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Severe atypical harlequin phenomenon in a near term infant to Covid-19 positive mother: A case report

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Abstract

We report an unusual, sever and transient skin color changes in a 36 weeks baby in his first day of life. The changes included sever blanching of the right toes and the penis, concomitant with sever mottling of the back and anterior abdominal wall mainly on the right side but were not well demarcated. The baby's clinical condition, transient course of illness and investigations ruled out sepsis or any evidence of thrombosis. Mum turned to have Covid 19 but no evidence it affected the baby, who was discharged on day 6 and is currently 8 months old with no recurrence of such event.

Keywords

neonatal thrombophilia; neonatal Covid 19; harlequin phenomenon; neonatal Raynaud's.

Case Report

A 36 weeks baby boy, admitted to NICU with respiratory distress. Mum had gestation DM and was controlled. He was born by elective section for maternal fever following PROM for 2 days. During delivery, there was a visualized placental separation and bleeding but blood transfusion to the mum was not required. No previous history or family history of thrombosis or abortions. Resuscitation was smooth and apart from tachypnea and grunting, initial systemic examination was normal including peripheral pulses. He was started on first line antibiotics for suspected sepsis and was stable on CPAP on 25% oxygen.

Following the usual baby care around 20 hours of life, he had an episode of melena which was concomitant with sever, diffuse and unusual changes of skin color of the lower back, right lower abdomen, right hemi-scrotum, penis, and toes of the right foot (Figures 1,2). The changes included sever blanching of the right toes and the penis, mimicking sever arterial flow obstruction and sever mottling of the skin of the

abdomen and back (Figures 3,4,5). There were no hemodynamic changes at that point or later, throughout his stay.

The color changes were extreme and did not follow a certain vascular anatomy or previous similar reports in literature. We needed to exclude sever sepsis and thrombophilia (particularly Protein C/S deficiency – Purpura fulminans). Immediate doppler scans did not show any arterial flow obstruction (from Comon femoral artery and to dorsalis pedis arteries on both limbs). Venous system was patent. Septic screen was normal. Initial thrombophilia screen septic screen, clotting profile and D-dimer were withdrawn and later appeared to be normal. It was noted that in the first 2 days, blood withdrawal was extremely difficult.

Within 30 minutes, color changes start to fade on positioning and hot fomentation. Whitish discoloration of the penis, skin of the back and abdomen completely resolved in 2 hours. The right foot went through a hyperemic phase suggesting reperfusion. The right hand and the sole of right foot remained mildly dusky for 3-4 days with very mild elevation of d-Dimer on follow up (Table 1). On immediate hematology consultation, the baby was started on prophylactic dose of LMWH and was given once FFP transfusion before all lab and duplex scans appeared but after clinical improvement started to happen. The next day, we were notified that maternal Covid-19 nasopharyngeal swab was positive (she had persistent fever). The baby's swab was negative.

While in NICU, the baby continued to be well clinically, CPAP was removed the same night. Melena recurred but there was no fresh bleeding or any fall in hemoglobin. It was attributed to swallowing maternal blood and not related to the event (Apt test was not available). Feeding was started and full feeds were reached by day 4 There was no family history of Raynaud's and maternal ANA and lupus anticoagulant were normal. The baby was continued on prophylactic dose of LMWH for 2 weeks, till all lab and thrombophilia screen were back and were considered normal for age and after the final senior hematology consultation concluded that it has no role in management. He was discharged on day 6 of life and till his current age (8 months), no similar episodes were reported.

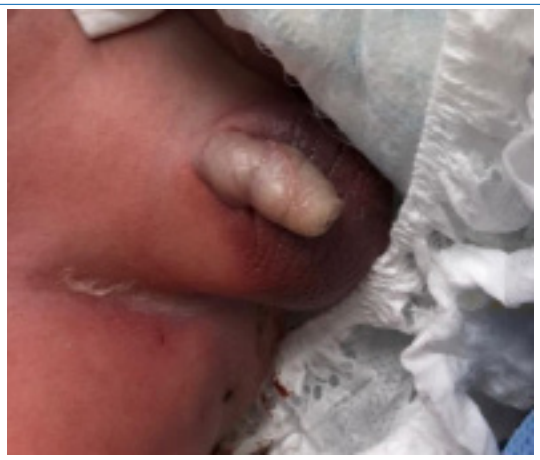


Figure 1: Extreme whitish appearance of the penis during the initial phase simulating the effect of sever arterial spasm.

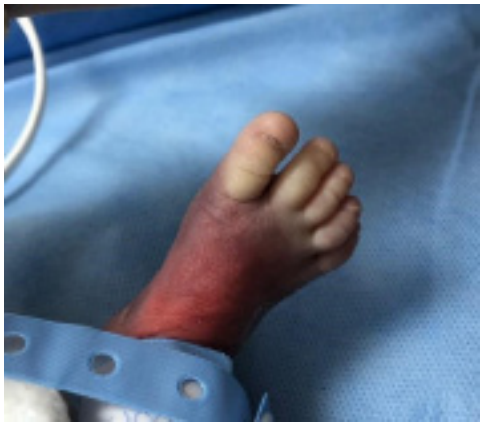
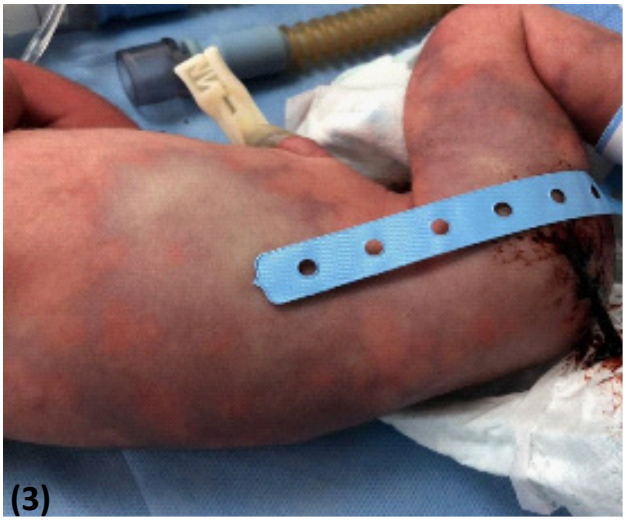
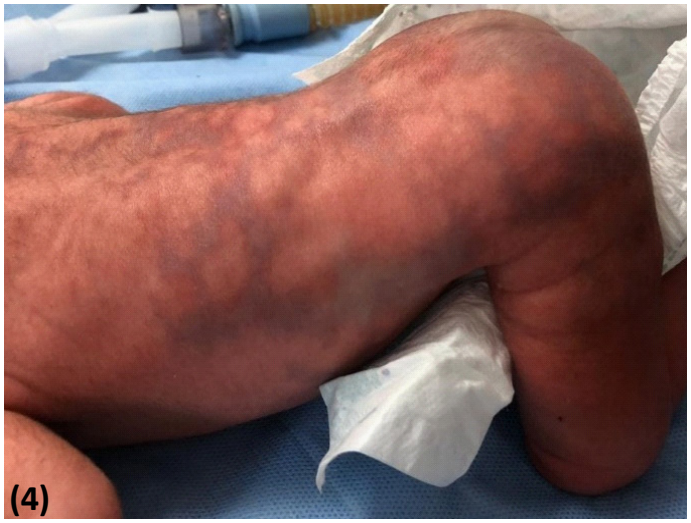


Figure 2: Whitish appearance of the toes of the right foot during the initial phase simulating the effect of sever arterial spasm.





Figures 3 & 4: Blotchy bluish and whitish discoloration of the skin of the back, abdomen, and right thigh concomitant with episode of melena.

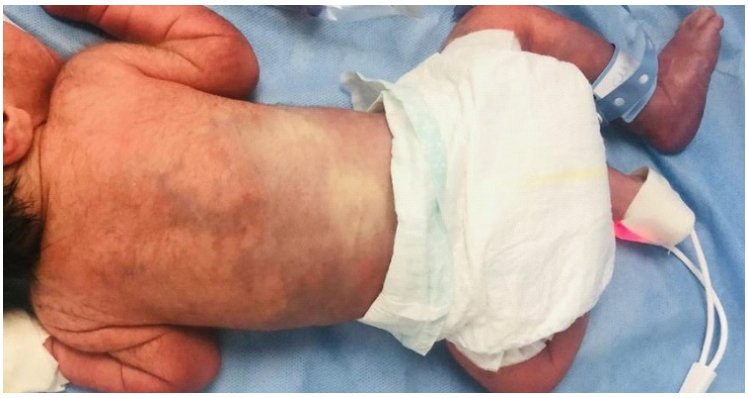


Figure 5: Whitish blanching of the back coupled with sever mottling.

Table 1: Is showing only the relevant initial lab and 5 days follow up. Septic screen was negative, and all biochemistry lab results were within normal range.

	16/10/2020	21/10/2020
WBC	7.4	8.4
Lymph	3.9	3.6
HBG	16.2	15.2
PLT	239	340
PTT	29.8	43
INR	1.15	1
D Dimer	0.58 ug/ml FEU (0.06-0.7)	0.8 ug/ml FEU (0.06-0.7)
Protein C activity	35% (N range :70- 130)	
Protein S level	37% (N range :70- 130)	
Maternal Lupus anticoagulant	Negative	
Covid 19 PCR	Negative	

Discussion

Our case was unique for the unusual presentation. Melena and discoloration -later proven to be unrelated- occurred at the same time which made pointing the diagnosis extremely challenging in a level 2 NICU. The patchy distribution of the affected areas was rather bizarre and didn't follow a specific arterial pathway.

The short clinical course suggests that the most likely diagnosis is sever transient neurovascular phenomenon, probably atypical form of harlequin color change (HCC). HCC was first described in 1952 and since then, reports of both typical and atypical forms have been reported in both children and adults [1]. The exact mechanism is unknown but thought to be an autonomic (sympathetic) dysfunction of the immature hypothalamic. Typical HCC occur in 2-5 days for seconds and up to 30 minutes [2]. It occurs more frequently in preterm babies and with certain medications (e.g anesthetic medications, PGE2) but can occur in 10% of healthy FT babies. It's characterized by color change that bisects the baby sharply into two halves, one hyperemic (usually the dependent side) and a pale side. Atypical forms might last longer (2 weeks in one case report) or have more patchy distribution [3-5].

Early on, we aimed to exclude purpura fulminans, a grave condition caused by congenital or acquired deficiency of protein C&S that presents with vascular thrombosis, skin necrosis and gangrene. Usually babies are in poor condition, lesions persist or worsen if no active treatment was provided. Active treatment includes fresh frozen plasma (FFP) transfusion, protein C concentrate and anticoagulants. There is definite radiological evidence of thrombosis in doppler studies and sometimes, abnormal levels of protein c & S (normal protein c and s in neonates are about 1/3 adult values) [6,7]. In a retrograde reflective manner, FFP and LMWH our baby received were not indicated.

While thrombosis is a reported complication of both Covid 19 infection and vaccine as well, there is no evidence that maternal covid might have anything to do with this presentation. A single case report of upper limb ischemia noted at birth in a preterm baby born to a mum with active Covid 19 was published suggesting possible inutero affection. Vascular occlusion was confirmed by scans and limb amputation was performed [8]. Our case didn't present since birth and was transient with no evidence of limb ischemia suggesting a reactive neurovascular event.

Other differentials of our case are neonatal Raynaud's and sever cutis marmorata. Raynaud's disease is an episodic condition caused by sever peripheral arterial vasospasm resulting from cold exposure resulting in peripheral cyanosis (typically bilateral), followed by the known color sequence It is very rare in infants and strong family basis is usually established [9]. Cutis marmorata is a benign condition that occur in all ages more frequent in neonates. It is characterized by diffuse, livedoid mottling of the skin in response to cold due to capillary and venular spasm [10].

Declarations

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References

1. Valerio E, Barlotta A, Lorenzon E, Antonazzo L, Cutrone M. Harlequin color change: neonatal case series and brief literature review. American Journal of Perinatology Reports. 2015; 5: e73-6.

2. Santos SF, de Abreu MR. Harlequin phenomenon at birth: and she was born with a white stocking. Case Reports. 2015; 2015: bcr2015210549.

3. Khemani S, Bahadur Ali S, Karim S, Yezdan, M. Harlequin colour change. Journal of the College of Physicians and Surgeons Pakistan. 2017; 27: S127-S128.

4. Rao J, Campbell ME, Krol A. The harlequin color change and association with prostaglandin E1. Pediatric dermatology. 2004; 21: 573-6.

5. Rao J, Krol A. The harlequin color change. New England Journal of Medicine. 2003 Sep 4; 349: 968.

6. Marlar RA, Neumann A. Neonatal purpura fulminans due to homozygous protein C or protein S deficiencies. InSeminars in thrombosis and hemostasis. 1990.

7. Andrew M, Paes B, Johnston M. Development of the hemostatic system in the neonate and young infant. The American journal of pediatric hematology/oncology. 1990; 12: 95-104.

8. Perveen S, Millington KA, Acharya S, Grag A, Boyar V. Neonate born with ischemic limb to a COVID-19 positive mother: management and review of literature. Case Reports in Perinatal Medicine. 2021; 10.

9-. Kobayashi M, Takano KI, Kamizono J, Ichikawa K. A serious case of primary Raynaud's phenomenon in an infant. Clinical case reports. 2018; 6: 2089.

10. Sharathkumar AA, Castillo-Caro P. Primary Raynaud's phenomenon in an infant: a case report and review of literature. Pediatric Rheumatology. 2011; 9: 1-6.

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