

A Different Harlequin Color Change Case

Farklı Bir Palyaço Renk Değişikliği Olgusu

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ABSTRACT Harlequin colour change is a transient condition that alarms parents usually. The skin area that infant lay on is erythematous, other part is pale and there is a sharp boundary in the middle of the body. Sometimes it does not occur on face and genital area, mucous membranes. Erythema and pallor replace by changing position. It is thought that the cause of it is vasodilatation due to lack of vascular tone depending on immature hypothalamic or sympathetic system. It is also reported that this condition may develop depending on traumas preponderant to sympathetic chain, tumours and vascular pathologies. We have reported a case due to it has differences about clinical presentation between other reported cases.

Keywords: Erythema; harlequin syndrome

ÖZET Palyaço renk değişikliği ebevnleri alarme eden geçici bir durumdur. Üzerine yattığı deri bölgesi eritemlidir; diğer bölüm ise soluktur ve iki vücut yarısı arasında orta hatta keskin bir sınır vardır. Bazen yüz, genital bölge ve muköz membranlar tutulmayabilir. Pozisyon değişikliğine bağlı olarak eritem ile soğuk bölge yer değiştirebilir. Hipotalamik ya da sempatik sinir sisteminin immatüritesine bağlı vasküler tonus azlığından kaynaklandığı düşünülmektedir. Ayrıca bu durumun sempatik sinir zincirine nazif travmalar, tümörler ve vasküler patolojiler sonucu da gelişebileceği bildirilmiştir. Literatürde bildirilen olgulardan farklı kliniğe sahip bir olgu sunduk.

Anahtar Kelimeler: Eritem; harlequin sendromu

Harlequin Colour Change was first reported and named by Neligan and Strang in 1952.¹ It usually starts to emerge in premature infants in the next 2 to 5 days after birth. The skin area that infant lay on is erythematous, other part is pale and there is a sharp boundary in the middle of the body.²⁻⁴ Informed consent form was obtained from the patient's mother and we have reported a case due to it has differences about clinical presentation between other reported cases.

CASE REPORT

12-day-old infant boy who was born on normal spontaneous time and was family's first child. His mother applied with redness complaint that emerged on one-half of her baby as he was 7 days old. There was not a known disease and a similar story in the family. There was diffuse erythema in the body of the newborn, which was unilateral and separated sharply from the

other side (Figure 1). Erythema did not continue in the head and genital area. The case was followed and it was seen that lesions did not repeat after 20th day of baby's life.

DISCUSSION

Harlequin colour change is a transient condition that alarms parents usually.^{5,6} It is stated that this case may occur in newborns with prostaglandin E1 applied congenital cyanotic heart disease. However, it is also stated that this case may have occurred as associated with hypoxia that is another possible launcher of it and may have been linked inadvertently to prostaglandin E1 application.⁷ It is thought that the cause of it is vasodilatation due to lack of vascular tone depending on immature hypothalamic or sympathetic system.^{5,8} It is also reported that this case may develop depending on traumas preponderant to sympathetic chain, tumours and vascular pathologies.⁹⁻¹¹

While it is stated in the literature that episodes take maximum 20 minutes, it has never disappeared in our patient since the emergence of diagnosed erythema, despite the occasional relief.⁵ It disappeared itself after 2 weeks from the moment it appeared. This situation, which resembles harlequin color

change, may be thought to be a different entity by not continuing after the 20th day, not changing the color with position change, and erythema which is constantly separated from the other side. This may be also due to the pathology of the hypothalamus, which is the center of vasomotor mechanisms. However, if this pathology of the hypothalamus continued, half-color change would continue. In addition, the harlequin color changes in the literature indicate that the erythema occurs on the side of laying (back or body front); In our case, there was unilateral sharply limited erythema at the main body.

With a boundary passes right in the middle of vertex - symphysis pubis line, one half of the body erythematous, as a result of hyperemia, seems more dimmed as compared to the other half. As in our case, sometimes it does not occur on face and genital area, mucous membranes. Erythema and pallor replace by changing position. Since hypothalamic centre that controls vasodilatation of the peripheral vessels has not developed sufficiently yet, the bottom half of the body seems red whereas the upper half seems light when the baby is in supine position and the colour distribution would be reverse as the position changes.⁵ While it is stated in the literature that this case is seen erythema in the



FIGURE 1: : Unilateral and sharply separated erythema in the front of the body and, diffuse erythema on the back.

bottom left area by rotating the patient and the upper part is seen pale, lesions that emerged regardless of the position were also not affected by the changes in position in our patient.

Episodes that extended up to maximum 4 hours were reported depending on Horner syndrome or after the interventions of sympathetic chain.¹²⁻¹⁴ It has never disappeared in our patient since the emergence of diagnosed erythema until the 20th day of birth.

If the color change is persistent, a large capillary malformation may be suspected. Probable differential diagnosis are Port-wine stain and nascent hemangioma of infancy, but these can be differentiated due to transient nature of this condition.¹⁵

While it is stated in the literature that this condition occurs usually in premature, newborn infants exposed to hypoxia, our patient was born on time and not exposed to hypoxia.^{3,4}

While it is stated in some sources that it is seen as high as about 10% of newborns, it is mentioned as a “rare” case in some of them.^{5,6}

Since parents are usually on alarm, it is important to recognize this condition of newborn and point out that it is a temporary situation without leaving sequellae.

Conflict of Interest

Authors declared no conflict of interest or financial support.

Authorship Contributions

Idea/Concept: Serkan Demirkıran, Özgür Gündüz; **Design:** Serkan Demirkıran; **Control/Supervision:** Serkan Demirkıran; **Data Collection and/or Processing:** Serkan Demirkıran, Özgür Gündüz; **Analysis and/or Interpretation:** Serkan Demirkıran; **Literature Review:** Serkan Demirkıran; **Writing the Article:** Serkan Demirkıran; **Critical Review:** Özgür Gündüz; **References and Fundings:** Serkan Demirkıran; **Materials:** Serkan Demirkıran.

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