

A 45-YEAR-OLD WOMAN WITH PERIORBITAL SWELLING AND ANEMIA

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Particulars of the patient

- Mrs X
- 45 years
- Housewife
- Gazipur Sadar, Gazipur
- Date of admission: 16-03-2016
- BSMMU Hospital

Presenting complaints

- Multiple nodular swelling around both eyes for 8 years
- Generalized weakness for 6 months
- Gum bleeding for the same duration

The course of illness...

- Periorbital nodular swelling for 8 years
 - Initially, single on right lower eyelid
 - Gradually increased in size and became multiple
 - Involved both eyes
 - Itching

The course of illness...

- No
 - Pain
 - Photophobia
 - Redness
 - Watering
 - Proptosis
 - Visual problem
- Similar lesion on upper back

The course of illness...

- Was admitted in NIOH on September 2009
 - Debulking surgery and biopsy
 - Antibiotic
 - Analgesic
 - Oral Prednisolone for short duration (14 days)

The course of illness...

- Histopathology report
 - Fibromuscular tissue
 - Collection of lymphoid cells and eosinophils
 - Formation of lymphoid follicles with prominent germinal center
 - Eosinophils are forming abscess in some areas
 - No granuloma or malignancy
 - **Diagnosis: Tissue from right eye (biopsy) – Inflammatory pseudotumor**

The course of illness...

Macroscopic Description

Specimen received in formalin with proper lab no and with patient's identification consists of six irregular grey brown pieces of tissue. The largest one measures about 2.5x1.0x1.0 cm. The cut surface is grey brown. Embedded four blocks.

Microscopic Description

Sections show fibromuscular tissue. Within the fibromuscular tissue, collection of lymphoid cells and eosinophils are seen. The lymphoid cells in most of the areas have attempted at formation of lymphoid follicles with prominent germinal center. The eosinophils are seen forming almost abscess in some areas.

No granuloma or malignancy is seen.

Dx

Tissue from eye right (biopsy) : Inflammatory pseudotumour.

The course of illness...

- Readmitted in NIOH on November 2009
 - Debulking surgery and biopsy
 - Antibiotic
 - Analgesic
 - Oral prednisolone for short duration

The course of illness...

- Histopathology report
 - Benign lesion
 - Fibroblast, collagen fibers, neurites
 - No malignancy was seen
- Diagnosis: Eyelid tissue (biopsy) - Neurofibroma

Macroscopic Description

Specimen received in formalin with proper lab no consists of an irregular grey brown measuring about 0.7x0.5x0.4 cm. Embedded whole in one block.

Microscopic Description

Sections show a benign lesion made of fibroblasts, collagen fibres and neurites.

No malignancy is seen.

Diagnosis

Dx

Eye lid tissue (biopsy) : Neurofibroma.

The course of illness...

- Not improved
- Repeatedly got herself admitted
- Given same treatment

The course of illness...

- Histopathology report
 - Neurogenic tumor with marked atypia
 - Sarcomatous change
 - Diagnosis: Eyelid tissue lower left (biopsy) – Malignant peripheral nerve sheath tumor

Macroscopic Description

Specimen received in formalin with proper lab no consists of an gray brown piece of tissue meas about 3.5x1.5x1.5 cm. The cut surface is partly gray white and gray brown. Embedded two blocks

Microscopic Description

Sections show fragmented pieces of tissue. It shows a neurogenic tumour with marked atypia. central area. it shows sarcomatous change.

Diagnosis

Dx

Eye lid tissue lower left (biopsy) : Features are suggestive of malignant peripheral nerve sheath tu

The course of illness...

- **March 2016**
- Gum bleeding for last 6 months
 - Associated with bleeding from nose
 - No H/O
 - Bleeding from other sites
 - Purpura
 - Hematemesis, Melena
 - Hemoptysis
- Generalised weakness
- Significant weight loss

The course of illness...

- No

- Fever, night sweats, anorexia
- Arthralgia
- Headache, nausea, vomiting
- Loose stool
- Heat or cold intolerance
- Food allergy
- Blood transfusion

Past history

- 1991: Obstructed labor with Eclampsia
 - Delivered a dead fetus
 - Complete perineal tear
 - Retained placenta
 - PPH and
 - Puerperal sepsis
 - D&C
 - Amenorrhoeic since then having no issue

Past history...

- Asherman syndrome
- Treated with different medications
- No improvement

Family History:

- Elder sister suffering from
 - Jaundice 5 years – cause?
 - Repeated blood transfusion 1 year
- Consanguinity of marriage between her parents

GENERAL EXAMINATION

General examination

- Anemia: Moderate
- Generalised pigmentation
- Vital parameters normal
- Poor oral hygiene and bleeding spot on gum



General examination ...

NO

- Jaundice
- Koilonychia
- Edema
- Lymphadenopathy
- Thyromegaly
- Bony tenderness
- Gum hypertrophy

SYSTEMIC EXAMINATION

Visual system...

- Multiple nodular skin lesion
 - Around both eyes
 - Blackish
 - Variable size, some have coalesced
 - Firm consistency



Visual system...

- Not
 - Tender
 - Fixed
 - Ulcerated
- No proptosis, lid lag
- Conjunctiva, cornea, sclera: Normal

Visual system...

- Pupils: Normal
- Visual acuity, color vision, field of vision: Normal
- Eye movement: Normal
- Fundoscopy: Normal

Abdomen

- Hepatomegaly
 - 4 cm from right costal margin along MCL
 - Firm
 - Non-tender
 - Smooth surface
 - Sharp margin
- Just palpable spleen

Abdomen...

- No abdominal lymphadenopathy
- No ascites

Other systems

- Unremarkable

DIFFERENTIAL DIAGNOSIS



INVESTIGATIONS

CBC

Hb%	8.7 mg/dl
ESR	45mm / 1st hour
RBC	2.88 mil/ cumm
WBC	10,000/cmm (N-71%, L-22%,M-05%, E- 02%)
Platelet	1,50,000/cmm
MCV	97.0 fl
MCH	30.0 pg
MCHC	31.0 g/dl
RDW(CV)	14%

Peripheral Blood Film

- **RBC:**

- Anisocytosis, anisochromia
- Dimorphic, target cell
- Spherocyte, micro spherocyte
- Polychromasia
- Rouleaux formation

- **WBC:** Mature with normal count

- **Platelet:** Normal

- **Comments:** Spherocytic anemia

Investigations...

- LDH: 242 U/L (*Increased*)
- Reticulocyte count: 6.42% (*Increased*)
- Coombs test (direct& indirect):
Negative
- Osmotic Fragility test: Normal

Osmotic Fragility test

AGE	:	45 yrs	Sex	:	F
OF THE TEST : OSMOTIC FRAGILITY TEST					
PERFORMED BY :					
OSMOTIC FRAGILITY TEST					
Initial lysis	:	5.5 g/l Nacl	Normal Range : 4.0 -4.5 g/l Nacl		
Complete lysis	:	3.0 g/l Nacl			
MCF (50% lysis)	:	4.25 g/l Nacl			
Comments	:	Normal Osmotic Fragility Test.			

LFT

- S. Bilirubin: 1.3 mg/dl
- SGPT : 62U/L
- SGOT: 92 U/L
- Total protein: 84 gm/l
- Albumin: 38 gm/l
- A/G Ratio: 1.83:1

Couagulation Profile

- BT : Normal
- CT: Normal
- PT: 14.3 s (INR-1.2)
- APTT: 28 s (normal)

USG of Whole Abdomen

- Liver enlarged
- Splenomegaly
- Cholelithiasis

Viral Markers

- HBsAg: Negative
- Anti HBc total: Negative
- Anti- HCV: Negative

Investigations...

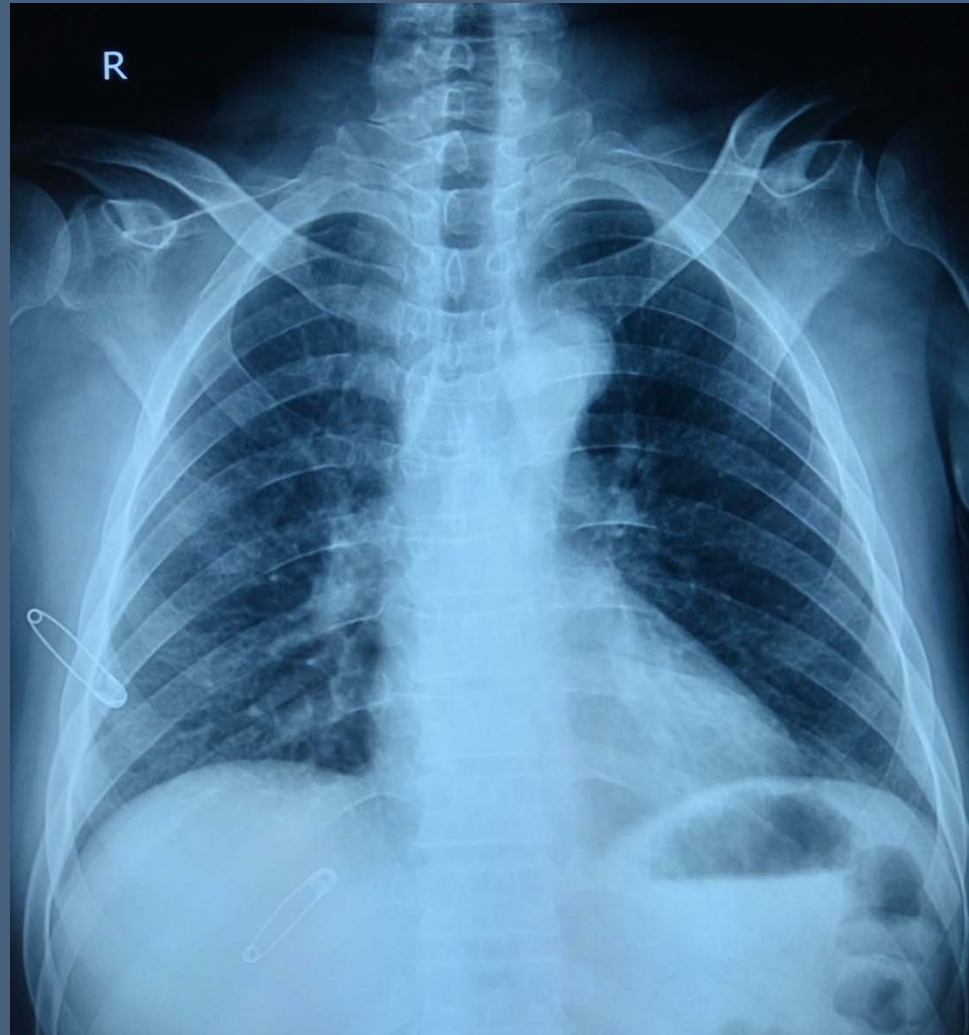
- Hb electrophoresis: Normal study
- Bone marrow examination: Reactive marrow
- ANA: Negative

Blood Biochemistry

- Fasting lipid profile
 - Total cholesterol: 217 mg/dl
 - HDL: 33 mg/dl
 - LDL: 148 mg/dl
 - TG: 181 mg/dl
- S. Creatinine: 0.7 mg/dl
- S. Electrolytes: Normal

Investigations...

- Chest X-ray P/A view: Mild cardiomegaly
- ECG: Normal study
- Echo-cardiogram: Normal study



Investigations...

- TSH: 7.95 mIU/L
- F-T4: 12.43 pmol/L
- Urine RME
 - Normal study



Histopathology

- Microscopic description:
 - Fibrocollagenous tissue
 - Infiltrated with chronic inflammatory cells
 - Foamy histiocyte
 - Multinucleated giant cells
- Diagnosis: Tissue from upper lid (rt):
Xanthogranulomatous reaction

Histopathology...

Gross Description

Specimen consists of four pieces of tissue largest one measuring about 2.0x0.8x0.60 cm.

Embedded one block. .

Microscopic description:

Sections show fibrocollagenous tissue infiltrated with chronic inflammation including foamy histiocytes and multinucleated giant cells.

No malignancy is seen.

DX: Tissue from upper eye lid, rt(Biopsy) : Xanthogranulomatous reaction

Investigations...

- S-100 protein: Negative

Final diagnosis



Final diagnosis

- Adult onset xanthogranulomatous disease with systemic involvement



Thank you!

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