WELCOME
TO
GRAND ROUND
02 MARCH 2013

A 25-year-old male presented with fever with altered level of consciousness

Presented by
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Particulars of the Patient

Name	: Mr.Mostofa Kamal
Age	: 25 years
Sex	: Male
Religion	: Islam
Marital Status	: Married
Occupation	: Businessman
Address	:Ataikula, Pabna
Date of Admission	:11.02.2013
Date of Examination	:11.02.2013

Chief complaints

- Fever for 2 days
- Altered level of consciousness for 1 day

Case summary

Mr. Mostofa Kamal, 25- year- old young Muslim, married, son of non consanguinous parents, diabetic, normotensive, non smoker, hailing from Ataikula, Pabna was admitted under MU-III at ShSMCH with the complaints of fever with altered level of consciousness for 2 days.

Case summary contd….

Fever was high grade, continued, associated with headache, chills and rigor, slightly reduced by taking anti pyretic drugs,highest recorded temperature 104°F. Fever was not associated with vomiting, loose motion, night sweats, burning sensation during micturition, cough, joint pain or skin rash.

Case summary contd….

Patient also developed altered level of consciousness 1 day after fever which was not associated with convulsion, history of head injury, discharge from ear or nose, taking CNS depressive drugs or intake of raw date palm sap and partially eaten fruits.

He had no history of Tuberculosis and contact with TB patients and traveling to malaria endemic area.

He gave history of gradual loss of libido and erectile dysfunction and gradual darkening of whole body since last few years with significant weight loss about 11 kg within last 7 months.

Case summary contd….

He had no history of leg edema, paroxysmal nocturnal dyspnea, black tarry stool or vomiting out of blood.

On query his attendant gave history of a mass in left upper abdomen which was first noticed at the age of 1 year and then gradually increased over time.

Case summary contd….

At the age of 8 years he got admitted in RMCH with the complaints of severe weakness, pallor and massive swelling of abdomen causing severe mechanical discomfort and transfused more than 20 units of whole blood within 3 months.

Case summary contd….

But the condition didn’t improved after blood transfusion. Then splenectomy was done. He got Vaccination before surgery but no history of re vaccination or penicillin prophylaxis after surgery. He also lost post splenectomy follow up and remain uneventful for last 16 years.

Case summary contd….

7 months back he was admitted in a local hospital of pabna with the complaints of gradual loss of weight and severe weakness and diagnosed as a case of Diabetes mellitus with severe anaemia and treated by insulin and 3 units of whole blood transfusion.

Case summary contd….

Since then he was on irregular insulin therapy with irregular follow up without history of any hypoglycaemic or hyperglycaemic crisis. Then he consulted with BSMMU out patient department and diagnosed as a case of **Hb E trait by Hb Electrophoresis**.

Case summary contd….

He got treated with **oral Iron chelator Tab Deferasirox (1200mg)** for 3 months due to features of Iron overload.

Case summary contd….

- **Hb electrophoresis: 12/9/2012**

Index	Band	Rel. Area	Range (%)
1	Hb A	50.3%	96.5-100
2	Hb E	49.7%	

Comment: Haemoglobin E trait

Case summary contd….

Iron Profile at 12/09/2012

Total Iron: 189 µgm/dl (25-150 µgm/dl)

S. Ferritin: 1993.17 µgm/l (30-300 µgm/l)

Case summary contd….

Patient is married for 4 years without having any issue, he has 3 siblings,all are in good health without having similar disease.He was immunized as per EPI schedule with no history of allergy to any drugs or food.

Case summary contd….

On general examination, the patient was drowsy, disoriented,with irrelevant talking,has depressed nasal bridge with malar prominence. Moderately anaemic, mildly icteric,

moderately dehydrated,

Temp-103°F,BP-80/50mmHg,pulse-120b/min, resp rate-28 breaths/min.

Case summary contd….

There was generalized hyper pigmentation with multiple discrete hyper pigmented macule over face and reduced male pattern hair distribution.

Neurological examination revealed patient was drowsy, disoriented with irrelevant talking, GCS score-8/15

Case summary contd….

Cranial nerves examination as far possible was normal with normal fundus, except bilateral planter extensor motor system could not evaluated , all signs of meningeal irritation present, sensory system and cerebellar sign could not evaluated.

Case summary contd….

Abdominal examination revealed an oblique scar mark about 11 cm extending from left costal margin upto umbilicus, liver is enlarged 4 cm from right costal margin,in right midclavicular line with sharp boarder, smooth surface, firm in consistency, non-tender,without hepatic bruit.

Case summary contd….

Testes are bilaterally small and soft with no ascites.

Cardiovascular system & Respiratory system reveals no abnormality.

Clinical Diagnosis

Meningoencephalitis with Septic shock

with hereditary haemolytic anaemia with post splenectomy status with secondary haemochromatosis with DM with hypogonadism.

Differential Diagnosis

- **Diabetic keto acidosis**

  with hereditary haemolytic anaemia with post splenectomy status with secondary haemochromatosis with hypogonadism.
- **Viral encephalitis**

  with hereditary haemolytic anaemia with post splenectomy status with secondary haemochromatosis with DM with hypogonadism.
- **Cerebral malaria**

  with hereditary haemolytic anaemia with post splenectomy status with secondary haemochromatosis with DM with hypogonadism.

Investigation

Complete Blood Count:

Hb	7.0 gm/dl
ESR	50 mm in 1st hour
Total Count of WBC	14,800/cumm
Differential Count:	
N	80%
L	15%
M	03%
E	02%
Platelet	300,000/cumm

Investigation contd……

- Peripheral Blood Film

RBC	Shows anisochromia and anisocytosis, target cells, a few polychromatic cells.
WBC	Increase total count with a few atypical cells.
Platelets	Adequate in number

Comment: Suggestive of Leuco-erythroblastic blood picture.

Investigation contd……

- **Urine R/E**

Albumin	+
Sugar	+++
Ketone Body	+
Pus Cell	8-10/HPF
RBC	Nil

- **S. Creatinine**: 1.2 mg/dl (0.4 to 1.4 mg/dl)

Investigation contd……

- **S. Electrolyte**

$[Na^+]$	141 mEq/L
$[K^+]$	4.8 mEq/L

- **Random Blood sugar-** 35 mmol/L
- **HbA1C-** 8.4%

Investigation contd……

CSF study:

Physical Examination	
Appearance	Hazy
Sediment	Absent
Biological Test	
Protein	76 mg/dl
Glucose	70 mg/dl

Investigation contd……

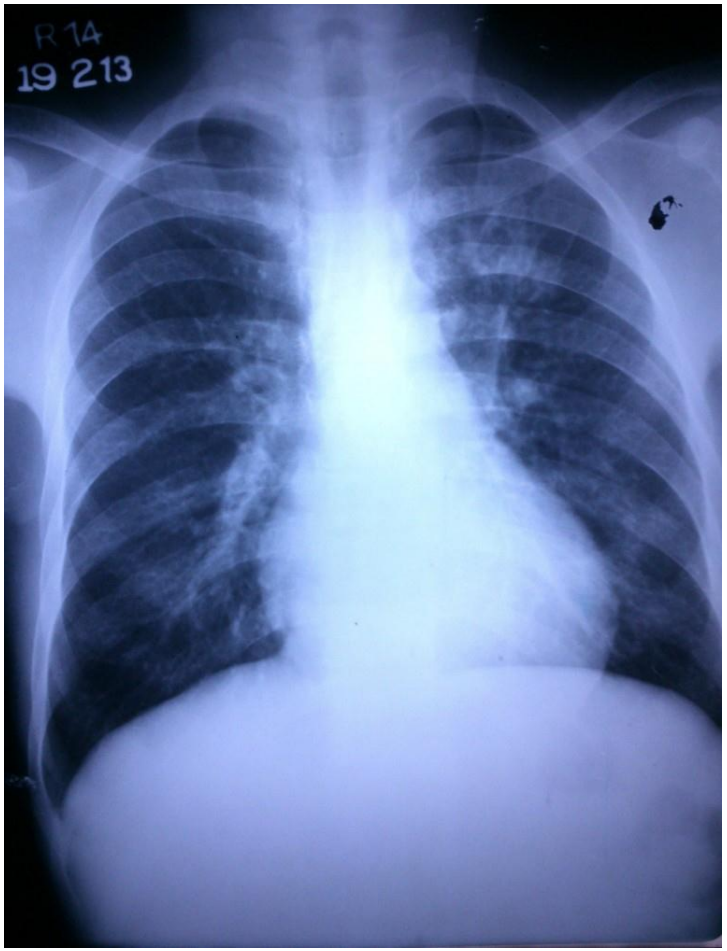
CSF study (contd.)

Microscopic Examination	
RBC	Nil
WBC	10-12/HPF
Total WBC	300/cumm
Neutrophil	80%
Lymphocyte	20%
AFB	Not Found
Gram Staining	Not found
CSF Culture	Yield no organism

Investigation contd……

ICT for Malaria	Negative
Blood for Malarial Parasite	Not found
Blood culture	No organism detected
X ray abdomen	Normal
ECG	Normal
ECHOCARDIOGRAM	Normal 2D/M-mode echo with good LV systolic function, LVEF-66%

Investigation contd……



Chest X ray- Cardiomegaly

Investigation contd……

USG of whole abdomen

Liver-Enlarged in size (16.7cm)with uniform echotexture

GB- Multiple bright echogenic structures casting posterior acoustic shadow are noted in GB Lumen.

Spleen – was not visualized (History of Spleenectomy)

Comments- 1.Hepatomegaly

2. Cholelithiasis

Investigation contd……

Serum bilirubin	3.0 mg/dl
SGPT	106U/L
SGOT	66 U/L
Total protein	5.8 gm/dl
Serum albumin	3.1 gm/dl
Serum globulin	2.7 gm/dl
A:G	1.14:1
HBsAg	Negative
Anti HCV	Negative

Investigation contd……

Investigation	Result	Normal value
S. TSH	12.7µIU/ml	0.4-4.0 IU/ml
Testosterone	29.5 ng/dl	270-1734ng/dl
FSH	0.84 mIU/ml	0.7-11.1 mIU/ml
LH	1.1 mIU/ml	0.8-7.6 mIU

Investigation contd……

Iron Profile At-20/02/2013

		Normal Value
Total Iron	**180 µgm/dl**	(25-150 µgm/dl)
S. Ferritin	**2635.24 µgm/l**	(30-300 µgm/l)
TIBC	**154 µgm/dl**	(250-425 µgm/dl)
TSAT	**117%**	[TSAT= (Total iron/TIBC)×100]

Confirm Diagnosis

Meningoencephalitis with septic shock with hereditary haemolytic anaemia (Hb E disease) with post splenectomy status with secondary haemochromatosis with DM with hypogonadism with subclinical hypothyroidism with cholelithiasis.

Management

Treatment on admission:

- Oxygen inhalation (high flow)
- Nasogastric feeding
- Inf. Normal saline
- Inj.Ceftriaxone (2gm) iv 12 hourly
- Inj. Vancomycin (1gm) iv 12 hourly
- Inj.Ranitidine (50 mg) iv 8 hourly
- Inj. Regular Insulin

  100 unit in 99 ml normal saline IV @ 6U/hour initially then titrated dose according to blood glucose level.

Plan of management

Patient education and counseling

- Patient should wear a bracelet informed about his post splenectomy status.
- Iron and iron containing diet are contraindicated.

Drug management

- Folic acid 5 mg daily.
- Tab penicillin V (500 mg) bd for life long.
- Iron chelation therapy.
- Vaccination
- Titrate Insulin dose to maintain DM.

Post splenectomy Vaccination

Streptococcus pneumoniae

Polyvalent pneumococcal vaccine (Pneumovax 23)

Haemophilus influenzae type B

Haemophilus influenzae b vaccine (HibTITER)

Neisseria meningitidis

- Age 16-55: Meningococcal (groups A, C, Y, W-135) polysaccharide vaccine
- Age >55: Meningococcal polysaccharide vaccine (A/C/Y/W-135)

Vaccine	**Revaccination**
Polyvalent pneumococcal	-Every 5 years
Quadravalent meningococcal polysaccharide	-Every 3-5 years
Haemophilus b	

Other management

- Blood transfusion to maintain the Hb level 10-12 gm/dl
- Hormone replacement therapy

Iron chelation therapy:

Inj. Deferoxamine (50 mg/kg/day) Intra venous or subcuteneous route

5 days in a week with oral vit C 1 Tab TDS.

OR

Tab Deferasirox (30 mg/kg/day) in a single dose every morning mixed with juice.

Follow up yaerly:

Target- Serum ferritin level < 1000 ng/ml

Thank you