Resumen *#825*

Pseudohipoparatiroidismo y embarazo

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Resumen:

El pseudohipoparatiroidismo (PHP) es una entidad rara que requiere modificaciones en el manejo durante el embarazo. La escasa literatura disponible motiva la siguiente presentación.

Paciente de 17 años primigesta fue derivada a las 11 semanas de gestación para control de PHP. El mismo fue diagnosticado a los 12 años a partir de crisis hipocalcémica (calcio total: 4.7 mg/dl; fósforo: 7.8 mg/dl; PTH: 653 pg/ml), tratado con calcio y vitamina D. Durante la pubertad, se diagnosticaron hipotiroidismo primario autoinmune y enfermedad celíaca. La paciente permaneció asintomática durante la gestación, en tratamiento dietario libre de gluten, calcio 2500 mg, calcitriol 1 mg y levotiroxina 112 mcg/día. El examen físico reveló P 57 kg; T 1.46 m(<Pc3). Cuello corto, facie redondeada, contextura robusta, manos pequeñas con acortamiento de falanges distales. Signo de Trousseau y Chvoztek negativos. Laboratorio del primer trimestre: TSH: 5.95 UI/L; T4 Libre: 1.21 ng/dl; PTH: 233 pg/ml; calcio total: 8.7 mg/dl; fósforo: 5.14 mg/dl. Los controles ecográficos fetales fueron adecuados. Durante la evolución de la gesta presentó controles de calcemia en rango normal bajo asociados a escasa adherencia al tratamiento. A las 37.5 semanas nació por cesárea programada (para evitar hipocalcemia materna secundaria a alcalosis metabólica por hiperventilación y riesgo de fracturas fetales asociadas al trabajo de parto) un recién nacido de sexo femenino, peso 3.250 g, 47 cm de longitud, Apgar 8/9. Prequirúrgicamente se administró gluconato de calcio EV por presentar hipocalcemia severa y se mantuvo hasta 24 h posparto. El laboratorio del recién nació reveló: calcio total: 8.9 mg/dl (VR: 9-11), fósforo: 5.9.mg/dl (VR: 4-6), PTH: 115.5 pg/ml (VR: 15-65); siendo el mapeo óseo normal. El puerperio fue normal y se les indicó alta hospitalaria a los tres días. A los 52 días de vida el valor de PTH descendió a 76 pg/ml, asociado a calcio total: 9.8 mg/dl y fósforo: 7.1 mg/dl.

El PHP resulta de una resistencia a la PTH, siendo considerado el embarazo de alto riesgo por posibles complicaciones obstétricas y neonatales. El manejo interdisciplinario de una patología de escasa prevalencia dan sentido al presente reporte.

Palabras Clave:
embarazo; pseudohipoparatiroidismo; manejo interdisciplinario.

Abstract *#825*

Pseudohypoparathyroidism and pregnancy

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Abstract:

Pseudohypoparathyroidism (PHP) is a rare disorder that requires specific modifications in its treatment during pregnancy. The scant literature available about this topic has motivated us to carry out the following presentation.

A 17-year-old patient bearing her first child was referred to our hospital in her 11th week of gestation for PHP control. She was diagnosed with PHP at age 12 after developing a hypocalcemic crisis (total calcium level: 4.7 mg/dL; phosphate: 7.8 mg/dL; PTH: 653 pg/ml), which was treated with calcium and vitamin D. During puberty, she was diagnosed with primary autoimmune hypothyroidism and celiac disease. The patient remained asymptomatic during gestation. She was on a gluten free diet, and she was prescribed calcium (2500 mg), calcitriol (1 mg) and levothyroxine (112 mcg/day). Her general physical examination revealed weight of 57 kg and height of 1.46 m (<Pc3), and her body features included short neck, round facies, robust build and small hands with shortness of distal phalanges. The examination also revealed negative Trousseau's and Chvoztek's signs. Blood test during the first trimester: TSH: 5.95 UI/L; free T4: 1.21 ng/dL; PTH: 233 pg/mL; total calcium: 8.7 mg/dL; phosphate: 5.14 mg/dL. Ultrasonographic parameters for fetal growth were found adequate. During the progress of gestation, calcemia levels were normal to low, which was associated to low compliance to the treatment. At week 37.5, she delivered a baby girl through scheduled caesarean section (to avoid maternal hypocalcemia that could be triggered by metabolic alkalosis through hyperventilation, and to prevent any risk of fetal fracture during labour). The infant weight was 3.250 kg, and her height was 47 cm, with Apgar scores of 8/9. Presurgically, the patient was administered IV calcium gluconate for up to 24 h postpartum after showing signs of severe hypocalcemia. The blood test of the infant showed the following: total calcium: 8.9 mg/ dL (VR: 9-11), phosphate: 5.9 mg/dL (RV: 4-6), PTH: 115.5 pg/mL (RV: 15-65); and the bone scan was normal. The puerperal period was normal, and the patient and her newborn were discharged three days after. At 52 days old, the infant's PTH levels dropped at 76 pg/mL, associated to a total calcium of 9.8 mg/dL and phosphate of 7.1 mg/dL.

PHP is a disorder resulting from a resistance to PTH, in which case the pregnancy should be considered high risk, as possible obstetric and neonatal complications could develop. This report aims to discuss the interdisciplinary management of such rare disorder.

Keywords:
Pseudohypoparathyroidism, PREGNANCY, multidisciplinary team