Case Report

Haemorrhagic Disease of Newborn presenting as Subdural Hematoma

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Introduction

Haemorrhagic disease of the newborn (HDN) can be defined as vitamin K deficiency related bleeding. HDN is one of the most frequent bleeding disorders in infancy [1]. It is classified as early, classical and late-onset disease according to the time bleeding occurs. Early HDN is diagnosed when the bleeding begins in the first 24 hours of postnatal period. It is frequently seen in babies whose mothers are on antitubercular (isoniazid and/or rifampicin), or antiepileptic (such as phenytoin and phenobarbitol) drugs. Classical HDN occurs between 2-5 days of neonatal period and life-threatening bleeding is rare. Late-onset disease can be seen during infancy but predominantly at 4-8 weeks of life. Its incidence is 4-25/1,00,000 births in the western countries but as high as 25-80/1,00,000 in eastern countries [2]. Late haemorrhagic disease is diagnosed if bleeding occurring after the 7th day of life with normal platelet count, prolonged prothrombin time (PT) and partial thromboplastin time (PTT) associated with stopping of bleeding and PT/PTT returning to normal after giving vitamin K [3].

Case Report

A six weeks male infant presented in the out patient department with acute onset recurrent vomiting, poor feeding and activity. He had a large subcutaneous lump over right thigh where DPT vaccine was given three days back. The child was the product of a full-term, normal pregnancy in a 25 year old gravida 2 mother with an uncomplicated antenatal period. Family history was negative for any form of hereditary or acquired bleeding disorder. He was delivered by spontaneous vaginal delivery at home (did not receive vitamin K at birth) and was on exclusive breast feeds. There was no history of antibiotic usage, protracted diarrhoea or head injury in the baby. Physical examination revealed a large haematoma over thigh (right) at the site of injection (DPT vaccine). He was comatose, with bulging anterior fontanelle, unequal pupils, bradycardia and weak respiratory effort. Severe pallor was noted. Prothrombin time (PT) and partial thromboplastin time (PTT) done at that time were markedly elevated. Platelet count was normal. Computed tomography (CT) scan of the head showed a right sided temporo-parietal subdural haematoma with mass effect (Fig.1). He was treated with intravenous vitamin K, after which PT and PTT were normal. He also received fresh blood (fresh frozen plasma could not be given due to non availability). Immediate neurosurgical intervention was done to drain the haematoma. Repeat CT scan at 48 hours showed evidence of ischemic infarct over the right cerebral area at the region of temporal cerebral hemisphere most likely as a complication of the subdural haematoma. He was discharged after one week without any clinical evidence of neuropsychological sequele. Monthly follow up was continued later, to monitor the coagulation status (PT and PTT) and detect other causes of vitamin K deficiency (e.g. liver disease, malabsorption). He was also advised to take phenobarbitone for seizure precautions. A repeat CT scan of brain done after three months was normal. His neurological examination was normal.

Discussion

HDN is a rare disease with high mortality and morbidity [4]. It is one of the most frequent causes of intracranial haemorrhage in the first year of life. Newborns have only 20-50% of adult coagulation activity. Lack of vitamin K administration at birth, exclusive breast feeding, chronic diarrhea and prolonged use of antibiotics make them more prone to vitamin K deficiency bleeding [1]. Almost 2/3rd of the babies with late HDN present with serious intracranial bleeds leading to high morbidity and subsequent mortality. Bleeding occurs because of insufficient vitamin K dependent coagulation factors such as factors II, VII, IX and X activity. Generally, the presentation occurs after the first month of life. HDN is the major probability in a bleeding infant if PT-PTT is higher and fibrinogen level and platelet count is normal. If the bleeding stops and PT
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returns to normal after vitamin K supplementation, diagnosis is more accurate. Protein induced in vitamin K absence (PIVKA) levels is not considered essential in the case definition (PIVKA-II levels are elevated in vitamin K deficiency).

Late HDN may be seen at any time after eight days and before twelve months, but is frequent between four to eight weeks. It generally presents with intracranial haemorrhage, injection haematoma and widespread deep ecchymosis. In addition, gastrointestinal system and superficial skin haemorrhage may be seen [2,5]. Breast-fed infants and newborns with inadequate vitamin K prophylaxis are under the risk of haemorrhagic disease. The amount of vitamin K in mother’s milk is not sufficient. HDN is more frequent in babies who are born at home [6]. Vit K deficiency can also occur due to secondary causes. Chronic diarrhea, cystic fibrosis, biliary atresia, celiac disease, alpha 1-antitrypsin deficiency, abetalipoproteinemia and a history of warfarin usage for a long period may induce vitamin K deficiency [6]. Risk of intracranial haemorrhage in late HDN is reported in 50-80% cases [7]. While subdural is the most common location for haemorrhage, subarachnoid haemorrhage is the second most common type. Zengin et al [8] have reported the rate of subdural, subarachnoid and intraparenchymal bleeding as 100%, 80% and 30%, respectively [8]. In another report, Pooni et al [2] reported subdural haemorrhage in 57.2% and subarachnoidal haemorrhage in 46.4% of the patients in their study. Late HDN can present with convulsions, poor sucking, irritability and pallor. Haemorrhages of gastrointestinal system, mucosal membranes and skin can accompany the disease. Intracranial haemorrhage is the major cause of morbidity and mortality. Mortality is reported in 14-50% cases by various authors [5].

Administering vitamin K to every newborn at birth can impede the disease, which has a high morbidity and mortality [5,6]. Oral prophylaxis of vitamin K is preventive against early and classical haemorrhagic disease, but parenteral administration of vitamin K is required for the late disease [7,9]. Current recommendations for vitamin K prophylaxis are to give vitamin K to all newborns as a single intramuscular dose of 0.5 to 1 mg (AAP policy statement 2006). Findings of Cochrane review (2009) are as follows – “A single dose (1.0 mg) of intramuscular vitamin K after birth is effective in the prevention of classic HDN. Either intramuscular or oral (1.0 mg) vitamin K prophylaxis improves biochemical indices of coagulation status at one to seven days. Neither intramuscular nor oral vitamin K has been tested in randomized trials with respect to effect on late HDN[10].

The low concentration of vitamin K in human breast milk and the predisposition to vitamin K deficiency bleeding following exclusive breast feeding is emerging as a matter of concern especially in developing countries where exclusive breast feeding is vigorously advocated to promote optimal health in the infant. Most reports of late HDN have been in babies born at home and not given vitamin K prophylaxis [5].

Conflicts of Interest
None identified

References

10. The Cochrane Database of Systematic Reviews 2009 Issue 2. Published by John Wiley and Sons, Ltd.