Case Report

Disseminated Intravascular Coagulation with Intracranial Haematoma in Neonatal Congenital Syphilis

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Summary

Disseminated intravascular coagulation (DIC) although a well known complication in neonatal sepsis is extremely rare in congenital syphilis and there are scanty reports of this entity in the literature. Intracranial bleeding following DIC in neonatal congenital syphilis is even rarer, and has been reported only once earlier. We are reporting the second case of neonatal DIC with intracranial haematoma due to congenital syphilis in a newborn. Our patient also had clinical and biochemical evidence of hepatitis which predisposes to DIC. Extensive investigations and emergent use of imaging modalities including ultrasound and CT scan led to early diagnosis and treatment in our patient, who could therefore be salvaged from an otherwise life threatening disease.

Key words: DIC, neonatal sepsis, congenital syphilis, intracranial haematoma, hepatitis, ultrasound, CT scan.

Introduction

Syphilitic infection in the foetus can be severe enough to cause stillbirth [1]. In live borns, hepatosplenomegaly, condylomata lata, rhinitis, meningitis and lymphadenopathy occur [1]. Nevertheless, most infants with congenital syphilis are asymptomatic at birth. Infants who develop clinical symptomatology during the first 2 years of life are considered to have early congenital syphilis, whereas features that appear later, usually near puberty comprise late congenital syphilis [1]. Disseminated intravascular coagulation (DIC) although a well known complication in neonatal sepsis is extremely rare in congenital syphilis and we found scanty reports of this entity in the literature [2–4]. Furthermore, intracranial bleeding following DIC in neonatal congenital syphilis has been reported only once earlier [2]. To the best of our knowledge, we are reporting the second case of neonatal congenital syphilis complicated by intracranial haematoma due to DIC in a newborn. Since in many reports of DIC in congenital syphilis the infants died, our report is unique, as our patient recovered on specific antibiotic and supportive therapy including fresh frozen plasma and platelet concentrate.

Case Report

A term baby boy weighing 2.1 kg was born to a third gravida mother with no previous live issues. The earlier two pregnancies had ended in still births. This pregnancy was unbooked but immunized; the mother had received two doses of tetanus toxoid. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy. Her VDRL was strongly positive (1:32 dilution) and she had received inadequate penicillin therapy.
treatment (warmth, intravenous fluids, oxygen) and was given injection penicillin in appropriate doses. CSF VDRL was reactive in 1:8 dilutions.

One Day 3 of life the baby developed conjugated hyperbilirubinemia; however, the cry and activity were normal. At this time the liver was enlarged 3 cm below costal margin and the spleen was just palpable. Abdominal sonography did not reveal any additional abnormality other than hepatomegaly. The liver function tests revealed, total S. bilirubin 18 mg% (direct 10 mg% and indirect 8 mg%), SGOT 121 IU; SGPT 298 IU and serum alkaline phosphatase 12 KA. Sepsis screening did not show evidence of infection and blood culture was sterile. Coomb’s test was negative. Considering this rise in S. bilirubin and raised transaminases as part of congenital syphilitic hepatitis, the baby was continued on supportive therapy, along with injection penicillin in appropriate doses. Radiographs of knees and wrists obtained to evaluate for osseous manifestations of congenital syphilis were normal.

On Day 6 the child developed ecchymosis below right eyelid, peri-oral cyanosis, with haemorrhagic gastric aspirate and tonic–clonic seizures. Seizures were controlled with luminol (phenobarbitone) therapy along with intravenous dextrose and calcium gluconate administration. Neuroimaging was undertaken immediately: cranial ultrasound revealed an echogenic space occupying lesion suggestive of haematoma in the right parietal lobe and the Non-contrast CT (NCCT) scan of the brain (CTDI—10 mGy) with adequate body shielding, confirmed the presence of large haematoma occupying the right parietal lobe (Fig. 1), causing significant oedema, mass effect, compression of ipsilateral (lateral) ventricle and midline shift (Fig. 2). The haematological profile at this stage revealed, Hb 15.4 g%, total lymphocyte count (TLC) 7400 mm$^3$ with polymorphs 50%, lymphocytes 48% and monocytes 2%. The platelet count was 90 000 mm$^3$, Prothrombin time (PT) was 62 s vs. control 13 s, Partial thromboplastin time with Kaolin (PTTK) was 100 s vs. 34 s, thromboplastin time (TT) 28 s vs. control 18 s, with increased fibrin degradation products (FDP). Prolonged PT, PTTK and TT with increased FDP

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FIG. 1. Non-contrast CT brain at the level of lateral ventricles shows a 2.8-cm size haematoma in the right parietal lobe with surrounding oedema and compression of ipsilateral lateral ventricle.

FIG. 2. CT scan at a more caudal level (quadrigeminal cistern), shows the extent of oedema in the right parietal lobe with compression of ipsilateral lateral ventricle and contra lateral displacement of interhemispheric fissure.
and decreased platelet count lead to the diagnosis of DIC. At this time the baby was stabilized with supportive treatment including fresh frozen plasma, platelet concentrate, vasopressor agents and anticonvulsants (phenobarbitone).

On Days 9 and 10, the baby continued to have similar episodes of bleeding. Blood component therapy comprising fresh frozen plasma and platelet concentrates were continued along with other supportive therapy. On Day 11, the baby’s condition showed signs of clinical improvement. At this time the platelet count was 100,000 mm$^{-3}$. PT was 20 s vs. control 13 s, PTTK was 59 s vs. control 34 s, TT was 22 s vs. control of 18 s and FDP was still positive.

On 15th day extreme pallor was observed. At this time the Hb was 9 g% and platelet count was 1,30,000 mm$^{-3}$. The coagulation profile PT 16 s vs. control 13 s, PTTK was 40 s vs. control 34 s and TT was 18 s vs. control 18 s. The FDP test was negative. Packed cell transfusion was given to boost the haemoglobin level to >10 g%. There was gradual clinical improvement after the 16th day of life and by the 20th day, the baby was accepting oral feeds. The infant was discharged from hospital on 25th day on anticonvulsants and breastfeeds, with advice to report back for regular follow up.

**Discussion**

Disorders frequently predisposing to neonatal DIC include foetal/neonatal conditions such as asphyxia, dehydration, sepsis and intravascular catheters, moreover, maternal diseases such as diabetes and cardiac diseases also contribute [5]. However, DIC is a rare manifestation of congenital syphilis and few reports of this entity are found in the literature [2]. Among these reports, we found intracranial bleeding as a manifestation of DIC in congenital syphilis, only in one case out of the series of 46 infants (of early congenital syphilis), reported by Freiman and Super in 1966 [2]. Mondanloiu and Linzy [5] reported fatal outcome in a neonate with congenital syphilis who had bleeding from a venepuncture site. In the other reports of DIC due to congenital syphilis, hepato-splenomegaly, liver failure, anaemia, purpura and sepsis were the leading manifestations [3, 4]. One of the recent reports of neonatal DIC due to congenital syphilis has been published by investigators from Hungary [3].

In our patient, we observed a bleeding tendency and deranged coagulation profile and raised FDP on the sixth day of life and an intracranial haematoma was also documented. The pathophysiology of DIC is triggered by endothelial damage and production of tissue factors. Both these pathologies then activate intrinsic and extrinsic pathways for disseminated intravascular coagulation. This process of DIC further causes consumption of platelets and clotting factors including fibrinogen, which in turn leads to a bleeding tendency. Simultaneous activation of plasmin increases formation of FDP, which is an indicator of the occurrence of DIC (the pathophysiology is summarized in Fig. 3) [6, 7]. In our neonate with DIC, we presume that vasculitis which is a known pathology of congenital syphilis, was the triggering mechanism which led to consumptive coagulopathy [1]. Secondary syphilitic hepatitis (which was also present in our patient), is also known to contribute to DIC [1].

Haematological abnormalities known to occur in congenital syphilis usually are anaemia and thrombocytopenia [1, 2, 8]. The mechanism for anaemia and thrombocytopenia is believed to be bone marrow infection and/or haemophagocytosis [8]. However, DIC is an unusual haematological manifestation of congenital syphilis which probably occurs due to vasculitis and/or liver failure [1]. The known laboratory indicators of DIC are thrombocytopenia, prolonged PT, prolonged PTTK, decreased fibrinogen levels, elevated FDP and presence of schistocytes or fragmented red blood cells in peripheral smear [6, 7]. In our patient thrombocytopenia, prolonged PT, prolonged PTTK and increased FDP were present as evidence of DIC. We believe that thrombocytopenia, syphilitic vasculitis and syphilitic hepatitis, all three of which are known pathologies of syphilis which occurred in our neonate, were responsible for precipitating this complication.

The treatment of DIC involves treatment of the primary cause and control of haemorrhage and...
anaemia. In our patient who was already on penicillin therapy, regular infusions of fresh frozen plasma and platelet concentrates prevented fatal outcome from an otherwise life threatening condition.

Our experience highlights not only a rare complication of DIC with intracranial haematoma in neonatal congenital syphilis, but also emphasizes the invaluable role of emergency cranial ultrasound and CT scanning in confirming the diagnosis.

References