Discussion

On the average, normal children walk alone when 13 months old; 90% of them achieve independent walking before 15 months. Children with MLD of late infantile type seem to sit, stand, and begin to walk with support like normal children, but they walk unaided considerably later than normal children. Fewer than 60% of them walk by 16 months of age, and 15% never achieve independent walking. This finding seems to have been overlooked in previous reviews of MLD. MLD is a degenerative disease of the nervous system in which sulpholipide begins to accumulate in various organs during intrauterine development, as has been shown by electron microscopical examination in affected fetuses after therapeutic abortion. Sulpholipide accumulation in peripheral nerve was demonstrated by biopsy shortly after birth in an affected case, and slowing of the motor nerve conduction velocity was documented at age 8 months in the same child. These observations support the impression that the first detectable sign in MLD is the early peripheral neuropathy. Therefore, it is not surprising that a delay in independent walking, most probably secondary to the peripheral neuropathy, is the first symptom of the disease in most children, as this study shows. Peripheral nerve motor conduction velocity measured at this early stage should have begun to show a significant reduction.

From the observation of 6 cases, and the review of 61 others, it seems that any description of the typical course of late infantile MLD should include a significant delay in independent walking. This delay may be the earliest clinical manifestation of the disease.

*Additional data on these 61 cases is available from the authors.

References


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Rubella and juvenile chronic arthritis

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SUMMARY A 9-year-old boy with a clinical illness similar to juvenile rheumatoid arthritis was found to have rubella virus in the synovial fluid. There was complete remission of symptoms after 3 months. The role of rubella virus as a possible aetiologcal agent in juvenile rheumatoid arthritis is discussed.

The systemic form of juvenile chronic arthritis (JCA) presents diagnostic difficulties. Similar clinical illnesses with arthritis have been described in association with coxsackie and adenovirus infections. Naturally-acquired rubella infection may be complicated by a polyarthritis although this is rare in children; it occurs more often in women. We describe a 9-year-old boy with a clinical history that suggested systemic onset JCA and in whom rubella virus was isolated from the synovial fluid.

Case report

A previously healthy 9-year-old white boy was admitted to hospital with a 3-week history of sore throat followed by high fever, muscle pains, and neck
and back pain. A 7-day course of penicillin had not affected the disease. Examination showed a fluctuating pyrexia, a patchy macular rash, stiff back and neck, a loud ejection murmur, and splenic (6 cm) and hepatic (2 cm) enlargement.

Preliminary investigations showed: erythrocyte sedimentation rate 69 mm in 1st hour (Westergren), mild polymorphonuclear leucocytosis, ASO titre 350 U/ml, normal chest x-ray film, and normal electrocardiogram. The disease persisted despite adequate doses of aspirin and he was transferred to Guy’s Hospital. Further investigation showed: IgG 13 g/l, IgA 1·1 g/l, and IgM 3·3 g/l. The serum rubella-specific haemagglutination inhibition titre was 160, 14 days later it was 320. ASO titre was 150 U/ml.

The following investigations produced normal results: rheumatoid factor, anti-nuclear factor, Mantoux test, renal scan, whole body computerised tomography scan, and urine and blood cultures. The eyes were normal on slit-lamp examination.

A presumptive diagnosis of JCA was made and aspirin restarted. The symptoms of pronounced pyrexia, episodic rash, and hepatosplenomegaly were unchanged. Three weeks after transfer two dental abscesses were treated by extraction and drainage under antibiotic cover, with no subsequent effect on the clinical illness. Nine weeks after the onset of symptoms he developed an inflamed painful left knee, with a moderate effusion. Rubella virus was grown from the synovial fluid; the histology of the synovial membrane was normal. After aspiration the effusion did not recur. One week later the fever settled and there were no further symptoms. The hepatic and splenic enlargement resolved during the next month and the patient has been well for the last 12 months.

Discussion

This patient illustrates the difficulty in diagnosis which may occur when systemic onset JCA is considered. A protracted illness, associated with arthritis, is most unusual in natural rubella infection. Also, the arthritis associated with rubella tends to occur either before the rash or within 3 days after the eruption.9

The possibility that rubella infection may play an aetiological role in JCA has been suggested by Ogra et al.,4 who observed rubella virus antigen in sediment smears of the synovial fluid in 33% of affected patients. Linnemann et al.5 reported raised rubella antibody titres in their series of patients with JRA, but suggested that this was a non-specific manifestation of increased immunoglobulins. Patients who develop JCA may have defective cellular immune responses. Jennings8 studied T-lymphocyte function and divided patients into two groups, those with normal responses and those with a significant reduction in T-lymphocyte function. The latter group presented mainly with polyarticular disease. Höyeraal7 observed impaired delayed hypersensitivity in his series and went on to describe humoral immunity in the same group.8 When compared with healthy controls, patients were found to have a normal primary immune response to brucella antigen and an increased secondary response to immunisation with diphtheria and tetanus vaccines. Therefore, while humoral immunity appears more than adequate, cellular immune responses may be deficient; an inverse relationship between the cellular and humoral immunity mechanisms has been suggested.9

It is not known whether illnesses initiated by viruses lead to JCA. We suggest that vigorous attempts to isolate viruses should be made in the early stages of illnesses of this type. Careful documentation of the follow-up of these patients will increase our knowledge of the natural history of this problem and may provide valuable clues to the aetiology of JCA.

References


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