CASE #2

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Cushing's syndrome was diagnosed in a 36-year-old man following which 3,000 rads were applied to the pituitary gland without relief of symptoms. One year later, he underwent subtotal adrenalectomy (for adrenal hyperplasia). Two years later he was begun on Orinase because of a borderline abnormal glucose tolerance curve. At age 43, the remnant of adrenal gland was removed because of persistent hyperadrenalism. (Gland showed diffuse hypertrophy and hyperplasia.) Steroid replacement was given. His subsequent course was as follows:

At age 47 - Increasing skin pigmentation; severe headaches; X-ray - ballooned sella. Later mild adrenal insufficiency, headaches, nausea and vomiting.

At age 49 - Pyrohypophysectomy done with post-operative bilateral 6th nerve palsies. Chromophobe adenoma diagnosed.

From age 50-52 - Repeated readmissions because of visual problems and Herpes Zoster infection of left eye. Additional craniostomies were done and 7,500 rads applied to head with some improvement of eye signs.

Final admission, one year later low back pain with bilateral generalized motor weakness without focal findings except for absent right knee jerk. Myelogram showed block at T_{12} and filling defects at L_{2-4}. Decompression and radiotherapy were done for intradural, extramedullary tumor. He died one week after the surgery.

Three Kodachromes are submitted as follows:

Figure 1: Base of brain, tumor on optic nerve and temporal lobe tip.

Figure 2: Tumor in spinal cord.

Figure 3: Microscopic photo, tumor in liver.

One microscopic slide stained with H & E containing tumor involving spinal cord.

Points for discussion:

1. What is the diagnosis of this neoplasm?
2. What is the relationship between the neoplasm and the endocrine dysfunction?