abnormality with perhaps secondary changes. The late onset of many of the reported cases could be ascribed to superimposed stresses, such as ageing producing a critical diminution of functional reserves in the cerebral circulation. It is not clear whether the network of collaterals is primarily a part of the initial malformation or a response to the progressive narrowing of the carotid system. The significance of the intimal thickening, the only consistent histological feature, is also not clear.

Other theories have even less to support them. These include a response to trauma or inflammation, the possibility of an autoimmune process, or a relation to atherosclerosis or hypertension.

**Summary**

A case is described of an Australian boy with moyamoya associated with peripheral vascular involvement manifest as severe chilblains since infancy. He developed an acute hemiplegia at age 13. Despite intensive investigation, no underlying cause was found for this condition. The early onset and diffuse occurrence of vascular disease gives some support to the theory that there is a primary abnormality of the blood vessels.

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**References**


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**Fatal septicaemia due to Moraxella non liquefaciens**

The organism *Mima polymorpha* or *Moraxella non liquefaciens* is known to cause serious infections. The members of the genus *Moraxella* are similar to *Neisseria* in morphology, biochemical reactions, growth characteristics, nutrient requirements, antibiotic sensitivity, and habitat, and there has been controversy regarding its taxonomy and bacterial sensitivity. We have encountered a case of *Moraxella non liquefaciens* septicaemia with bilateral adrenal haemorrhages clinically resembling meningococcaemia.

**Case report**

A 5-month-old boy, born after term normal delivery with birthweight 3355 g to healthy parents, was admitted to hospital with a history that he was unwell with coryza for one week. He had been refusing feeds, and became drowsy, lethargic, and febrile one day before admission.

At the age of 9 weeks he had been hospitalized with an attack of fulminating bronchopneumonia and was critically ill; he had been treated with ampicillin and recovered completely in 10 days. Subsequently he had two episodes of upper respiratory tract infection and an attack of croupy cough. These illnesses were treated by the family doctor and required no hospitalization.

On examination he was found to be endomorphic in build, weight 13.5 kg, temperature 39.5°C, pulse rate 168/min, respiratory rate 35/min. He was pale, peripherally cyanosed, drowsy, and dehydrated, irritable on waking and showed no interest in feeds. Mucoid secretions were seen with crusting at the nostrils, there was no skin rash and no lymphadenopathy. The liver and spleen were not palpable. Cardiovascular and respiratory systems were normal. No abnormal neurological signs were elicited. Hb 10.6 g/100 ml; packed cell volume 34%; mean corpuscular haemoglobin concentration 35%; platelets 200,000/mm³; RBC 4 million/mm³; WBC 3600/mm³; polymorphs 9%; lymphocytes 89%; monocytes 2%. Peripheral smear showed normochromia with mild variation in size. Plasma sodium 139 mEq/l, chloride 104 mEq/l, potassium 4.9 mEq/l, urea 59 mg/100 ml, creatinine 0.8 mg/100 ml; blood pH 7.13; Po2 97.6 mmHg, PCO2 51.8 mmHg; buffer base 33, standard bicarbonate 16, base deficit —13 mEq/l. Chest x-ray showed overdistension of the lung fields with no areas of consolidation or collapse. CSF was slightly blood-stained with 1560 RBC/mm³, lymphocytes 3/mm³; protein 36 mg/100 ml; sugar 88 mg/100 ml; culture was negative. Urine and blood cultures grew Gram-negative organisms after 48 hours incubation, identified as *Moraxella non liquefaciens* sensitive to penicillin, ampicillin, tetracycline, streptomycin, and cephaloridine.

**Bacteriological findings.** A nonmotile Gram-negative coclobacillus was isolated after 48 hours' incubation at 37°C from each of two blood cultures containing cooked meat medium (oxoid) plus 0.05% liquoid. The properties of the organism were as follows. On horse blood agar, nonhaemolytic colonies aerobically, but no growth anaerobically. On MacConkey agar, no
growth. On Hugh and Leifson's O-F medium with 1% glucose, no growth. Serum water sugars showed glucose fermentation with production of acid but no fermentation of lactose, sucrose, or mannitol. Results of other biochemical tests were oxidase positive; catalase negative; nitrate reductase negative; gelatin liquefaction negative; indole production negative; citrate utilization negative. The media were prepared and tests carried out according to Cowan and Steel (1965).

Management. An intravenous drip was inserted with 0.45% normal saline and 2.5% dextrose to correct the dehydration and 40 mEq sodium bicarbonate was given intravenously to correct acidosis. Crystalline penicillin 500,000 units 3-hourly and sulphadimidine 350 mg 4-hourly were given also intravenously.

Over the next 2 hours the child's general condition deteriorated and he developed a maculopapular rash over the chest; the lesions progressed rapidly in size and number, and within the next hour he became cyanosed with confluence of the rash and went into a state of shock. A provisional diagnosis of meningococcaemia was made and he was given heparin intravenously (100 mg/kg), hydrocortisone 150 mg 6-hourly, and 200 ml stable plasma protein solution intravenously to combat shock. The prothrombin time was 16 sec (control 12 sec), prothrombin index 74%, partial thromboplastin time 95-5 sec (control 55 sec), and the platelet count dropped to 56,000. The F1 latex fixation test for fibrin split products titre was 1:16, and the haemagglutination inhibition test for fibrin split products titre was 323 μg/ml, which was abnormally high. The serum immunoglobulins were IgA 20 mg/100 ml; IgG 210 mg/100 ml; IgM 50 mg/100 ml. He deteriorated rapidly, the whole body being covered with blotchy haemorrhagic lesions, he became hypothermic, and died within 9 hours of admission.

Necropsy revealed congestion of the larynx, trachea, and the main bronchi with no consolidation of the lungs. Both kidneys showed some fetal lobulation. The adrenal glands each weighed 5 g and were extremely haemorrhagic. The CSF in the subarachnoid space was turbid, but examination of the brain after fixation revealed no abnormality. Histologically, the spleen was extremely congested, the adrenal glands were autolysed and haemorrhagic and the lymph nodes were congested and showed reactive hyperplasia.

Discussion

The clinical course of this child suggested meningococcaemia with Waterhouse-Friderichsen syndrome. Though the organism isolated was sensitive to many antibiotics in vitro, including those given on this occasion, the clinical course was not altered. Burrows and King (1966) and Townsend, Hersey, and Wilson (1954) have described cases of Waterhouse-Friderichsen syndrome due to Mima polymorpha. The work of Tracey and Moir (1960) has shown differing antibiotic sensitivities. It has also been shown that various organisms identified as Mima polymorpha may not be identical (Christensen and Emmanouilides, 1967; Townsend et al., 1954). There has been much confusion regarding classification of this organism. Pickett and Manclark (1965) believe that this organism should be classified as Moraxella duplex or Moraxella non liquefaciens. It has also been suggested that oxidase-negative Mima polymorpha is a member of the genus Achromobacter. However, either Moraxella duplex or Moraxella non liquefaciens seems to be the name of choice. Irving and Herrick (1967) have discussed the clinical significance and means of identifying these organisms. Christensen and Emmanouilides (1967) have reported bacterial endocarditis due to Moraxella in a case of ventricular septal defect. None of the case reports so far have correlated the severity of the infection with any immunological deficiency state.

Summary

A fulminating septicaemic illness in a 5-month-old boy was due to infection with the Gram-negative organism Moraxella non liquefaciens and, despite treatment with penicillin to which the organism was sensitive, the illness was rapidly fatal.

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References


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