Mrs DT

DR GWYNN MATTHIAS

HAEMATOLOGY SPECIALITY DOCTOR

87 year old lady

Rheumatoid arthritis diagnosed before 1996

Factor V Leiden diagnosed 2001

Back pain

Depression

Macular degeneration

Old right retinal vein occlusion

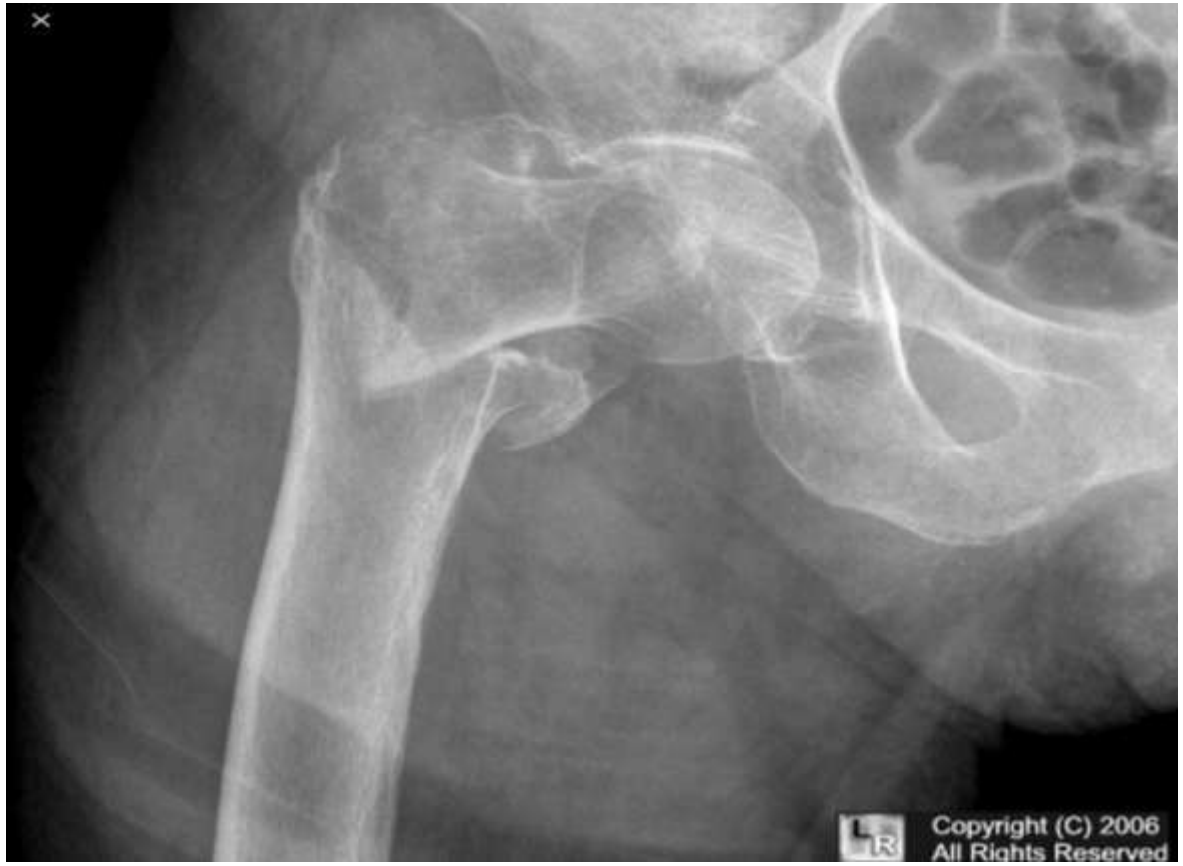
Early cataracts

Registered partially sighted

Medicines

Methotrexate 7.5 mg s/c once per week	**Bisoprolol**	**Omeprazole**
Folic acid 5 mg po once per week	Clopidogrel	Metoclopramide
Calcichew and vit D	Simvastatin	Co codamol
Paracetamol	Sertraline	Alendronic acid stopped Feb 16

Admission to ED QA 7/6/16



Admitted to ED 7/6/16 Fractured neck of femur

Normal U&E's

Normal LFT's

Normal magnesium

Glucose 6.8

Normal INR & APTR

Hb 118

WBC 13.1

Pl 277

Neut 10.8

Group &screen and 2 units of blood requested

SR Pad

Ig A deficient!

What would you do?

Solutions

Confirm it's true

2010- Ig A level < 0.0016, Ig A antibody titre-0

This was repeated to confirm levels

Ig G, Ig M normal levels

Phone a friend

Advice

No need to give Ig A deficient products but observe closely

Unlikely to develop antibodies as very low level Ig A level

Repair of fracture 8/6/16

16.00 post op Hb sent

Junior surgical team want advice

Pr-op Hb was 118

What would you do?

My advice

What is her blood pressure and heart rate

How much blood do we think was lost at surgery?

Give tranexamic acid

Do not transfuse unless Hb is <80

What happened?

Junior doctor at night recorded

Hb 78 transfuse 2 units – overnight!

Patient had no reaction- phew!

What happened next

16/6 admitted to a rehab ward

20/6 severe right upper abdominal pain

Investigations 20/6/16

Bilirubin 87 (3-20)

Albumin 21 (35-50)

ALP 456. (30-130)

ALT 178. (7-35)

CRP 120. (0-7)

Hb 101

WBC 12.2

Pl 3.8

Neut 10.6

Antibody screen Anti c & Anti E

Readmitted to SAU 21/6/16

Bilirubin 90

ALP 382

ALT 124

Albumin 19

CRP 146

Hb 89

WBC 11.8

Pl 311

Neut 9.6

Abdominal ultrasound

Extremely distended gallbladder containing a large quantity of biliary sludge.

No calculi.

No obstructive features

Multiple bilateral renal cysts 10 cmx14cm

Kidneys appear unremarkable

Post ERCP 23/6/16

Bilirubin 43

Albumin 18

ALP 311

ALT 55

CRP 130

Hb 84

WBC 6.5

Pl 259

Neut 4.7

Ig A deficiency- a few facts

Common 1:600

Inheritance is variable

Ig A levels don't usually change

Acquired :drugs, post infection, chemicals

Symptoms

Maybe asymptomatic and donate blood

Increased risk of infection associated with mucosal surface e.g. Lung and gut

Anaphylaxis to blood products

Associated with drug allergy

Autoimmune conditions

Atopy

Can also be associated with reduced classes of Ig G but total Ig G maybe normal

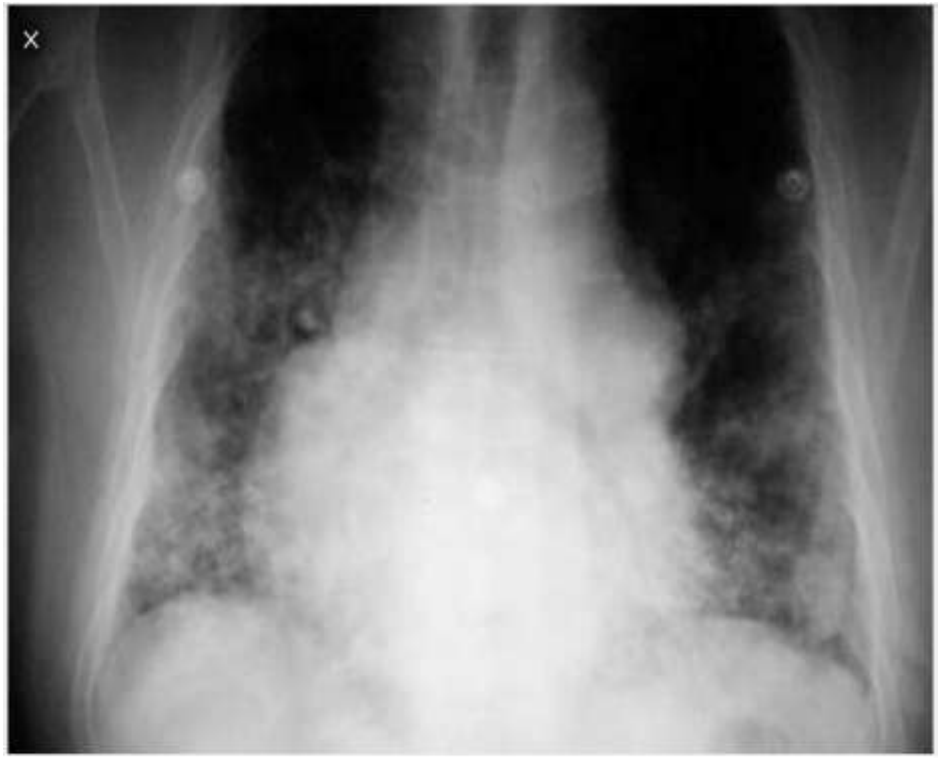
Associations of Ig A deficiency

Table – Clinical manifestations of SIGAD

Recurrent sinopulmonary infections	Autoimmune disorders	Other manifestations
Bronchiectasis	Addison disease	Systemic infections
Bronchitis	Celiac disease	Meningoencephalitis
Pneumonia	Chronic nephritis	Septicemia
GI tract infections and disorders	Dermatomyositis	Viral hepatitis
Cholelithiasis	Evans syndrome	Allergic reactions
Chronic active hepatitis	Hashimoto thyroiditis	Allergic rhinitis
Giardia lamblia infection	Henoch-Schönlein purpura	Atopic dermatitis
Inflammatory bowel disease (including Crohn disease and ulcerative colitis)	Isolated hemolytic anemia	Conjunctivitis
	Isolated idiopathic thrombocytopenic	Urticaria
Intestinal disaccharidase deficiency	Juvenile idiopathic arthritis	Malignancies
Lactase deficiency	Myasthenia gravis	Acute lymphoblastic leukemia
Lupoid hepatitis	Pernicious anemia	Gastric and colonic adenocarcinoma
Pancreatic deficiency	Pulmonary hemosiderosis	Hepatoma
Primary biliary cirrhosis	Purpura	Lymphosarcoma
	Sarcoidosis	Malignant thymoma
	Sjögren syndrome	Melanoma
	Skin disorders (including pyoderma gangrenosum, paronychia, and vitiligo)	Multiple myeloma
		Ovarian cancer
	Systemic lupus erythematosus	Squamous cell carcinoma
	Type 1 diabetes mellitus	

SIGAD, selective IgA deficiency.

Consequences of Ig A deficiency



Medscape Reference

Immunoglobulin A Deficiency: Background, Pathophysiology, Epidemiology

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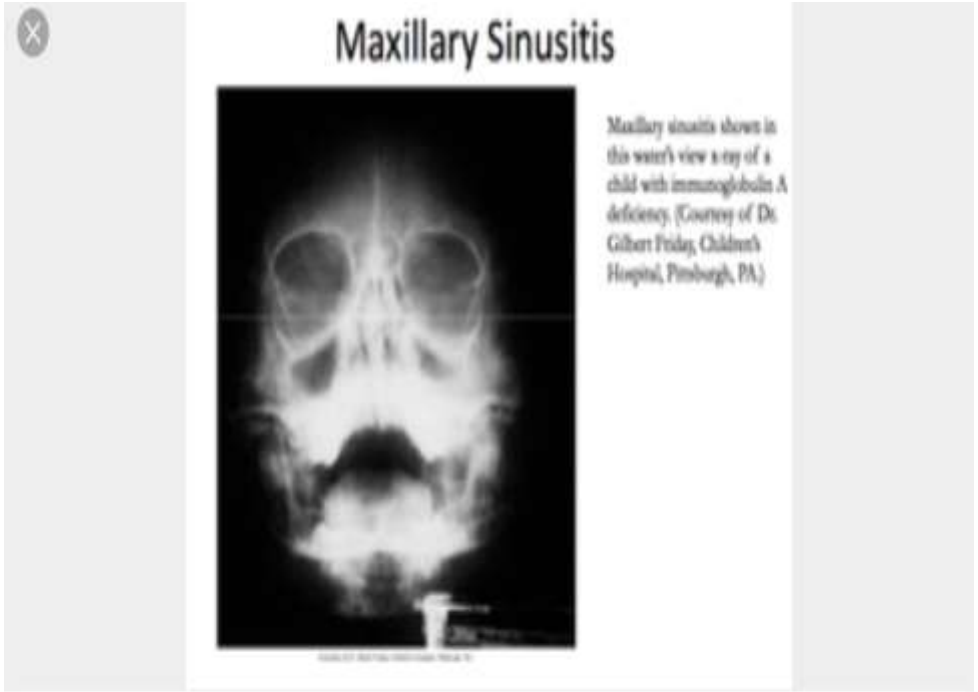

Quizlet

Pathophys Week 1: Immunology Flashcards | Quizlet

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Complications of Ig A deficiency

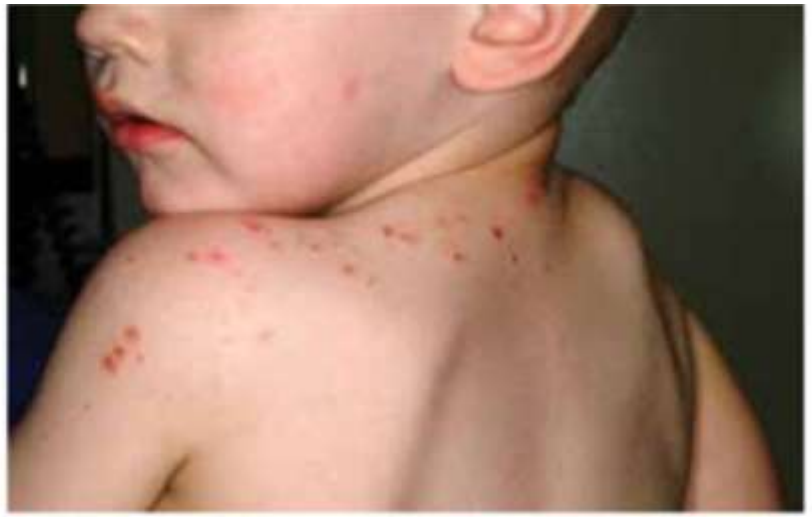
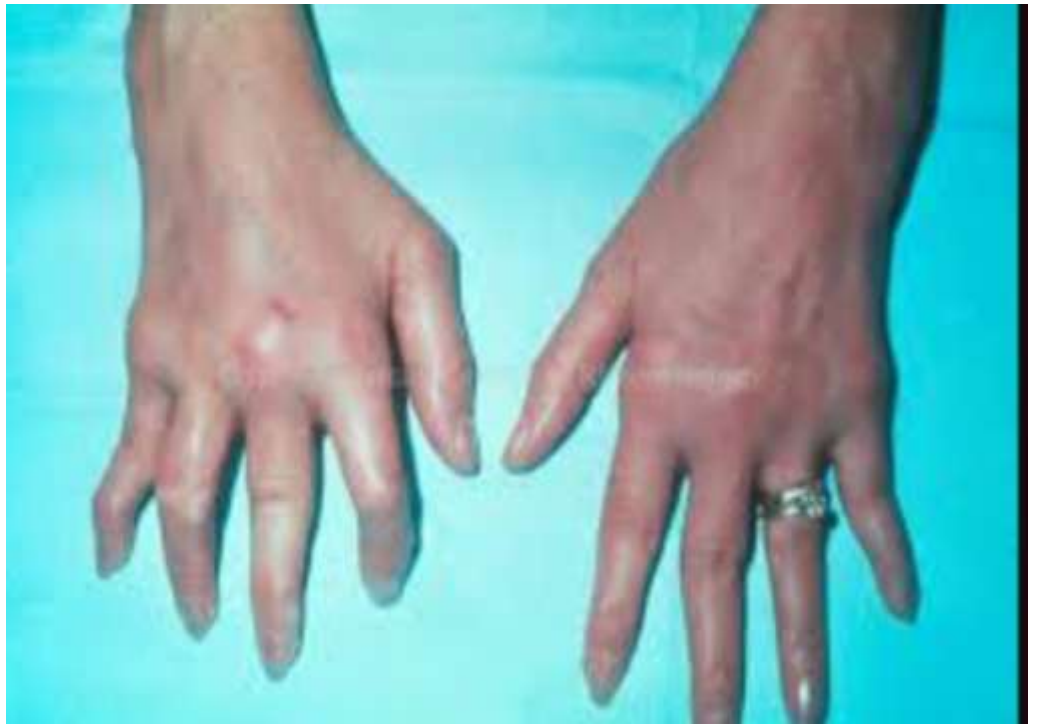


FIGURE 1: *Dermatitis herpetiformis (DH). Most patients with DH have concomitant intestinal mucosal changes of celiac disease on biopsy even in the absence of gastrointestinal symptoms. Both the rash and intestinal changes improve on a gluten-free diet.*



Anaphylaxis