CONGENITAL LEUKAEMIA IN DOWN'S SYNDROME

by

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Kelsey and Anderson (1939) reviewed the literature on congenital leukaemia and accepted 9 authentic cases. Merrit and Harris (1956) accepted 21 cases. There is a distinct association between leukaemia and Down's Syndrome (Mongolism). According to Krivit and Good (1957), the frequency of leukaemia in Down's Syndrome is at least 3 times greater than chance alone would explain. It is of interest to recall that 3 of the 21 cases of congenital leukaemia, reviewed by Merrit and Harris, occurred in Mongol infants. Brill and Forgotson (1964) draw attention to the greater frequency of Down's Syndrome among siblings of leukaemic children. The present communication relates to an infant with Down's Syndrome who succumbed to acute congenital leukaemia in the neonatal period.

Case Report

A 3-day-old baby was brought for medical attention with history of convulsions. The infant, who weighed 6 lbs. 3 oz., was the first born of a 37 year old woman and had been delivered at term. The antenatal period had been uneventful and history of maternal irradiation could not be elicited. The mother noticed that the infant had become 'Jittery' since the second postnatal day. An episode of frank convulsions involving all the four extremities had occurred 57 hours after birth. Features of cerebral irritation, including the characteristic cerebral cry were evident on physical examination. It was also observed that the infant was a Mongol. Various stigmata of Down's Syndrome like microcephaly, low set ears, flat facies with hyperteleorism, arched gothic palate, cutis laxa, small hands and feet, marked hypotonia, diastasis recti, incurving of the little fingers, simian crease in the left palm and single crease in the right fifth finger were present. The pouting facial expression on crying and intermittent protrusion of the tongue were also diagnostic. An arresting feature was the presence of haemorrhagic spots over the extremities. A few discrete spots were also visualized on the torso. The infant was given Vitamin K and placed on barbiturates. Haematological investigations were suggestive of congenital leukaemia. R.B.C. — 5.8 million/c.m..; haemoglobin — 18.3 gm%; W.B.C. — 1,87000/c.mm. with 62 per cent blast forms; reticulocyte count — 6%; blood group — 'A' (mother's blood group: 'A'). Rh typing could not be carried out. Skiagram of the skull did not reveal any cranial fracture, and that of the chest was normal. No evidence of infection anywhere in the body could be discerned. The baby had another bout of convulsions 2 hours later and expired soon after.

Comments

It is evident that the infant had congenital leukaemia and succumbed to intracranial haemorrhage stemming from the leukaemic process. The distinct association between older maternal age and Down's Syndrome has been well documented. Stewart *et al.* (1958) point out that Down's

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Syndrome is encountered among children dying of leukaemia almost 20 times more often than among neonates. Smith (1964) states that although the effect of advanced maternal age has generally been attributed to meiotic nondisjunction, the fault could occur in a postzygotic mitotic division. Cytogenetic studies have established trisomy of chromosome number 21 as the basis of Down's Syndrome. The relationship between chromosome number 21 and leukopoiesis remains to be elucidated. In the case of the Mongol infant with congenital leukaemia described by Honda *et al.* (1964), a small percentage of leukocytes exhibited one, and occasionally, two deleted small acrocentric chromosomes. Ross *et al.* (1963) opine that the regulatory mechanism for the production of or maturation of leukocytes is defective in such infants. Infection as also blood group incompatibilities are known to result in leukaemoid reactions. Honde *et al.* point out that of the reported cases of Mongol infants who succumbed to congenital leukaemia, four had blood group incompatibilities and two had co-existent septicaemia. That folic acid deficiency could also precipitate a similiar reaction was convincingly demonstrated by Lahey *et al.* (1963). Extremely high leukocyte counts have been described in congenital leukaemia. In the case reported by Pein and Garvie (1950), the count was as high as 1,600,000/c.mm. Leukaemia has not been demonstrated in the mother of any infant with congenital leukaemia.

Summary

The case of a newborn infant with Down's Syndrome and congenital leukaemia is reported, reference being made to relevant literature.

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