Rare Case of Nonarteritic Anterior Ischemic Optic Neuropathy and Blindness Due to Calciphylaxis

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Introduction:

Calciphylaxis is a disorder of small vessel calcification associated with the development of progressive cutaneous plaques and ulcers due to ischemia. It occurs primarily in patients with CKD and is especially seen in those with ESRD who are undergoing hemo- or peritoneal dialysis. Calciphylaxis of temporal artery in ESRD leading to nonarteritic anterior ischemic optic neuropathy and blindness has rarely been described and on review of literature, only 4 cases were reported.

Case presentation:

A 69-year-old male patient, smoker, hypertensive and diabetic for 30 years. He presented with rapidly progressive visual diminution in both eyes secondary to anterior ischemic optic neuropathy. MRA revealed optic nerve ischemia/inflammation, but with negative complete immune panel. Temporal artery biopsy was performed and revealed coarse marked calcifications in the muscular layer of the arterial wall with NO vasculitis. There was laboratory evidence of anemia, hyperphosphatemia and hypercalcemia and rising kidney functions. Imaging revealed widespread arterial calcification. This was shortly followed by the development of painful necrotic skin ulcers on the proximal and distal lower limbs as well as the glans penis.
The patient gave history of aortic stenosis and valve replacement and has been on coumarin derivative since then. He had atrial fibrillation, severe tricuspid regurge with progressing pulmonary hypertension and volume overload. The progressing odema was refractory to diuretics with rise of kidney functions necessitating intermittent hemodialysis for 8 months prior to his presentation.
The diagnosis of calciphylaxis was postulated based on the characteristic skin lesions and cluster of risk factors as history of diabetes, use of coumarin derivatives, hemodialysis and increased Calcium x Phosphorus product ($76mg^2/dL^2$). Accordingly punch biopsy of the thigh lesion was performed and confirmed the diagnosis of calciphylaxis. After hospitalization, broad spectrum antibiotics were given, wound care and debridement of the left thigh was carried out. Low calcium dialysate and high dose of sevelamer hydro-chloride was administered with a cinacalcet hydrochloride dosage of 180mg per day. Frequency of hemodialysis was increased and the patient improved regarding dyspnea and O2 saturation levels. High pressure angioplasty was performed for the left superficial femoral artery with excellent angiographic results. He was evaluated by the urology team who diagnosed him with dry gangrene of the glans penis 2ry to calciphylaxis and was scheduled for a partial pencetomy. 2 months later, the patient showed dramatic improvement of his skin lesions.

Conclusion:

Calciphylaxis is characterized by: Arteriolar medial calcification, thrombotic cutaneous ischemia, necrotic skin ulceration and high mortality rate especially in proximal skin lesions. It is crucial to maintain calcium and phosphorus homeostasis to prevent both acute and chronic complications for CKD patients. Despite a multi-interventional approach for calciphylaxis, the disease remains associated with a high morbidity and mortality especially in the presence of proximal skin lesions, in light of this, the dramatic improvement in the afore-mentioned patient was striking.Calciphylaxis of temporal artery in ESRD leading to nonarteritic AION and blindness has rarely been reported.

Key words:
Calciphylaxis, Anterior ischemic optic neuropathy, blindness.