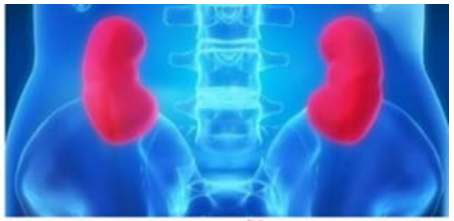
قطب علمی آموزشی نفرولوژی مرکز تحقیقات نفرولوژی

In The Name of God

Case Presentation

A 23 Y O Girl with Hamaturia

Patient Data:



قطب علمی آموزشی نفرولوژی مرکز تحقیقات نفرولوژی

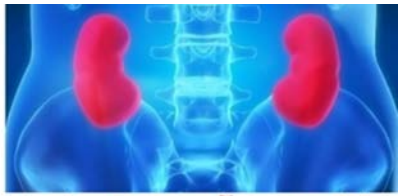
A 23 Y o girl with hamaturia (Not in period time) and HTN

BP and azotemia: 160/90

Family history: Negative

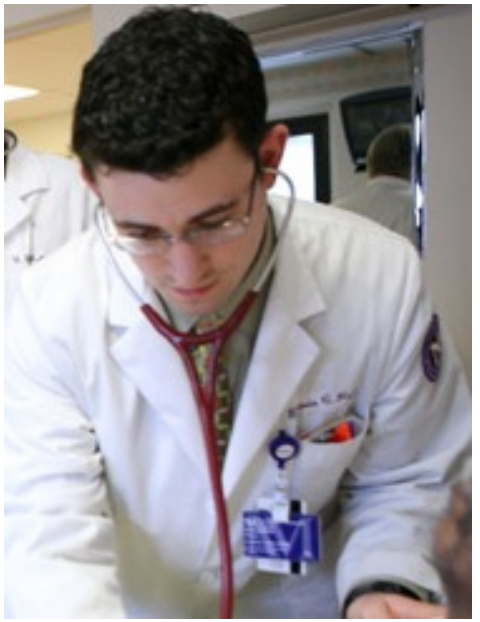
Weakness, Paleness, Headache

Anemia can cause fatigue, headaches, pale skin, and other symptoms.



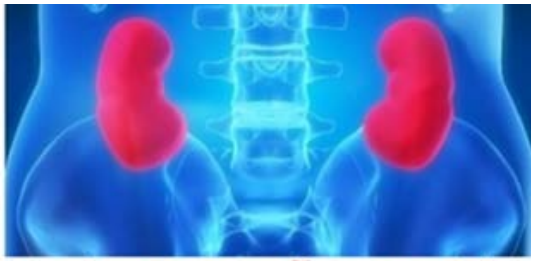
A **targeted approach** should be performed in patients with an **unusual presentation** of **HTN and Hematuria.**

What is your Assessments?



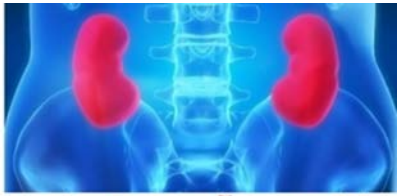
Laboratory Tests

- Electrolytes(Na, K, & serum Cr (Calculate eGFR)
- FBS
- Urinalysis
- Lipid profile (total-, HDL- & LDL-Ch & TG)
- ECG



مرکز تحقیقات نفرولوژی قطب علمی آموزشی نفرولوژی

Imaging



قطب علمی آموزشی نفرولوژی مرکز تحقیقات نفرولوژی

-Sonography

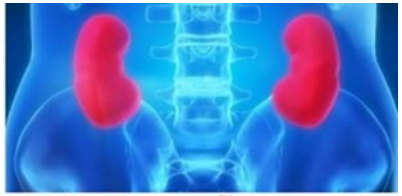
-Doppler Sonography

-CT Scan

-CT Angiography

-Angiography

Patient Data:



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WBC: 6700, Hb: 10/5, Plt:

FBS: 98 mg/dl

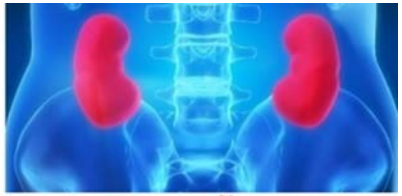
BUN: 32 mg/dl, Cr: 2/6 m

U/A: Protein 2+, RBC: 10-12,

Anemia can cause fatigue, headaches, pale skin, and other symptoms.

- *Red Urine & Common Cold was Coincident*

Patient Data:



قطب علمی آموزشی نفرولوژی مرکز تحقیقات نفرولوژی

-Sonography Normal

-Dysmorphic RBC was seen.

Anemia can cause fatigue, headaches, pale skin, and other symptoms.

What is new assessments?

Na, K, IgA, HBSAg, HCVAb, HIVAb, Anti GBMAb, ANA, AntidsDNA, C3, C4, CH50, ANCA(P&C), Cryoglobulin, Urine Output,

-Sonography

-Doppler Sonography

& maybe…

-CT Scan?

-CT Angiography?

-Angiography?

-Kidney Biopsy?

Patient Data:

IgA titer is high and Kidney biopsy show IgA deposition in mesangium, mesangial & endocapillary hypercellularity

قطب علمی آموزشی نفرولوژی مرکز تحقیقات نفرولوژی

IgA nephropathy is the most common pattern of primary glomerulonephritis seen in the Western world.

Seminars in Nephrology

Volume 28, Issue 1, January 2008, Pages 27-37

Cases of
familial &
secondary IgA nephropathy
show
underlying genetic &
environmental triggers for this
disease.

Secondary IgA nephropathy is seen most commonly in patients with **liver disease** or **mucosal inflammation**, in **particular** affecting the **GI** tract.

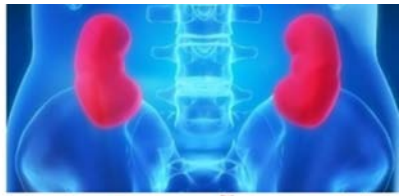
Dietary & microbial Ags have been identified in circulating IgA immune complexes & mesangial IgA deposits, suggesting that environmental factors may play a role in the pathogenesis of IgA nephropathy.

IgA nephropathy

-Present at any age

-Peak incidence: second & third decades of life.

-Male/female ratio: 2:1



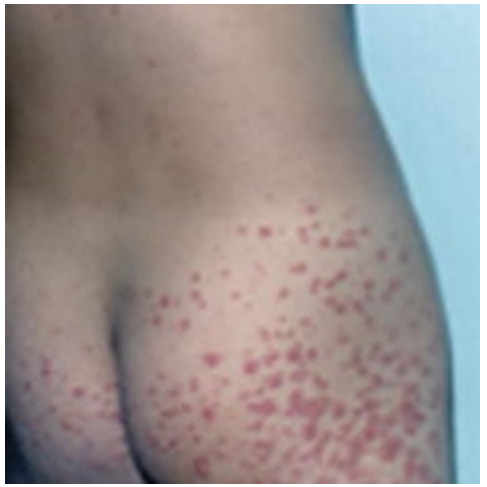
In a Chinese study of 13,519 renal biopsies, IgA nephropathy constituted 45% of all cases of primary GN .

IgA deposition does not necessarily followed by nephritis

There are a number of reports documenting IgA deposition in other forms of GN include

- -Thin basement membrane nephropathy,
- -Lupus nephritis,
- -Minimal Change Disease, &
- -Diabetic nephropathy.

The immunofluorescence findings in IgA nephropathy are indistinguishable from those in **Henoch-Schönlein purpura (IgA vasculitis)**



Oxford classification of IgA nephropathy

-Mesangial hypercellularity

-Segmental glomerulosclerosis

-Endocapillary hypercellularity

-Tubular atrophy/interstitial fibrosis

CLINICAL FEATURES

Visible hematuria (40-50%) usually following an upper respiratory infection (**synpharyngitic hematuria**), bacterial tonsillitis, or by other viral upper respiratory infections even with **tonsillectomy**.

Low-grade fever

Microscopic hematuria (30-40%)

Less than 10 % **nephrotic syndrome or acute RPGN**

Rarely, **AKI with or without oliguria**.

IgA nephropathy

Transmitted as an **autosomal dominant** trait with incomplete penetrance.

CLINICAL ASSOCIATIONS

- Cirrhosis and other forms of severe liver disease
- Celiac disease
- HIV infection
- Minimal change disease and membranous nephropathy
- Wegener
- Dermatitis herpetiformis,
- Seronegative arthritis (particularly ankylosing spondylitis),
- Small-cell carcinoma,
- Lymphoma (Hodgkin's lymphoma and T-cell lymphomas, including mycosis fungoides),
- Disseminated TB,
- Bronchiolitis obliterans, and
- IBD

Treatments

Nonimmunosuppressive (ACEis or ARBs, and fish oil).

Immunosuppressive therapy (Glucocorticoids &…)

Isolated hematuria & minimal proteinuria (<500-1000 mg/day), & normal GFR are typically not treated & often not biopsied.

This group eriodically monitored at 6-12 month intervals for increases in proteinuria, BP & Cr.

Proteinuria >500-1000 mg/d

A slightly reduced GFR that is not declining rapidly & only mild to moderate histologic findings on renal biopsy are initially managed with nonimmunosuppressive (ACE i or ARB).

The goal is urine protein<500- 1000 mg/day & BP<130/80 mmHg.

Fish oil (3.3 g/d or more) can be tried in proteinuria >1g/d despite 3-6 mo of therapy with an ACEi or ARB.

Glucocorticoids recommended in active disease (hematuria in addition to one or more of the following):

-Increasing Cr

-Proteinuria >1 g/d after maximal antiproteinuric nonimmunosuppressive therapies

-Morphologic evidence of active disease in kidney biopsy (proliferative or necrotizing glomerular changes)

Glucocorticoid Therapy

IV methylprednisolone, 1 g, for 3 consecutive days at the beginning of months 1, 3, and 5, combined with 0.5 mg/kg of oral prednisolone given on alternate days for 6 months.

Or

Prednisone, 0.8 to 1 mg/kg/day, for 2 months followed by monthly dose reductions of 0.2 mg/kg/day during the next 4 months).

Or

Combined immunosuppressive therapy can be considered in patients with more severe disease as defined by a more rapidly progressive clinical course and/or histologic evidence of severe active inflammation.

Occasionally IgA nephropathy present with nephrotic syndrome, no hematuria, NL kidney function, minimal glomerular changes on light microscopy & diffuse fusion of the foot processes of the glomerular epithelial cells on electron microscopy.

These histologic findings are characteristic of minimal change disease.


THANK
YOU