



The Royal Free Hospital
HAEMOPHILIA CENTRE & HAEMOSTASIS UNIT

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Telephone
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GRO-C

GRO-B

26 October 1987

Dr GRO-B

GRO-B

Dear Dr GRO-B

GRO-B 51

GRO-B

Diagnosis: Severe Haemophilia A
Human Immunodeficiency Virus antibody positive

I saw GRO-B at the Haemophilia Centre on 14 October for a review. Generally, he is very well, he continues to live at home with his parents. His current drug therapy is Indomethacin 1 suppository at night and a 25 mg tablet taken at mid-day which seems to control the pain he was having in his left ankle. He also told me that he has intermittent back pain particularly when he has to stand up for long periods and tends to tire easily and often takes a little sleep in the afternoon or early evening. The only other problem is that he has lost a significant amount of weight (10 kgs) over the last 10 years.

On examination he looked well, his weight was 67.2 kg, his mouth was clean but there was ulceration on his inner lower lip. There was no palpable lymphadenopathy. Examination of the chest was clear, pulse was 100 per minute and regular. Blood pressure 120/90, and examination of the abdomen was normal.

Investigations showed a normal blood count

Haemoglobin 15.2 g/dl

White cell count $5.4 \times 10^9/l$

Platelets $160 \times 10^9/l$

Liver function tests showed elevation of aspartate transaminase and gamma GT consistent with his chronic non-A, non-B hepatitis.

The immunology showed a remarkably reduced level of the T helper count (T4) at $0.19 \times 10^9/l$ (normal lower limit 0.6).

In conclusion GRO-B is generally well at the present time but he has lost a significant amount of weight over the last 2 years and this may be related to HIV infection. In addition there is laboratory evidence of immunosuppression and any infection needs to be treated seriously. I have arranged to see him again in 3 month's time when he will also be seen by the dental surgeon at this hospital.

Yours sincerely

GRO-C

Elizabeth Miller, MRCP MRCPATH, Hon Lecturer

R.F.H. DEPARTMENT OF HISTOPATHOLOGY
P.M. REPORT

PHYSICIAN OR SURGEON Dr Kernoff

WARD

GRO-B

P.M No GRO-B

Hospital No.

GRO-B

Surname

First Name

GRO-B

M/F
M/S/W

D. of B.

GRO-B-51

M

Date of Admission: 22-Dec-87

Time of Admission:

Date of Death: GRO-B-87
00:35 hrs

Time of Death:

Date of Postmortem: GRO-B-87

Time of Postmortem: 12:00 hrs

Prosecutor:

Dr J E McLaughlin

Student:

PROVISIONAL ANATOMICAL DIAGNOSIS

CAUSE OF DEATH

- 1(a) Pneumocystis pneumonia
- 1(b) HIV infection
- 2 Haemophilia A

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PATHOLOGIST

Autopsy Number
Patient
Hospital Number

GRO-B

GRO-B

CLINICAL SUMMARY

The notes were available at the time of postmortem.

The brief clinical history is that the patient was diagnosed at the age of 16 months as a severe haemophilia A sufferer. His early problems were related to haemarthrosis following episodes of trauma and more recently he was diagnosed as having eczema. For the last year he has been known to be HIV sero positive and has a low grade chronic hepatitis. He was admitted on the 23 December 1987 with a two and half week history of cough which he attributed to upper respiratory tract infection and over the previous five days increasing shortness of breath. Chest X-Ray showed bilateral interstitial shadowing and a presumptive clinical diagnosis of pneumocystis pneumonia was made. He was treated with high dose septrin but died of respiratory failure six days after admission.

EXTERNAL APPEARANCES

The body was that of normally nourished white caucasian male (height 185cm, weight 98kg) who looked the stated age. There were flexion deformities of the knee joints but no other major external abnormality was noted.

INTERNAL APPEARANCES

Examination was limited to inspection of major organs.

Cardiovascular System

The pericardial surfaces were smooth and no fluid was present in the pericardial cavity. The heart appeared externally unremarkable.

Respiratory System

A little straw coloured fluid was present in both pleural cavities particularly the right but the pleural surfaces were smooth. Both lungs appeared externally anatomically normal but were extremely heavy.

Alimentary System

The peritoneal surfaces were smooth and no fluid was present in the peritoneal cavity. The liver was of normal size but showed a fine white stippling of the capsular and the cut surface. The intestines appeared externally unremarkable.

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Uro-genital System

The left kidney was removed and showed no external abnormality.

Lymphoreticular System

No enlarged lymph nodes were noted. The spleen was enlarged to approximately three times the normal size but the capsular surface appeared unremarkable.

Endocrine System

The left adrenal was unremarkable.

Musculo-skeletal System

No abnormality was seen in any bone, joint or muscle apart from the flexion deformities of the knees consistent with the history of haemophilia and haemarthrosis.

Central Nervous System

Meninges and external surface of the brain were unremarkable.

Summary

Limited postmortem examination on this patient with haemophilia A and HIV positivity shows splenomegaly consistent with the diagnosis of haemophilia and bilateral consolidation of both lungs consistent with pneumocystis pneumonia. Further investigation will be performed on the the fixed organs.

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