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Urological management of the metabolic complications of the distal tubular acidosis (Albright's syndrome)

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Introduction: The Albright's syndrome or distal renal tubular acidosis reflects the primary kidney's inability to excrete the acid ions. The association with a hearing impairment is common as in many hereditary kidney diseases and suggests either structural or functional similarities between the two bodies, or common steps during embryonic development. As a well defined clinico-biological entity, the primitive kidney's lesion is responsible for metabolic disorders that may be the cause of urological complications requiring repeated surgery.

Material and Methods: We present the observation of two patients with Albright syndrome complicated by a large mass calculi. We discuss the aspects of urological and nephrological management of this condition whose ultimate evolution is chronic renal failure.

Results: The Immunohistochemical analysis of renal biopsies of our patients revealed a lack of H+ATPase pump in the intercalated cells of the collecting ducts. This metabolic acidosis causes hypercalciuria and other negative effects on calcium balance. Our two patients have an occurrence of kidney stones and calcifications. This complication during follow-up despite early diagnosis and treatment is due to hypercalciuria, which is an independent risk factor (age and acidosis severity) of nephrocalcinosis. The recurrence of these kidney stones is well known for its adverse effects on renal function with early onset chronic renal failure. In the two observations, we performed a multitude of interventions including flexible ureterorenoscopy, PCNL and SWL with the persistence of residual stones.

Conclusion: The Albright's syndrome is related to an accumulation of acid in the body. This is a very rare condition especially in adults characterized by a lack of acidification of urine. It is the cause of nephrocalcinosis and kidney stones. Its management requires a close collaboration between nephrologists and urologists.

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