CASE STUDY: GOODPASTURE'S SYNDROME

Goodpasture’s Syndrome: A Case Study from Southeastern New Mexico

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Abstract
Goodpasture’s syndrome is not a common cause of renal failure but was observed with higher than expected frequency in dialysis clinic admissions in southeastern New Mexico. A case study is presented of the patients with this diagnosis. Comparisons and contrasts of the patients suggest that while patients had many similarities there were some common threads of low adherence to treatment plan including unmanaged high blood pressure, elevated potassium and phosphorus levels, missing and shortening treatments, a relatively young age with expressed interest for transplant, and alleged drug use. The low level of personal acceptance of the patients’ disease state along with their socioeconomic challenges of drug use contributed to a shorter than expected survival rate.

Introduction
Time is a luxury for the Registered Dietitian/Nutritionist (RDN) working with patients with renal impairment.1 Patients needing nutrition care for Chronic Kidney Disease (CKD) and end-stage renal disease (ESRD) continue to grow in number.2 The Centers for Medicare and Medicaid Services (CMS) continue to increase expectations of the entire health care team to improve care quality and reduce costs for dialysis patients.3 Many patients admit to care with common disease states that the RDN is familiar with; however, when a patient with a rare disease comes into care, more time is needed to study and develop an individualized plan of care. The following introduces an uncommon contributor to renal failure observed at a higher than expected frequency in two clinics in southeast New Mexico.

Case Background and Literature
Goodpasture disease describes glomerulonephritis with or without pulmonary hemorrhage and the presence of circulating anti-glomerular basement membrane (anti-GBM) antibodies. The incidence of anti-GBM disease is estimated to be 0.5-1.8 cases per million annually in both European white and Asian populations and is responsible for 1-5% of all types of glomerulonephritis and for 10-20% of crescentic glomerulonephritis.4,5 This disease occurs more often in white people than black people and has been seen more commonly in certain ethnic groups such as the Maoris in New Zealand. The age distribution has been found to be bimodal, 20-30 years and 60-70 years and occurs more often in men in the younger group and women in the older group.6

Anti-GBM disease is an autoimmune disorder. There are numerous methods to classify and diagnose specifically through pathological studies. The key factor appears to be the cellular changes increasing the vascular permeability to the alveolar capillaries in the lungs. Factors that increase this permeability include increased capillary hydrostatic pressure, high concentrations of inspired oxygen, bacteremia, endotoxemia, exposure to volatile hydrocarbons, upper respiratory infections and tobacco smoking in addition to a genetic predisposition to the disease.7 Environmental factors creating the initial insult to the pulmonary vasculature exposing the alveolar capillaries to the anti-GBM antibodies include exposure to organic solvents or hydrocarbons, smoking, infection, cocaine inhalation, exposure to metal dusts, lymphocyte depletion therapy, and extracorporeal shock wave lithotripsy.7

Treatment of Goodpasture disease includes plasmapheresis, corticosteroids, and immunosuppressive agents to increase the 5-year survival rate to 80% with fewer than 30% needing renal replacement therapy (RRT) when progressing to ESRD. Unfortunately, the aggressive treatments have not been found successful for patients already at ESRD.6 The median survival rate overall is 5.93 years. Early recognition and treatment of the disease are critical because the prognosis for recovery of renal function depends on the initial extent of the injury.

Teague et al., documented 29 cases of Goodpasture’s syndrome in New Zealand over a 13-year period.8 While presentation was varied, 12 of the 29 required dialysis with six needing bilateral nephrectomies early in the disease presentation. Those studied had varying survival rates of less than one week up to two years who needed dialysis. Participants who did not need dialysis survived 30 weeks up to 14 years. More of the patients were ages 25 and under and there were more males than females.9 A review of the occupations of those studied suggests that many could have been exposed to chemicals in their workplace. Renal health has been noted as negatively impacted by organic solvent exposure.9

Introduction to Case Environment
Three patients with Goodpasture’s syndrome were identified in two southeastern New Mexico hemodialysis clinics between 2012-2017, which is higher than expected presentation rate considering the average census of the two clinics combined was 110. A review of water quality from the community where two of the three patients with Goodpasture’s disease resided suggested that the water provided by the municipality did not have solutes above safe levels.12 However, many people in this area, due to socioeconomic conditions, rely on well water that is not processed by the municipality.

The southeast New Mexico region has one of the highest deposits of pure potash (potassium chloride and sodium chloride) in the United States.10 The minerals are mined underground and brought
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to the surface for manufacturing, most commonly into fertilizer products. A review of the ground-water conditions of southeast New Mexico in the mid-20th century suggested that water needs were filled almost entirely from ground water in aquifers. It was noted that the water supply was already being contaminated with seepage of oil-field brines and other oil-field byproducts. Oil field production has skyrocketed in this area in the 21st century. These factors contribute to a more unique environment and thus is of note when looking at onset of rare conditions.

Analysis of Patient Data

Three patients were admitted to the hemodialysis clinics with diagnosis of Goodpasture's syndrome. The population of the centers combined were 100 with one designated full time RD with a 75-mile travel trip in between the clinics. Time allocated was 32 hours per week for the larger center of 80 and 8 hours a week for the more remote center with 30 patients. Data was collected from the patient medical records both electronic and paper based and identifying medical information was removed (Table 1).

The two males presenting with the disease were young (between 20-35 years of age) and their course on dialysis was rather short ranging from 1 year and 2 months to 2 years 7 months. The female patient had been diagnosed prior to starting dialysis while the males presented to the emergency department (ED) with serious symptoms. Two of the three patients worked in customer service while the other patient chose not to work. The female patient worked in an environment with potential exposure to hydrocarbons (auto parts and repair).

Family situations were unstable for all patients. Two of the three did not live with their nuclear family but did not live independently. The female lived at her parents with her children. Two patients were financially unstable. Housing situations were substandard for two of three patients living in homes without adequate utilities or repair. All three were substance users, took anti-anxiety and pain medications and used tobacco.

Providers’ notes included frequent hospitalizations, missing treatments, and inconsistency of taking medications. Nursing also noted missing treatments, refusal to remove catheter and documenting the advanced bone disease of Patient A. The social worker followed each case extensively due to the high needs resulting from family and financial issues, problems with transportation and referral needs to services.

Nutrition care was complex due to chronic elevations of phosphorus and potassium issues. Patients were not able to make progress with nutrition care tools due to their unstable situations of being incoherent during treatment or expressing disinterest in counsel to improve. Routine labs were rarely close to target ranges. Drug tests were ordered for patients by providers due to behavior in clinic. These results were not shared with the clinic staff.

Two of the patients had additional providers on the case. One male was a potential transplant candidate whose family insisted on living donation only. He also saw the rheumatologist for his very advanced tophaceous gout but did not live long enough to have additional treatment. This patient also sought help from a pain specialist. Of note all three patients expressed high levels of pain which were difficult to manage. The female had been treated prior to admission for the Goodpasture’s diagnosis.

All causes of death were tragic and untimely. Two of the three patients died due to a drug overdose. The third patient died unexpectedly of cardiac arrest.

Discussion

The home situations of the patients were substandard and likely included use of a well at their home and insufficient plumbing. These patients may have increased their likelihood to develop Goodpasture’s with the level of contaminants and hydrocarbons in the ground water.

The age of the patients contributed to a low acceptance of the disease process and thus cooperation with their plan of care. The patients were in difficult family situations and attempted to seek some level of independence even though financially they were unable to live on their own. Their resistance to following directions about their medical care and substance use reflected this behavior. Age and poor outcome were also noted by Teague et al. Patient reliance on and time spent with providers were higher than the other patients in the clinic. Numerous interventions, clinical notes, narratives from providers, nurses, social workers and dietitians suggest that even though the patients weren’t able to manage their own care, they were willing to share their concerns and seek interventions from caregivers when caregivers were easily accessible. While one patient had frequent hospitalizations and sought other consultations, most of the requests and interaction were in the clinic to address patient concerns. The remote location of one of the units may have hindered a more effective connection with the patients as the one day a week the RD, social worker (SW) and providers rounded at the unit possibly coincided with a missed treatment or a day when the patient was not open to counsel.

All three patients had chronic complaints of intense pain and anxiety. They were prescribed pain and antianxiety medications but the patients chronically requested increases in the dosage. Patient A sought a pain specialist who remarked, "Patient reports self-medicating large doses of hydrocodone and oxycodone up to 8-12 tablets of 10 mg daily. Started on Dilaudid and fentanyl.

Table 1 Abbreviations

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
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<tr>
<td>c/o</td>
<td>complaining of</td>
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<tr>
<td>CKD</td>
<td>Chronic Kidney Disease</td>
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<tr>
<td>CXR</td>
<td>Chest X-Ray</td>
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<tr>
<td>ED</td>
<td>Emergency Department</td>
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<tr>
<td>HTN</td>
<td>Hypertension</td>
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<tr>
<td>iPTH</td>
<td>intact Parathyroid Hormone</td>
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<tr>
<td>IDWG</td>
<td>intradialytic weight gain</td>
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<tr>
<td>K</td>
<td>potassium</td>
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<tr>
<td>KDOQOL</td>
<td>Kidney Disease Quality of Life (assessment tool for social workers)</td>
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<tr>
<td>MCS</td>
<td>Mental Component Score of KDOQOL</td>
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<tr>
<td>PCS</td>
<td>Physical Component Score of KDOQOL</td>
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<tr>
<td>PD</td>
<td>Peritoneal dialysis</td>
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<tr>
<td>Phos</td>
<td>phosphorus</td>
</tr>
<tr>
<td>SW</td>
<td>social worker</td>
</tr>
<tr>
<td>RCS</td>
<td>Role Social Component Summary of KDOQOL</td>
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<tr>
<td>THC</td>
<td>Tetrahydrocannabinol</td>
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**Table 1: Clinical Course of Goodpasture Patients**

<table>
<thead>
<tr>
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<th>Patient A</th>
<th>Patient B</th>
<th>Patient C</th>
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</thead>
<tbody>
<tr>
<td><strong>Age Range</strong></td>
<td>20-25</td>
<td>30-35</td>
<td>20-25</td>
</tr>
<tr>
<td><strong>Gender</strong></td>
<td>Male</td>
<td>Female</td>
<td>Male</td>
</tr>
<tr>
<td><strong>Time on Dialysis</strong></td>
<td>2 years 0 months</td>
<td>2 years 7 months</td>
<td>1 year 2 months</td>
</tr>
<tr>
<td><strong>Cause of Death</strong></td>
<td>Cardiac arrest</td>
<td>Drug overdose</td>
<td>Drug overdose</td>
</tr>
<tr>
<td><strong>Access History</strong></td>
<td>Catheter to PD catheter, back to catheter and graft</td>
<td>Catheter 2 years, 6 months to graft 1 month</td>
<td>Catheter 5 months to fistula 9 months</td>
</tr>
<tr>
<td><strong>Initial Hospital Course</strong></td>
<td>Presented to ED</td>
<td>Diagnosed with positive antiglomerular basement antibodies for &lt; 6 months, HTN, CKD prior to admission</td>
<td>Presented to ED</td>
</tr>
<tr>
<td><strong>Vocation</strong></td>
<td>Customer service</td>
<td>Customer service</td>
<td>None</td>
</tr>
<tr>
<td><strong>Family Situation</strong></td>
<td>Preferred to stay with grandparents, mother frequently brought inappropriate foods to treatment</td>
<td>Lived at home with parents and her children; food insecurity expressed</td>
<td>Estranged from parents, stayed with siblings</td>
</tr>
<tr>
<td><strong>Substance Use</strong></td>
<td>Tobacco use, ½ pack per day. Patient denied heavy alcohol or illicit drug use; pain medications – hydrocodone, oxycodine</td>
<td>Tobacco, clonazepam</td>
<td>Hydrocodone, temazepam, oxazepam, THC, opiates</td>
</tr>
<tr>
<td><strong>Nephrology Team Follow</strong></td>
<td>6 hospitalizations recorded in 7 months</td>
<td>Notes include chronic catheter presence with missing appointments and elevated phosphorus and blood pressure – not refilling/taking medications. Pt faithful with Requip and Xanax refills</td>
<td>Seen monthly, patient was asked to be referred to psychological counseling due to issues expressed to provider</td>
</tr>
<tr>
<td><strong>Nursing Follow</strong></td>
<td>Catheter removed 8 months prior to death, often arrived late with shortened treatments; family was counselled about milkshakes brought to unit month prior to death; patient unable to move due to joint bone pain and started to arrive in a wheelchair.</td>
<td>Noted as missing treatments or having a clotted access requiring return to catheter several times.</td>
<td>Missed treatments or was medicated per pt request to sleep through treatment.</td>
</tr>
<tr>
<td><strong>SW Notes</strong></td>
<td>Last KDQOL listed PCS and MCS including burden of disease. No concerns noted in prior assessment by SW while pt on PD</td>
<td>Patient received assistance with rides to doctor appointments. Unable to locate other SW records</td>
<td>8 months prior to death- RCS and MCS scores within average. Financial issues. Multiple referrals for psychological intervention, patient did not attend appointments</td>
</tr>
<tr>
<td><strong>Nutrition Care Summary</strong></td>
<td>Logged consistent elevated potassium and phosphorus during last 6 months of life, Phos up to 12, K up to 7.1. C/o diarrhea. Brought 32 oz milkshakes from fast food chains to treatment.</td>
<td>Chronic elevated phosphorus due to not taking binders and diet with highly processed foods. Weight was challenged several times. Elevated K with occasional 1K bath.</td>
<td>Frequent mention of having no food. Inconsistent labs of elevated phos, potassium and then not. Albumin normalized after 6 months of treatments</td>
</tr>
<tr>
<td><strong>Pertinent medical information prior to death</strong></td>
<td>Labs: Albumin 3.7 (oral supplement), K 6.5, Phos 8.9, Calcium (total) 9.2, iPTH 810, IDWG: 3.09 kg</td>
<td>Drug tests ordered 2 and 1 month prior to death</td>
<td>Noted with borderline cardiomegaly CXR 4 months prior to death</td>
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every 2 hours but always rated pain as a 10. Patient has a low
time threshold, inappropriate expectations of relief and worsened
chronic depression.” Separating the pain of their disease process
from their complex socioeconomic situation may provide
additional insight of the disease progression from a clinical
standpoint. All three patients resisted removal of catheters. Patient
B did not have hers removed until the month of death. A common
practice of individuals using recreational drugs is to utilize
catheters for increasing the effect of the drug.13

All three patients died soon after starting treatment.
Socioeconomic complexities combined with an advanced disease
state increased the difficulty for both the interdisciplinary team
(IDT) and the patients to manage their disease effectively for
longer term survival.

Conclusions

Working with individuals with Goodpasture’s syndrome
presented clinical and social challenges while providing
hemodialysis treatment. These patients required a high number
of hospitalizations, physician time, and interdisciplinary team
member effort and counsel without improvement in condition or
outcome. This case study was affected by the relatively low age
of the patients and their socioeconomic challenges especially drug
use. However, the patient data may contribute to helping future
Goodpasture’s patients that present to these clinics.. The patient
survival rate was similar to prior observed cases with the younger
Goodpasture’s patients that present to these clinics..

The interdisciplinary team of this rather rural, remote area
worked forward with a more cohesive communication system but
the case continues to present a quality improvement opportunity
to improve future outcomes and longer survival on hemodialysis.
Literature following patients with Goodpasture’s through the
clinical course of kidney disease is limited. More work is
needed to further develop characteristics, clinical course, and
successful interventions in working with these patients. Future
data collection is likely to include more focus on environmental
factors to better understand the association between the unique
physical environment of this geographic area.

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