

History of Case Submitted by K. M. Earle, M.D.
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This 15 year old boy died and an autopsy was performed. The brain was sent to me.

Records obtained from the hospital indicate that his birth was full term and uncomplicated. He developed normally for the first ten months at which time convulsions began and he was diagnosed as having "cerebral palsy" and "epilepsy". He was one of four children. One sister has epilepsy, but the other children and the parents are living and well. The exact nature of these convulsions and exact medications are not known. History was difficult to obtain from informants.

The boy was admitted to a hospital a first time but the family took him home against advice and all that is known is that he was having convulsions at the time. He was readmitted two months later with "convulsions and fever". An informant stated that he had been unable to feed himself since the first admission and had been fed by a nasal tube. Shortness of breath, episodic coughing and loss of sphincter control had been present since.

On admission to hospital he was described as a slender boy who was small for his age. He was comatose and respirations were labored. His neck was "stiff" and the pupils did not react to light or accommodation. Coarse rales were present in all lung fields. The extremities showed increased tone. Reflexes were hyperactive but there were no pathological reflexes.

Laboratory studies showed: RBC 4,000,000; Hgb. 11.5 gms.; WBC 18,450, Stabs 1, segs 76, neutro. 77, lymph 14, mono 1, eso. 8.

Urine showed 20-30 pus cells per h.p.f.; 6-8 rbc, sp. gr. 1.000; alb. and sugar negative.

V.D.R.L. negative.

X-Rays of the chest showed evidence of pulmonary edema. Despite therapy respiratory difficulty worsened and he died on the third hospital day. Spinal fluid examination is not recorded.

Autopsy revealed "multiple, pearly white, shiny nodules on the face, varying from 0.1 to 0.4 cm., which are well circumscribed, firm and distributed over the nose, cheeks and eyelids". Areas of brownish pigmentation and several large pigmented nevi were scattered over the abdomen and trunk. The kidneys contained well circumscribed, firm, round and oval nodules of light yellow and grey tumor masses in the cortex and medulla. The tumors varied from 0.2 to 4 cm. in diameter. The bladder showed evidence of acute cystitis and the lungs showed pulmonary edema and thickening of alveolar walls. The brain weighed 1165 gms. and showed greyish white nodules in the cortex and projecting into the ventricles. Some of these nodules were calcified.