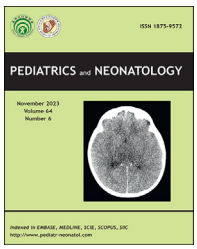
Pediatrics and Neonatology 64 (2023) 690–691

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.pediatr-neonatol.com



Images

Preterm ovarian hyperstimulation syndrome mimicking clitoromegaly

Zeynep Alize Ay [a], Saygin Abali [b,*], Ayse Korkmaz [c], Serdar Beken [c]

[a] *Acibadem Mehmet Ali Aydinlar University, School of Medicine, Turkiye*
[b] *Acibadem Mehmet Ali Aydinlar University, School of Medicine, Department of Pediatrics, Division of Pediatric Endocrinology, Turkiye*
[c] *Acibadem Mehmet Ali Aydinlar University, School of Medicine, Department of Pediatrics, Division of Neonatology, Turkiye*

Received Feb 13, 2023; received in revised form Apr 5, 2023; accepted Apr 20, 2023
Available online 16 June 2023

A premature female infant, born after 23 weeks of gestation through a cesarean section of a 23-year-old mother, was admitted to the neonatal intensive care unit with a birth weight of 600 g. On the postnatal day 103 (corrected gestational week, cGW, 37^{+4}), labial edema and clitoral swelling were observed (Fig. 1A). Blood tests showed high follicle-stimulating hormone (FSH) and luteinizing-hormone (LH) concentrations (19.5 mIU/mL [normal range 1.2–12.5] and 35.4 mIU/mL [<7]), and extremely high estradiol (138 ng/L [<20]) levels. Pelvic sonography showed multiple ovarian follicles in both ovaries, the largest was 11 mm in diameter (Fig. 2), and an enlarged uterus with a size of 34 × 20 × 9 mm. The patient was diagnosed with preterm ovarian hyperstimulation syndrome (POHS). The external genitalia swelling, and gonadotropin and estradiol concentrations started regressing by postnatal day 165 (corrected age 46 days) with no medical intervention (Fig. 1B). No vaginal bleeding was detected during observation. The infant's follow-up laboratory results showed a decrease in FSH, LH, and estradiol concentrations (cGW, 44), 17.7 mIU/mL (1.2–12.5), 4.13 mIU/mL (<7), and 79.6 ng/L (<20), respectively. Due to the gradual clinical resolution, no further follow-up was needed. POHS is a rare self-resolving disorder, presenting with increased levels of gonadotropins in preterm infants, that occurs due to hypothalamic–pituitary–gonad axis immaturity with diminished negative feedback and early disappearance of placental steroids.[1] It manifests in preterm newborns with edema in the vulva, upper leg, hypogastric region, and ovarian cyst/cysts, and with elevated gonadotropin and estradiol levels.[2] The differential diagnoses include clitoromegaly, which is most commonly related to androgen excess, and less well-known nonandrogenic disorders, such as neurofibromatosis, epidermoid cysts, and other tumors, and conditions including Apert and Fraser syndromes. In this case, hyperandrogenism was excluded through laboratory testing of testosterone and dehydroepiandrosterone sulfate (DHEAS) levels. Misdiagnosing the clitoral swelling as clitoromegaly in these patients causes unnecessary laboratory tests for hyperandrogenism.[3] However, virilization can be excluded by the absence of labial fusion, the most important finding of antenatal hyperandrogenism, and the presence of separate vaginal and urethral openings. The clinical

☆ Approval was obtained from Acibadem University and Acibadem Healthcare Institutions.

* Corresponding author. Acibadem Atakent Hospital, Halkali Merkez, Turgut Ozal Bulvari No:16, 34303 13 Kucukcekmece/Istanbul, Turkiye.
E-mail address: saygin.abali@acibadem.edu.tr (S. Abali).

https://doi.org/10.1016/j.pedneo.2023.04.008

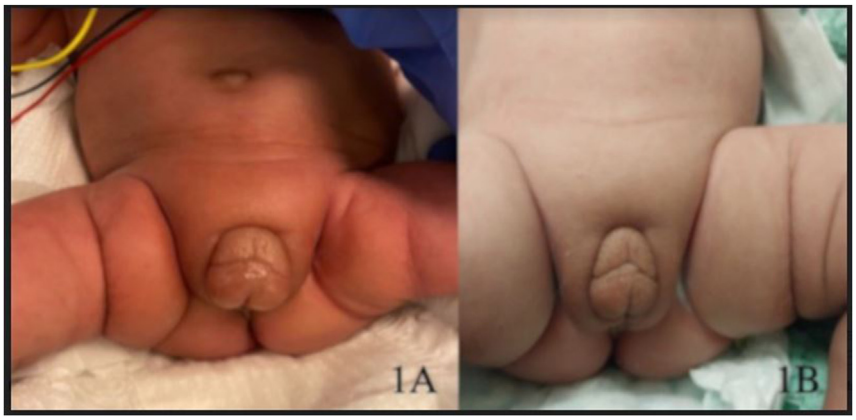


Figure 1 A. The external genitalia of the patient on the postnatal 103rd day showed clitoral swelling and labial edema. B. On the postnatal 165th day showed reduced edema.

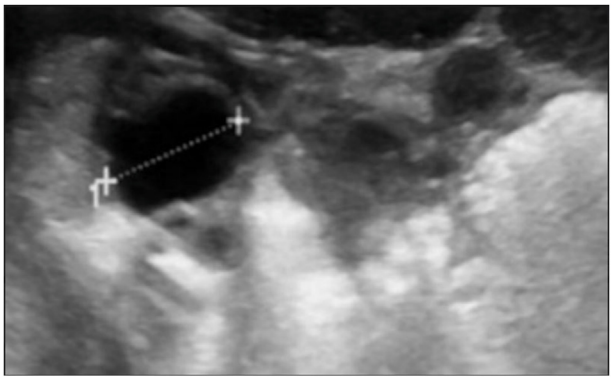


Figure 2 Pelvic ultrasonography showed large ovarian follicles in the right ovary. The largest diameter was 11 mm.

diagnosis of POHS is important to prevent unnecessary laboratory testing in infants with clitoral swelling.

Contributors

ZAA: writing up the report, SA: named consultant, writing up the report, obtaining parenteral consent, SB: named consultant, writing up the report, AK: named consultant, writing up the report.

Funding

The authors haven't received a specific grant for this research from any funding agency in the public, commercial, or not-for-profit sectors.

Patient consent for publication

Parenteral/guardian consent is obtained.

Declaration of competing interest

No Declared.

References

1. Durst MA, Wicklow B, Narvey M. Atypical case of preterm ovarian hyperstimulation syndrome. *BMJ Case Rep* 2017;**2017**:bcr2016217517.
2. Altuntas N, Turkyilmaz C, Yuce O, Kulali F, Hirfanoglu IM, Onal E, et al. Preterm ovarian hyperstimulation syndrome presented with vaginal bleeding: a case report. *J Pediatr Endocrinol Metab* 2014;**27**:355—8.
3. Lee YL, Jamli FM. Preterm ovarian hyperstimulation syndrome presenting as clitoromegaly in a premature female infant. *Arch Dis Child* 2022;**107**:166—7.