cases of JRA are presented i.e. one is complicated with pulmonary tuberculosis and the other with Thalassemia minor and heart failure. The latter is very suggestive of a simultaneous appearance of Rheumatic fever. The literature of JRA is also briefly discussed. Unfortunately, Rheumatoid factor can-

not be examined in our clinic up to now due to lack of facilities.

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#### REFERENCES

1. ANSELL, B.M.: Still's disease. Progress in clinical Rheumatology (Churchill, London 1965) as cited by Laaksonen (1966).
2. BURDA, C.D.: Chronic Post-rheumatic fever (Jaccoud's arthritis). Arch. intern. Med. 120 : 712 (1967).
3. BYWATERS, E.G.L.: The relation between heart and joint disease including "Rheumatoid Heart disease" and post Rheumatic arthritis (type Jaccoud). Brit. Heart J. 12 : 101 (1950).
4. CALABRO, J.J.: Rheumatoid arthritis. Clin. Symp. 23 : 1 (1971).
5. DIETERLEN, M.: Three cases of Rheumatic fever of the small children. Pediatrics. 26 : 1 (1971); cited from Excerpta med. 8 : 2 (1972).
6. LAAKSONEN, A.L.: A Prognostic study of JRA. Act. ped. scand. Suppl. 166 (1966).
7. NELSON W.D.: Textbook of Pediatrics. 9th ed. (Saunders, London 1969).
8. ROTHFIELD, N.F.: Diagnosis of Lupus Erythematosus and Rheumatoid Arthritis in children. Ped. clin. N. Am. 18 : 1 (1971).
9. SCHALLER, J.: Arthritis as presenting manifestation of malignancy in children. J. Ped. 81 : 3 (1972).
10. STEINBROCKER, O., TRAEGER, C.H. and BATTERMAN, R.C.: Therapeutic criteria in Rheumatoid arthritis. J. Amer. Med. Ass. 140 : 659 (1949).
11. TORRIGIANI, G., ANSELL, B.M., CHOWN, E.E.A. and ROITT, I.M.: Raised IgG antiglobulin factors in Still's disease. Ann. Rheum. 10 : 83 (1967) as cited by Rothfield (1971).
12. ZVAIFLER, N.J.: Chronic Post-rheumatic fever (Jaccoud's Arthritis). New Engl. J. med. 267 : 10 (1962).