

CASE 7

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This was a 63-year-old female who presented in with abdominal pain. She underwent a laparotomy and was found to have "retroperitoneal fibrosis". Three months later she was admitted with emaciation, anasarca and ascites. Over the next eleven months these signs progressed and she developed an insatiable thirst "•••she would sneak fluids everytime she got a chance•••". The thirst was attributed to psychogenic polydypsia and no endocrinological investigation was instituted. The patient died shortly thereafter. Prior to her death her serum electrolytes were normal, urine specific gravity was 1.006 and a skull X-ray showed no abnormality of the pituitary fossa.

General Pathology: The retroperitoneum and mediastinum were diffusely infiltrated by firm white tissue which surrounded the aorta, inferior vena cava, ureters, adrenals, pelvic organs and extended along penetrating vessels into the hepatic and pulmonary parenchyma. The thyroid was normal.

Neuropathology: The pituitary gland measured 1 x 1 x 1.5 cm. The pars posterior was replaced by a uniform, firm, white mass. The brain was normal.

Sections submitted:

- A. horizontal section of the pituitary gland, H&E and unstained
- B. mediastinal infiltrate, H&E

Points for discussion:

1. What is the relationship of the lesion in the pars posterior to Hnad-schuller-Christian disease, Letterer-Siwe disease and other forms of histiocytosis?
2. How often and to what extent is the central nervous system involved in the above condition?
3. What is the relation of the pituitary lesion to the retroperitoneal lesion which we believe to represent the retroperitoneal xanthogranuloma of Oberling?
4. Are cases 6 and 7 variants of the same disease process?
5. What is the etiology of this condition?