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MYSTERIOUS RASH NOTICED IN HEMODIALYSIS PATIENT: HERPES ZOSTER OPHTHALMICUS

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Introduction

Mortality in ESRD patients with an inpatient zoster diagnosis is increased with older age and higher severity of clinical comorbidities. The role of zoster vaccination on mortality in this population remains to be defined. Herpes zoster (HZ), which is caused by the reactivation of a latent varicella-zoster virus (VZV) infection within the cranial or dorsal root ganglia, typically manifests as a characteristic vesicular rash with a unilateral distribution limited to a single dermatome. In one study, using iron preparations and 1α-hydroxylated vitamin D was potentially associated with less risk of developing HZ reactivation in maintenance hemodialysis patients.

Patient presentation

An 82-year-old female patient, known to have congestive heart failure, chronic hypercapnic respiratory failure on long-term oxygen, dyslipidemia, osteoporosis with a history of left neck of femur fracture, and end-stage renal disease requiring initiation of hemodialysis.

She came to the outpatient dialysis unit for her second scheduled session following hospital discharge with a vesicular rash on her right forehead involving the right eyelid and redness, there was no discharge.

Examination

Vital signs showed a respiratory rate of 22 breaths per minute, her blood pressure was 151/79 mmHg and oxygen saturation was 95% on room air. Eye examination showed reactive pupils bilaterally, right upper lid edema, erythema, and ruptured vesicles, involving the right side of the forehead. There are no vesicles on the lid margin, the cornea is clear in both eyes, and there is no pseudo-dendritic ulcer. She was started on renal dose intravenous and topical acyclovir. She had a reduction in her level of consciousness and was started on intravenous antibiotics to cover the possibility of meningitis and encephalitis including ampicillin, ceftriaxone, and vancomycin.

Progression

She developed septic shock with high inflammatory markers. Her chest x-ray showed an enlarged cardiac silhouette and left-sided blunting of costophrenic angle representing pleural effusion. It increased Hazy opacification of both lung fields more prominent in the inferior zones. Abdominal x-ray showed gas-filled distended loops of bowel noted in 4 quadrants of the abdomen with no transition or significant air-fluid levels, in a picture of pseudo-obstruction. Computerized tomography of the brain showed age-related changes noted in brain parenchyma. Old infarcts in occipital regions on both sides. No fresh intracranial bleed, midline shift, or any mass effect was seen.

Conclusion

Herpes zoster can affect immunocompromised patients including ESRD on hemodialysis, It can carry high mortality among those patients.

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IT'S RARE BUT THERE: SEVER ANGIOEDEMA FOLLOWING RITUXIMAB TREATMENT FOR SEVER LUPUS NEPHRITIS

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Introduction

Rituximab is a genetically engineered chimeric murine/human monoclonal IgG1 kappa antibody directed against the CD20 antigen. Rituximab has been associated with hypersensitivity reactions (HSRs), which can be classified as infusion-related, cytokine-release, type I (IgE/non-IgE), mixed, type III, and type IV reactions. Immediate infusion-related reactions to rituximab are quite common and decrease in frequency with subsequent infusions. However, severe infusion-related reactions develop in about 10% of patients, which prevents its use. Some of the immediate infusion reactions are due to cytokine release, but some raise concerns for type I (IgE/non-IgE) hypersensitivity. Recent studies have shown the presence of serum anti-rituximab antibodies, represented by the IgG or IgE isotype.

A newer classification includes five groups: (Type α) high cytokine levels, (Type β) hypersensitivity reactions, IgE, IgG, and T cell mediated reactions, (Type δ) immune imbalance syndrome, (Type γ) cross-reactivity with native proteins, and (Type ε) non-immunologic adverse effects.

Case presentation

A 17-year-old girl, a known case of Systemic lupus erythematosus was diagnosed in 2021, with antiphospholipid syndrome. A kidney biopsy done in 2022 confirmed the diagnosis of lupus nephritis class IV. She was on tacrolimus 2 mg, mycophenolate mofetil 1 g bid, prednisolone telmisartan-amlodipine 80mg-10mg, and hydroxychloroquine 300 mg. She presented to the emergency room with complaints of joint pain mainly in the left-hand small joints and left knee. She was admitted with a lupus flare-up with oliguric AKI. Her creatinine on admission was 165 micromol/L peaked at 391 micromol/L, with severe acidosis and hyperuricemia 923 micromol/L and she was started on hemodialysis. Her urine analysis showed RBCs >100 x10^6 and a protein creatinine ratio of 5.74 g/g, her C3 and C4 were very low. She underwent a kidney biopsy which showed diffuse lupus nephritis ISN/RPS Class IV-G, active with membranous lupus nephritis ISN/RPS Class V. She was started on plasma exchange for her severe flareup, and received rituximab infusion after the third session which was complicated by severe allergic reaction. She had severe swelling of her tongue with severe oral bleeding complicating her angioedema. She had an emergency tracheostomy underwent bilateral lingual artery embolization and was admitted to intensive care.

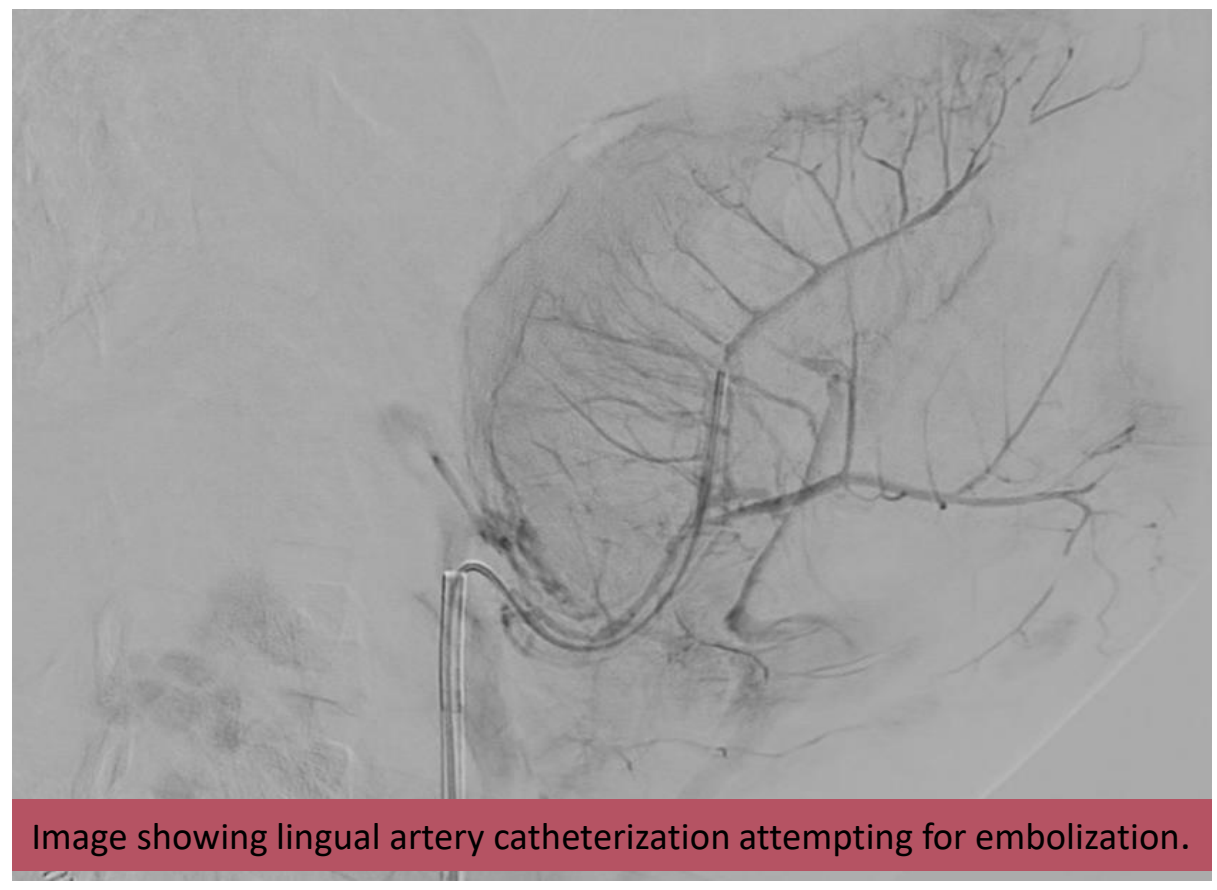

Image showing lingual artery catheterization attempting for embolization.

Results

She completed 5 sessions of plasma exchange and remained on dialysis-dependent for 3 months. Her admission was complicated by bilateral thigh cellulitis with necrotic changes and necrotizing fasciitis was ruled out. She underwent multiple surgical debridement and her wound condition improved. Her renal function improved dramatically, her urine output increased gradually to more than 1 L and her repeat lupus markers showed normal double-stranded DNA and high normal C3 and C4. MMF was increased to 1g.

Conclusion:

Rituximab can cause a life-threatening reaction, part of the treatment includes emergency tracheostomy for airway compromise, IV steroids, and plasma exchange following the rituximab exposure that can remove up to 50 % of the amount. Our patient had renal recovery and was dialysis-free at 3 months of her hospitalization.

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2 CASES WITH IGM NEPHROPATHY

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Introduction

The reported frequency of IgMN in the literature has varied from 1.8% to 18.5% in native biopsies. IgMN appears to have different clinical outcomes at different ages. It mainly presents proteinuria or haematuria in young adults or children. Proteinuria in IgMN can range from asymptomatic proteinuria to nephrotic syndrome. IgM deposits may be found in patients with MCD, FSGS, and mesangial proliferative glomerulonephritis. The presence of IgM deposits in a patient with MCD signifies a poorer prognosis compared with MCD without deposits, as fewer than 50 % of such patients respond to glucocorticoids.

Case 1

19-year-old male patient, known to have nephrotic syndrome since the age of 3 with frequent relapses. He received multiple courses of immunosuppression medications, including multiple courses of steroids, tacrolimus, 5 doses of rituximab, and cyclophosphamide for 3 months in 2010. He was admitted to our facility with a relapse of his nephrotic syndrome in the form of generalized body edema and weight gain of 9kgs, his urinary protein was >6g/L, started on oral pulse steroids in the form of prednisolone at 1 mg/kg/day.

Case2

37-year-old male known case of mesangioproliferative GN proved in a biopsy done 2 years ago following Covid infection. He was on mycophenolate mofetil, but it was stopped for financial reasons. He presented with generalized and facial edema and acute kidney injury with proteinuria of 6.49 g. his serum albumin was 6 g/. he was started on pulse steroids with a kidney biopsy done and showed IgM (++) on immunofluorescence and effacement of the podocyte food process of more than 80 % which is consistent with severe podocytopathy. He restarted MMF and prednisolone with tapering dose and his urine protein dropped to 129mg/L in 2 months time.

Results

Kidney biopsy images for the first patient showing mild mesangial expansion, Unremarkable tubules, and interstitium with IgM (2+) and C1q (1+), kappa (1+) and lambda (1+) light chains . The glomeruli show no immunoreactivity to IgA, IgG, C3, fibrinogen, and albumin. Tubular protein reabsorption droplets are immunoreactive for IgA, IgG,C3, albumin, kappa, and lambda light chains. Electron Microscopic study showed focal small mesangial electron-dense deposits. The podocytes are vacuolated and swollen. The podocytes' foot processes are severely effaced and show diffuse effacement with an overall involvement of approximately 90% of the capillary surface. The features are consistent with IgM nephropathy.

Conclusion

IgM Nephropathy is considered a distinctive pathological entity with a lot of controversy and may not be distinguished from Minimal change disease or FSGS, it carries variable response to steroids.

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PREGNANT PATIENT WITH SEVER PREECLAMPSIA AND SEVER IUGR RECEIVING SODIUM ZIRCONIUM

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Introduction

Gordon syndrome is a rare inherited monogenic form of hypertension, which is associated with hyperkalaemia and metabolic acidosis. Since the recognition of this predominantly autosomal dominant condition in the 1960s, the study of families with Gordon syndrome has revealed four genes WNK1, WNK4, KLHL3, and CUL3.Pregnancy in women with Gordon syndrome appears to be associated with a significant risk of adverse pregnancy outcomes, particularly where there is maternal hypertension preconception. No pregnancy registry exists for Gordon syndrome. The available data is limited to case reports and small case series and may be affected by bias. However, the medication US FDA pregnancy category is not assigned. Sodium zirconium is not systematically absorbed following oral administration; therefore, it is not expected to cause fetal exposure. Animal studies did not indicate direct or indirect harmful effects to reproductive toxicity.

Methods

21-year-old female patient known to have renal tubular acidosis type IV due to Pseudo-hypoaldosteronism type IID \"Gordon syndrome\" confirmed by genetic studies (KLHL3- homozygous variant) and chronic hyperkalemia. There is a Strong family history of PHA2 and a similar gene mutation (KLHL3- homozygous variant) with her brother and cousin. Her home medications include hydrochlorothiazide and sodium bicarbonate. She presented to the prenatal clinic for follow-up. She was para 1 +0 at 31 weeks. She had lower limb swelling, and her BP was elevated at 140/100 mmHg. Her K level was 6.8 mmol/L, then dropped to 5.7 mmol/L. She was started on sodium zirconium 10 mg three times a day and a low K diet 24 hours before her delivery. Her antenatal ultrasound showed abnormal Umbilical Artery Doppler: absent end diastolic flow. She was given dexamethasone, IV labetalol, and mg sulfate, and she had brisk reflexes. Emergency C/S was performed, and the baby cried immediately with a weight of 1090 g.

Results

She was started on sodium zirconium 10 mg three times a day and a low K diet 24 hours before her delivery. Her antenatal ultrasound showed abnormal Umbilical Artery Doppler: absent end diastolic flow, she was given dexamethasone, IV labetalol, and mg sulfate, and she had brisk reflexes. Emergency C/S was performed, and the baby cried immediately with a weight of 1090 g.

Conclusion:

Rare conditions like Gordon syndrome are difficult to manage during pregnancy. The use of new medications like Sodium zirconium is assumed safe during pregnancy as it is not systematically absorbed however in such a complex case relationship between medication rare risk of edema and patient symptoms cannot be completely excluded.