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# DEPARTMENT OF NEUROPATHOLOGY AUTOPSY REPORT

CH 334505 Page:

Autopsy No.: NPPM 55/2003

Name: MITCHELL, Conor

Date of Birth: 12/10/87

Sex: F

Hospital No.: CH 334505 CAH B63929

Hospital: RBHSC

Ward: PICU

Pathologist: Dr B Herron

cian: Dr Rothwell

Clinician: Dr Bothwell Date of Autopsy: 13/5/03

Date of Admission: (RVH) 9/5/03

Date of Death: 12/5/03 Time of Death: 3.45 pm

Time of Autopsy: 2.30 pm Restrictions: Coroner's Case

Organs/Tissue Samples Retained: Brain and spinal cord.

# ANATOMICAL SUMMARY

## SNOMED 3 CODES

Acute cerebral oedema with coning, patchy bronchopneumonia, no evidence of meningitis or encephalitis.

History of cerebral palsy, bilateral porencephalic cysts.

Bu 2/3/4

P3-42000 T-A0100 M-36300

## CLINICAL SUMMARY

He presented with a 10 day history of general malaise with a reduced oral intake and vomiting. It was presumed he had a viral illness and he was admitted to Craigavon Area Hospital. He was commenced on Penicillin for an ear or throat infection and improved and then started vomiting. Vomiting settled 3 days before admission (admission was on 8/5/03). He was able to keep down some food and fluids. His mother noticed sediment in his urine 2 days before admission and it was strong smelling. He had spasms which had increased over the previous week but no cough or sputum and he had occasional absence seizures (he was on Epilim). He lived with his mother and grandmother.

On examination he was drowsy and pale. His temperature was 36.7 with oxygen saturation of 97 and blood pressure of 118/69. His chest was clear on examination. His pulse was 72 and his abdomen was soft and non tender.

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The impression was that he had a urinary tract infection. His haemoglobin was 13.6 and his white cell count was 19.1 with a sodium of 138, urea of 7.8 and a creatinine of 5 on 8/5/03. A note on 8/5/03 said that he had 3 seizures since Christmas which were petit mal and usually when tired. He stared then sighed. He vomitted about 10 days before admission, then developed sore ears and throat and a temperature. It was queried whether he had a rigor and was started on penicillin which was changed to Amoxicillin. He was given Cyclizine for nausea.

At 6.30 pm on 8/5/03 a rash was noted on his abdomen but on examination there was no rash seen by the medical staff and there was no photophobia. On examination he was dry with a pulse of 96 and a blood pressure of 118 over 68. His chest was clear. The impression was that he was dehydrated with a urinary tract infection and a viral illness. The urine showed protein, blood and ketones. A seizure followed lasting seconds with no respiratory effort being made. He was bagged and masked. A Consultant Paediatrician was present and an anaesthetist called. His pupils were dilated and unreponsive to light. His pulse was 130 with a pressure of 112/78. He was given iv Phenytoin; Acyclovir was added and radiology was contacted to arrange urgent CT. There are no signs of intracranial iritation. The CT brain provisional report said that the scan was very abnormal with a large left sided porencephalic cyst but there was also high signals suggesting subarachnoid haemorrhage with tight basal cisterns. Intensive care was contacted at 2100 hours. The patient then had a coma scale of 3 with a pressure of 84/40. On arrival at Intensive care the pulse was 80 with blood pressure 84/48 and coma scale of 3 as mentioned above. The case was discussed with Mr Cooke in Neurosurgery and there was no indication for neurosurgical intervention.

On 9/5/03 at 10.05 am the pupils were dilated with no reaction to light and no independent breathing with a coma scale of 3 and a pulse of 145. There were no cranial reflexes. He was transferred to the Royal Victoria Hospital on 9/5/03. A repeat CT scan showed diffuse brain swelling including midbrain and brain stem with loss of grey/white matter differentiation suggestive of infarction although the cause wasn't apparent but ischaemic changes would give this appearance. He did not improve and died on 12/5/03.

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# **EXTERNAL EXAMINATION**

The body is that of a child weighing 24 g and measuring 1.3 m. His head circumference is 57 cm. External examination is unremarkable apart from slightly prominent upper teeth and mild joint contracture. He is otherwise quite thin and has the appearance of someone younger than his 15 years.

# INTERNAL EXAMINATION

#### **BODY CAVITIES:**

There is no pleural or pericardial effusion and there is no ascites.

## HAEMATOPOIETIC SYSTEM:

Spleen The spleen weighs 66 g.

Histologically this shows some reactive changes with increased neutrophil polymorphs.

Lymph Nodes There is no lymphadenopathy.

Histologically they show no abnormality.

<u>Bone Marrow</u> Histologically this shows blood cells in normal haemic proportions.

# MUSCULO-SKELETAL SYSTEM:

There is no asymmetrical muscle wasting. There is no obvious bony abnormality.

Histologically a section of muscle shows no atrophy.

#### RESPIRATORY SYSTEM:

<u>Lungs</u> The right lung weighs 320 g and the left lung 280 g. Both show oedema and pneumonia but not a lot of haemorrhage to suggest aspiration.

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Histologically bilateral oedema is present but there is no haemorrhage within the alveoli to suggest aspiration. There is however patchy bronchopneumonia.

# CARDIOVASCULAR SYSTEM:

Heart The heart weighs 166 g. It is normally formed with normal coronary arteries, artria, ventricles and valves. The rest of the vascular system is normal.

Histologically the myofibres show increased eosinophilia and contraction but no myocarditis. There is no established ischaemic necrosis.

### DIGESTIVE SYSTEM:

Oesophagus There is no lesion in the oesophagus.

Histologically there is no abnormality.

Stomach The stomach is slightly congested.

Histologically there is no abnormality.

<u>Intestines</u> The small and large intestine appear normal.

Histologically there is no abnormality (although autolysed).

<u>Liver</u> The liver weighs 716 g and appears normal.

Histologically this is congested and shows increased neutrophil polymorphs sinusoids in keeping with a septic state.

Gall Bladder The gall bladder is normal.

Pancreas The pancreas is normal and weighs 55 g.

Histologically there is no abnormality.

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## GENITO-URINARY SYSTEM:

<u>Kidneys</u> The right kidney weighs 95 g and left 100 g. The collecting system is normal.

Histologically there is no abnormality.

Bladder The bladder shows focal congestion (catheterization).

## ENDOCRINE SYSTEM:

<u>Thyroid</u> The thyroid is symmetrical and not enlarged.

Adrenals The adrenals weigh 11.5 g in total.

Histologically there is no abnormality.

Pituitary Histologically this is necrotic.

Parathyroids There is no parathyroid enlargement.

## **NERVOUS SYSTEM:**

<u>Unfixed Brain</u> The brain was oedematous, showed focal subarachnoid haemorrhage but no subdural or extradural haemorrhage.

#### **BRAIN DESCRIPTION**

<u>Following fixation</u> On examination of the dura there is some yellowish discolouration which lines the whole inside surface of the dura.

The brain weighs 1148 g. There is no extradural or subdural haematoma and there is no meningitis. There is no sinus thrombosis. On examination of the fixed brain the most remarkable abnormality is the left sided large cystic area in keeping with a porencephalic cyst. This extends from the left frontal area close to the frontal pole into the left parietal area. It extends into the left sylvian fissure.

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In addition to this the brain is extremely congested but there is no purulent exudate. A small focus of subarachnoid haemorrhage is present in the right mid frontal area. On further examination of the right side a smaller porencephalic cyst is present. This has similar anterior boundaries to the left sided one but does not extend posteriorly to the same degree. On examination of the base of the brain the arteries appear normal. The unci are markedly swollen and the tonsils are necrotic.

On examination of the coronal sections the presence of bilateral porencephalic cysts is confirmed. Otherwise the brain is extremely congested with very prominent vascular markings bilaterally including grey and white matter. No tumour is present.

## HISTOLOGY

<u>Dura</u> There is no significant inflammation or haemorrhage. However focal haemosiderin staining is present.

<u>Hypothalamus</u> There are similar changes to the rest of the cerebral hemispheres with tissue necrosis, a patchy neutrophilic infiltrate in response to the tissue necrosis. Occasional vessels show a very thin cuff of lymphocytes.

<u>Right Hippocampus</u> There is acute hypoxic ischaemic necrosis with a scattering of neutrophils in response to the tissue necrosis. The dentate fasciculus shows no duplication.

Midbrain There is no established acute meningitis or lymphocytic inflammation. The vessels and meninges do show peripheral neutrophilia with neutrophils in the lumen of the vessels and also attached to the endothelial surface. This also occurs in the vessels within the midbrain. There is compression of the midbrain by the adjacent medial temporal structures. All of these are necrotic. The medial temporal structures (uncus) shows neuronal eosinophilia and nuclear pyknosis. Many of the neurones also within the midbrain show similar features. The aqueduct appears compressed but there are no polymorphs within the aqueduct. No abnormal inclusions are seen in the midbrain.

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<u>Pons</u> Similar changes are at present in the pons (as the midbrain). In addition secondary brain stem haemorrhage is present. There is no central pontine myelinolysis. There is extensive oedema in the periaqueductal region with fluid in the surrounding tissues. In the meninges surrounding the brain stem fragments of cerebellum are present in keeping with tonsillar necrosis.

<u>Left Basal Ganglia</u> There is oedema with neuronal necrosis and perivascular haemorrhage suggestive of perfusion following a period of tissue necrosis (similar to post arrest).

Right Basal Ganglia Similar findings to left basal ganglia.

<u>Left Frontal</u> For the most part the cortical lamination is normal. There is some increase in white matter neurones but the main abnormality is the presence of diffuse cortical haemorrhage similar to that seen in the basal ganglia with marked oedema, hypoxic ischaemic necrosis and reperfusion haemorrhage. In areas the meningeal blood vessels are quite prominent but don't show the typical features of an arteriovenous malformation.

<u>Right Frontal</u> For the most part the cortical lamination is normal. There is some increase in white matter neurones but the main abnormality is the presence of diffuse cortical haemorrhage similar to that seen in the basal ganglia with marked oedema, hypoxic ischaemic necrosis and reperfusion haemorrhage.

<u>Left Porencephalic Cyst</u> Tissue not related to the cyst appears to have good lamination. That closer to the cyst wall appears to be more complex with architectural disturbances in the pattern of migration.

<u>Cerebellum</u> There is dentate necrosis although the Purkinje cells are relatively well preserved. Tonsillar necrosis is confirmed histologically.

<u>Spinal Cord</u> No intrinsic abnormality is present but tonsillar tissue is present over the surface of the cord.

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## COMMENT:

To summarize the brain pathology. He had a history of cerebral palsy and epilepsy. His brain showed bilateral porencephalic cysts. In addition there were acute changes to this brain with coning of cerebellar tonsils, cerebral oedema, cerebral perfusion failure, global tissue necrosis with an early inflammatory response. There was haemorrhage into the brain which may be secondary to reperfusion following a respiratory or cardiac arrest due to leakage of damage vessels.

19/6/03/hb 4/12/03/hb 26/2/04/hb

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