Case 1

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Clinical Abstract:

At age 45, this woman had Grave's disease and was treated with methimazole. She developed progressive ophthalmopathy and an autonomous toxic nodule was treated with radioactive iodine. She, then, received synthetic thyroid hormone for hypothyroidism. At age 46, in an evaluation of a urinary tract infection, plain radiograms of the abdomen revealed calcifications. At laparotomy, there was retroperitoneal fibrosis, with compression of both ureters; bilateral nephrostomies were performed. She received corticosteroids for the ophthalmopathy and retroperitoneal fibrosis. Serum anti-parietal antibody, rheumatoid factor, and FANA were negative. At age 51, she was hospitalized with progressive uremia and had several episodes of cardiac arrhythmias. There was decreased sensation of the larynx and pharynx, with absent gag and cough reflexes. Direct laryngoscopy demonstrated mobile vocal cords. Tracheostomy was performed, nasogastric tube feedings were initiated, and hemodialysis begun. She had a seizure and remained unresponsive for 3 min., with her eyes rolling to the right then to the left. She awoke but later that day, after hemodialysis, she developed a right motor seizure. She could follow simple commands but did not visually follow an object. The eyes deviated downward, more on the right than on the left. There was loss of horizontal conjugate gaze. Pupils and fundi were unremarkable. There was a right central facial paresis and slight weakness of the legs. Bilateral Babinski signs were present.

A CT scan revealed a 5 cm diameter low density lesion adjacent to the left cerebello-pontine angle. CSF was normal. Dopamine was required to maintain blood pressure. Two days after the first seizures she became comatose, hypothermic and responsive only to noxious stimuli. There was disconjugate gaze with roving eye movements and recurrent seizures. The corneal reflexes were absent. The cold caloric test produced tonic deviation of the eyes. She died 7 weeks after she was hospitalized.

The general autopsy revealed generalized anasarca, biventricular cardiac hypertrophy, bilateral hydronephrosis secondary to retroperitoneal fibrosis. Diffuse fat necrosis with proliferation of fibroblasts and infiltration by lymphocytes and foamy histiocytes were present in the retroperitoneum, mediastinum and adventitia of the aorta. Amyloid angiopathy was found in these sites, in almost all viscera, dura and choroid plexus.

The dura was infiltrated by multiple soft, yellow, greasy, nodular or flat deposits adjacent to the superior sagittal sinus over a region 10x2 cm. A soft, solid, bosselated, tannish-yellow mass, measuring 6x4x2.5 cm. was firmly attached to the inferior surface of the left leaflet of the tentorium cerebelli and a similar smaller mass was present in the right leaflet. Their cut surfaces were tan-yellow, smooth and greasy. These masses indented the inferior surface of the left occipital lobe and the superior surface of both cerebellar hemispheres, especially on the left. There was a deep groove along the left tonsil.

Material submitted: One H&E stained section from the tentorial mass

Point for discussion: What is the relationship of the dural lesions to the patient's systemic disorder?