Case Report

Nodular Purpura and Intracranial Bleeding due to Late Onset Hemorrhagic Disease of Infancy: A case report

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Abstract

UHQ, a four -month-old female child, was rushed to the Emergency Medicine Department (EMD) due to severe spontaneous bleeding. This exclusively breast fed child had a history of diarrhea, vomiting and taking broad-spectrum antibiotics over the last two weeks. She developed nodular purpuric spots for about three days prior to the intracranial bleeding. She was unconscious and responding only to pain, and was severely pale. She was transfused with two units of whole blood and underwent a craniotomy for draining the hemorrhage. She was treated with Vitamin K supplements. Her OFC was 44cm on admission. She was discharged after one week, with OFC 41cm. On follow-up, she is developing normally.

Introduction

Late onset hemorrhagic disease of the newborn or Vitamin K Deficiency Bleeding (VKDB) is a recognized entity usually occurring between 2-12 weeks, most commonly in exclusively breast-fed infants who did not receive vitamin K prophylaxis at birth. Vitamin K content is low in human milk and more than half of these infants present with acute intracranial hemorrhages [1]. Several cases reports have been described in young exclusively breast fed infants with gastroenteritis, whereby the intricate balance of Vitamin K is deranged, leading to intracranial bleeding [2,3,4]. Newborn infants are at risk of developing vitamin K deficiency due to various reasons. Transplacental transfer of vitamin K in pregnancy and storage in neonatal liver is also limited. Secondly the bacteria that produce the active form of vitamin K (K**2**) that's readily absorbed by mammals are absent in the intestines of newborns. [5] Breast milk is a poor source of vitamin K (1-4 ug/) while recommended dietary intake of vitamin K is 1 ug/kg/d [6]. Formula milk can

contain up to 100 ug/L and therefore babies who are formula fed receive nearly 100 times more vitamin K than those breastfed. There are virtually no reports of VKDB occurring in infants who are
formula fed [7]. Human breast milk is a unique fluid and supplies all dietary essentials needed for first six months of life except vitamin K. Curiously, vitamins are lower than their formula counterparts but apart from vitamin K, deficiencies are unusual due to their high bioavailability [7].

Vitamin K is an essential cofactor in the production of coagulation factors that help the blood to clot; hence deficiency can lead to spontaneous bleeding from various sites such as gastrointestinal, epistaxis, intracranial bleeding and others.

The postnatal administration of vitamin K has dramatically reduced the incidence of VKDB during the first weeks of life, although sporadic cases with late-onset hemorrhage are described among exclusively breastfed infants who did not receive additional prophylaxis [7] .

Case report

A four-month-old female baby UHQ was seen at an outpatient clinic with a history of severe gastroenteritis and electrolyte imbalance for two weeks. She was admitted for IV replacement fluids and was also treated with zinc supplementation, Oral Rehydration Salt (ORS) solution and azithromycin for three days. She was exclusively breast fed. On the 13th day of the gastroenteritis, she developed spontaneous nodular purpura, first on the left thigh and then several on the right groin and on the chest. She became lethargic, pale and was unable to breast feed. She was rushed to the EMD at this hospital after a CT scan from another hospital showed intracranial bleeding. She was found to be severely pale, unconscious, and responding only to pain. Her vitals were stable, except for tachycardia, and was saturating at 90-95 percent with face-mask oxygen. Her Occipital Frontal Circumference (OFC) on admission was 44 cm. The CT Scan showed left sided fronto-parieto-occipital sub-acute subdural hemorrhage with midline shift and sub-falcine herniation. Her initial haemoglobin was 3.2gms/dl and platelets were normal (345K/cmm). The PT was 22s [12-22 seconds] and APTT was 42 [30-40 seconds], with an INR of 3 on admission. She was transfused fresh blood, given an injection of Vitamin K 3mg stat and underwent a craniotomy to release the intracranial hematoma. Approximately 100mls of blood was removed, and the drain was kept for 24 hours. The PT and APTT reverted to 12s and 32s respectively, with an INR of 1.21 on the second day post-operatively. Fibrinogen levels were not tested. There was no further bleeding. The child improved significantly, although there were associated residual seizures which were managed with phenobarbitone and carbamazepine. The OFC at discharge was 41cm. She continues to be followed up in the OPD clinic.

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Figure 1: Nodular purpura on left lower thigh, chest and right upper thigh

DISCUSSION

The incidence of intracranial hemorrhage in the late form of Vitamin K deficiency bleeding (VKDB) is not known in our environment. There are several factors which indicate that the most likely cause of the bleeding in this child is Vitamin K induced haemorragic disease. This includes the history of gastroenteritis in an exclusively breastfed infant, recent use of antibiotics, and deranged coagulation tests prior to vitamin K administration. The occurrence of nodular purpura in a young infant with risk factors such as gastroenteritis, broad spectrum antibiotic use and exclusively breast-fed infant who is not supplemented with Vitamin K should raise the high possibility of Vitamin K related bleeding.

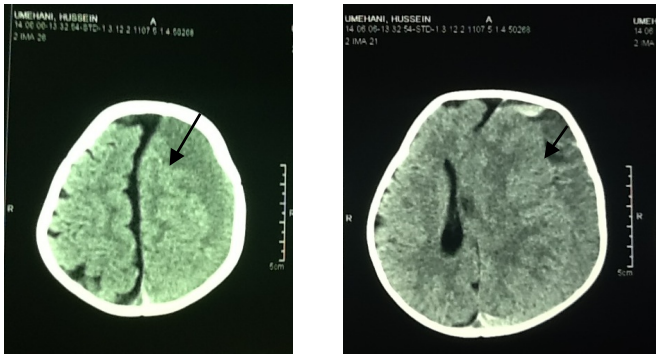

Figure 2: CT-Scan Imaging (without contrast) of the brain showing extensive intracranial hemorrhage, with shift of the midline.

Nodular purpura is uncommon and its presence heralds potentially serious Vitamin K deficiency bleeding in young infants. In a case report from India, an 8-month-old exclusively breastfed male infant presented with multiple purpuric, nodular non-collapsible swellings on the trunk for four days, and later presented with intracranial hemorrhage. Investigations showed raised PT and PTT . Although skin and mucosal bleeding may occur in 1/3rd of infants with VKDB, 'nodular purpura' is not a common presenting feature. Earlier recognition of VKDB and immediate investigation and treatment helps prevent the potentially fatal outcome of the disease [8].

Although the Standard Treatment Guidelines (STG) of Tanzania from Ministry of Health (MoH) recommend 1mg of vitamin K intramuscularly at birth to all newborns as a preventive measure, [9] most hospitals do not adhere to the routine practice. In a study done at Muhimbili National Hospital (MNH), vitamin K prophylaxis is only given to neonates/infants with "high risk" of bleeding [10]. There is a need to change practice and implement guideline surrounding vitamin K administration to prevent against, Vitamin K deficiency bleeding in infants [3].

This case report calls attention to the following issues:

1. The supplementation of Vitamin K to all newborn infants must be implemented in Tanzania
2. Recognition that late hemorrhagic disease of infancy exists and may present with nodular purpura, which heralds severe intracranial bleeding. [2, 4, 6]
3. Use of broad-spectrum antibiotics in treatment of diarrhoea in infants is not warranted.
4. Infants with gastroenteritis, who are exclusively breast fed, should have a dose of Vitamin K injection if they haven't started complimentary feeds.

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