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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Refractory hypokalemia associated with obstructed hemivagina and ipsilateral renal agenesis

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2026, 43(Συμπλ 1):143–147

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In complex congenital anomalies such as obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome, electrolyte disturbances may have multifactorial etiologies, including renal anatomical abnormalities and hormonal dysfunction. This case report aims to present a case of refractory hypokalemia associated with subclinical hypothyroidism in a patient with OHVIRA syndrome. A 28-year-old woman with a history of OHVIRA syndrome since early life presented with recurrent episodes of muscle weakness and fatigue. She had persistent refractory hypokalemia that did not improve with oral potassium supplementation. Laboratory tests showed persistent hypokalemia (2.9–3.0 mmoL/L), sodium levels ranging from 139 to 150 mmoL/L, elevated thyroid stimulating hormone (TSH) (8.4 μIU/mL) with normal free T4, along with mild leukocytosis and thrombocytosis. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) confirmed unilateral renal agenesis on the same side as the obstructed hemivagina. She was treated with oral levothyroxine with gradual dose adjustments and potassium supplementation. Monthly laboratory follow-up showed improvement in potassium levels and resolution of symptoms in parallel with normalization of TSH. In conclusion, subclinical hypothyroidism may be an important contributing factor to refractory hypokalemia in patients with OHVIRA. A multi-organ evaluation is essential to identify the underlying cause and to design an optimal management strategy.

Ανθεκτική υποκαλιαιμία που σχετίζεται με αποφραγμένο ημι-κόλπο και ομόπλευρη νεφρική αγενεσία

Περίληψη στο τέλος του άρθρου

Key words

Electrolyte imbalance
Hypokalemia
Hypothyroidism subclinical
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Electrolyte disturbances, particularly hypokalemia, are common clinical problems that can affect neuromuscular, cardiovascular, and cellular metabolic functions.[1] Globally, hypokalemia has been reported in 14–20% of hospitalized patients and is associated with increased morbidity and mortality.[2,3] The condition may result from potassium loss through the kidneys or gastrointestinal tract, intracellular potassium shifts, or inadequate dietary intake.[4] Renal potassium loss is frequently observed in patients with congenital or acquired kidney disorders, where increased filtration load and compensatory mechanisms may trigger excessive potassium excretion.[5,6] This mechanism bridges the link between electrolyte imbalances in general and specific conditions such as congenital renal anomalies, which have significant implications for electrolyte homeostasis.[7]

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare congenital anomaly characterized by uterus didelphys, obstructed hemivagina, and renal agenesis on the same side.[8] In addition to affecting reproductive and urinary tract function, this anomaly may increase the risk of electrolyte disturbances through potassium loss secondary to compensatory hyperfiltration in the solitary kidney.[9] Hypothyroidism, particularly in its subclinical form, may also influence potassium balance by reducing Na^+/K^+-ATPase activity and altering cellular metabolism.[10,11] Several reports have suggested a link between hypothyroidism and hypokalemia; however, clear evidence of a link between these variables in patients with OHVIRA has been limited.[12] The lack of evidence raises question about whether an interaction exists between anatomical

and hormonal factors in patients with OHVIRA and refractory hypokalemia.[13] The purpose of this case report is to present a potential association between subclinical hypothyroidism and refractory hypokalemia in a patient with OHVIRA syndrome. We believe that treating the underlying hormonal disorder may help correct the electrolyte disturbance in such cases, offering new possibilities for evidence for clinicians when evaluating and managing hypokalemia in patients with complex congenital anomalies.

CASE PRESENTATION

A 28-year-old woman with a history of OHVIRA syndrome, diagnosed in early life, presented with recurrent episodes of muscle weakness and easy fatigability. Her medical history revealed persistent refractory hypokalemia that was difficult to correct despite standard potassium supplementation, suspected to be related to a combination of unilateral renal anatomical abnormality and endocrine dysfunction. She had no history of hypertension. Physical examination showed short stature with mild clinical features suggestive of hypothyroidism.

Echocardiography revealed an aortic root diameter of 17 mm (smaller than the normal range of 20–39 mm), a left atrial diameter of 24 mm with an elevated left atrial to aortic root (LA/AO) ratio of 1.57 (normal <1.3), and left ventricular dimensions of left ventricular internal diameter in diastole (LVIDd) 30 mm and LVIDs 18 mm (both smaller than normal). The ejection fraction (EF) was preserved at 70%, indicating normal systolic function; however, the mean E/e' ratio was 20.25, suggesting elevated left ventricular filling pressures consistent with diastolic dysfunction. The interventricular septal thickness (IVSd) and left ventricular posterior wall thickness (LVPWd) were both at the upper limit of normal (12 mm), suggesting possible mild hypertrophy. These findings are illustrated in figure 1, with figure 1A showing cardiac dimensions (slightly enlarged left atrium and reduced aortic root diameter) and figure 1B demonstrating preserved systolic function (maintained EF) alongside elevated left ventricular filling pressures.

Arterial blood gas and electrolyte analysis demonstrated persistent hypokalemia (2.9–3.0 mmoL/L), serum sodium ranging from 139 to 150 mmoL/L, a normal blood pH (7.367), slightly elevated PCO_2 (42.1 mmHg), elevated PO_2 (166.1 mmHg), and oxygen saturation of 99.4%. Bicarbonate levels (HCO_3) were within the normal range at 24.4 mmoL/L. Thyroid function testing revealed elevated thyroid-stimulating hormone (TSH) at 8.4 uIU/mL with a normal free T4 level (17.52 pmoL/L), consistent with subclinical hypothyroidism. Hematology results showed mild leukocytosis (11.3×10^3/μL) and thrombocytosis (499×10^3/μL), with hemoglobin and hematocrit levels within normal limits.

Abdominal computed tomography (CT) imaging demonstrated unilateral renal agenesis on the same side as the obstructed hemivagina. Figure 2A shows a coronal section depicting the liver, kidney, spleen, and major vessels (aorta and inferior vena cava). Figure 2B presents a sagittal section illustrating the vertebral column and abdominal organs relative to the posterior wall. Figure 2C displays a coronal section in a different vascular phase, and figure 2D provides an additional coronal view for comprehensive assessment of abdominal organ structures and vascularization. Prior abdominal magnetic resonance imaging (MRI) confirmed unilateral renal agenesis and Müllerian duct anomaly.

Based on the overall evaluation, the most likely cause of refractory hypokalemia in this patient was a combination of subclinical hypothyroidism and congenital renal abnormality, both contributing to increased renal potassium loss. She was treated with oral levothyroxine with gradual dose adjustments alongside potassium supplementation. Figure 3 shows a laboratory timeline demonstrating improvement in serum potassium levels in parallel with TSH normalization, accompanied by a reduction in muscle weakness symptoms.

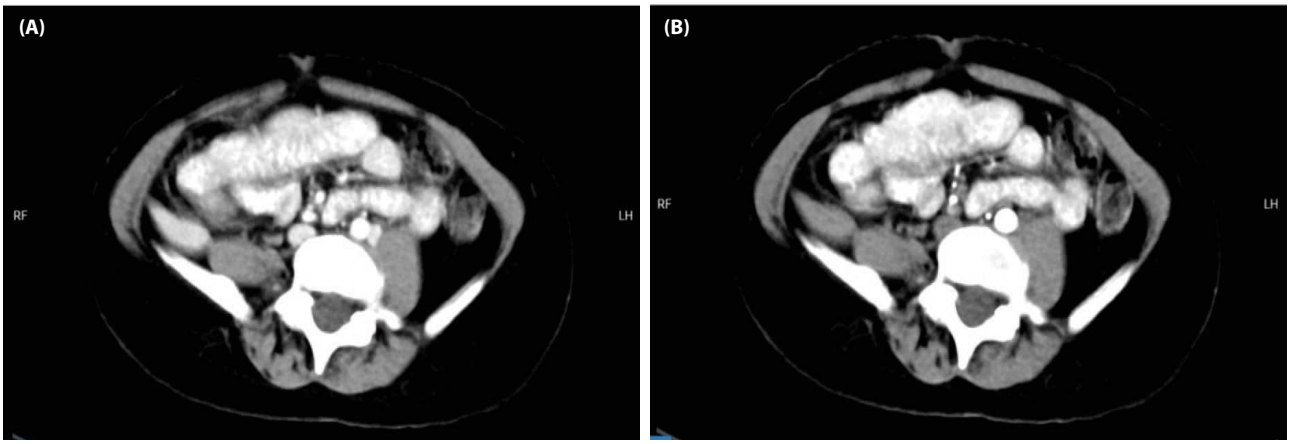

Figure 1. Echocardiographic findings in our patient. **(A)** Cardiac dimensions demonstrating a mildly enlarged left atrium and a smaller-than-normal aortic root diameter. **(B)** Cardiac function showing preserved ejection fraction with evidence of increased left ventricular filling pressure.

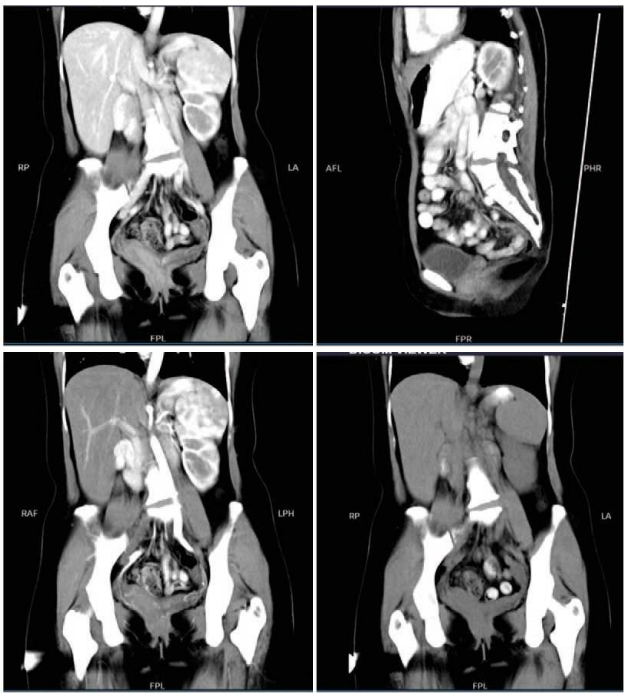

Figure 2. Abdominal computed tomography (CT) scan in coronal and sagittal views. **(A)** Coronal view showing the liver, kidneys, spleen, and major abdominal vessels, including the aorta and inferior vena cava, providing a front-to-back anatomical perspective. **(B)** Sagittal view illustrating the spinal column and abdominal organs, such as the kidneys and bowel loops, in relation to the posterior abdominal wall. **(C)** Coronal view at a different slice level or vascular phase, highlighting abdominal vasculature and its relationship to surrounding structures. **(D)** Additional coronal view offering further assessment of abdominal organs and vasculature across multiple planes for comprehensive evaluation.

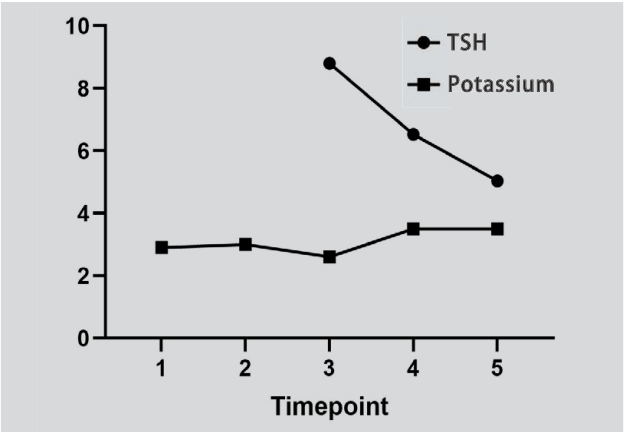

Figure 3. Timeline of TSH and blood potassium levels in patient laboratory follow-up.

COMMENTS

This case describes a 28-year-old woman with a history of OHVIRA syndrome diagnosed in early life. She presented with recurrent episodes of muscle weakness and fatigue, with persistent refractory hypokalemia that remained difficult to correct despite oral potassium supplementation. Echocardiographic evaluation revealed an aortic root diameter of 17 mm (smaller than normal), an elevated LA/AO ratio of 1.57, smaller-than-normal left ventricular dimensions with preserved systolic function (EF 70%), and an average E/e' ratio of 20.25, indicating diastolic dysfunction.[14–16] These findings suggest that chronic electrolyte disturbances may influence structural cardiac adaptation.[17] Arterial blood gas and electrolyte testing showed potassium levels of 2.9–3.0 mmoL/L, sodium levels of 139–150 mmoL/L, normal blood pH, and slightly elevated PCO_2. Thyroid function testing demonstrated elevated TSH (8.4 uIU/mL) with normal free T4, consistent with subclinical hypothyroidism.[18] Abdominal CT confirmed unilateral renal agenesis with ipsilateral hemivaginal obstruction, without evidence of other pathological masses.

The combination of subclinical hypothyroidism and congenital renal anomaly was considered the primary cause of this patient's refractory hypokalemia.[19] Hypothyroidism can impair potassium regulation by reducing Na^+/K^+-ATPase pump activity, while a solitary kidney with compensatory filtration load may promote excessive renal potassium loss.[19] Based on these findings, the patient was treated with oral levothyroxine with gradual dose adjustment and potassium supplementation. The therapeutic approach aimed to correct the underlying hormonal disturbance while restoring electrolyte balance. Monthly laboratory follow-up showed a gradual decrease in TSH levels with concurrent improvement in potassium levels, in line with reduced muscle weakness.

This case demonstrates that multi-organ evaluation is especially relevant in someone with complex congenital malformations like OHVIRA syndrome.[8] The exact cause of refractory hypokalemia is not all due to renal anomalies; concurrent endocrine anomalies may also play a role.[17] The pathophysiology in this case was likely due to the solitary kidney excreting potassium, and then the hypothyroidism negatively affecting potassium metabolism, as well as affect cell membrane function.[20] Consequently, the most appropriate management would counteract the root cause and not just provide potassium replacement. Clinically, this would prevent the patient from developing hypokalemia again and ideally protects the cardiovascular system from potential long-term complications of chronic electrolyte disturbance.[21]

The report offers clinical lessons. First, it emphasizes the importance of a multi-organ assessment approach in

patients with OHVIRA syndrome or other complex congenital defects when there are challenges with uncorrectable electrolyte abnormalities. Second, addressing the root cause (e.g., subclinical hypothyroidism) may ameliorate refractory hypokalemia without solely supplementing a potassium deficiency. Third, including assessments of cardiology, endocrinology, and nephrology in the management of this condition may help identify early complications (for example, diastolic dysfunction from chronic hypokalemia). Fourth, the report highlights that any assessment of hypokalemia must include certain hormonal factors aside from renal considerations. Lastly, the case gives rise to a potential opportunity for further research into relationships between hypothyroidism and electrolyte disturbances in individuals with congenital anatomical anomalies that could inform better screening and treatment protocols.

This study also has limitations. First, as a single case report, the findings cannot be generalized to all OHVIRA patients with refractory hypokalemia. Second, the absence of a control group limits the ability to quantitatively assess treatment efficacy. Third, confounding factors such as nutritional status, daily potassium intake variations, or medication use could not be fully controlled. Fourth, the descriptive nature of this report precludes definitive conclusions about a causal relationship between hypothyroidism and hypokalemia in this patient. Fifth, additional investigations such as detailed renal function testing and genetic analysis were not performed, leaving the possibility of other unidentified mechanisms.

In conclusion, refractory hypokalemia in patients with OHVIRA syndrome may be influenced by hormonal factors, such as subclinical hypothyroidism, in addition to underlying renal anatomical abnormalities. Comprehensive multi-organ evaluation is essential to determine the cause and guide appropriate treatment. Addressing the underlying etiology, such as through levothyroxine therapy, can improve potassium levels and clinical symptoms while preventing recurrence.

ΠΕΡΙΛΗΨΗ

Ανθεκτική υποκαλιαιμία που σχετίζεται με αποφραγμένο ημι-κόλπο και ομόπλευρη νεφρική αγενεσία

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Σε πολύπλοκες συγγενείς ανωμαλίες, όπως το σύνδρομο OHVIRA, οι διαταραχές των ηλεκτρολυτών μπορεί να έχουν πολυπαραγοντικά αίτια, περιλαμβανομένων των ανατομικών ανωμαλιών των νεφρών και της ορμονικής δυσλειτουργίας. Παρουσιάζεται μια περίπτωση ανθεκτικής υποκαλιαιμίας που σχετιζόταν με υποκλινικό υποθυρεοειδισμό σε ασθενή με αποφραγμένο ημι-κόλπο και ομόπλευρη νεφρική αγενεσία (OHVIRA). Επρόκειτο για μια 28χρονη γυναίκα με ιστορικό συνδρόμου OHVIRA από μικρής ηλικίας, που παρουσίαζε επαναλαμβανόμενα επεισόδια μυϊκής αδυναμίας και κόπωσης. Είχε επίμονη ανθεκτική υποκαλιαιμία που δεν βελτιωνόταν με την από του στόματος χορήγηση συμπληρωμάτων καλίου. Οι εργαστηριακές εξετάσεις ανέδειξαν επίμονη υποκαλιαιμία (2,9–3,0 mmoL/L), επίπεδα νατρίου 139–150 mmoL/L, αυξημένη θυρεοειδοτρόπο ορμόνη (TSH) (8,4 μIU/mL) με φυσιολογική ελεύθερη T4, μαζί με ήπια λευκοκυττάρωση και θρομβοκυττάρωση. Η αξονική και η μαγνητική τομογραφία κοιλίας επιβεβαίωσαν μονόπλευρη νεφρική αγενεσία ομόπλευρα με τον αποφραγμένο ημι-κόλπο. Η ασθενής αντιμετωπίστηκε με από του στόματος λεβοθυροξίνη με σταδιακές προσαρμογές της δόσης και συμπλήρωση καλίου. Η μηνιαία παρακολούθηση των εργαστηριακών εξετάσεων έδειξε βελτίωση στα επίπεδα καλίου και υποχώρηση των συμπτωμάτων παράλληλα με την ομαλοποίηση της TSH. Συμπερασματικά, ο υποκλινικός υποθυρεοειδισμός μπορεί να συνιστά σημαντικό παράγοντα στην ανθεκτική υποκαλιαιμία σε ασθενείς με OHVIRA. Απαραίτητη είναι η πολυοργανική αξιολόγηση για τον εντοπισμό της υποκείμενης αιτίας και τον σχεδιασμό μιας βέλτιστης στρατηγικής διαχείρισης.

Λέξεις ευρετηρίου: Αγενεσία νεφρού, Ανωμαλίες του σωλήνα Müller, Διαταραχή ηλεκτρολυτών, Υποκαλιαιμία, Υποκλινικός υποθυρεοειδισμός

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