

Case 3

Submitted by: Dr. J.M. Bilbao, Department of Pathology  
St. Michael's Hospital, University of Toronto

Clinical Abstract:

While vacationing this 29-year-old man developed pain and weakness of right leg. He had experienced for the previous two years, dysesthesia in the same limb intermittently. He was transferred back home and hospitalized. CT scan revealed two space occupying masses extradurally at L4-5 on the right side and at T4 on the left side. The lumbar lesion had eroded into bone.

Medical History: He was normal up to the age of 14 except for cutaneous lesions which were resected and diagnosed as myxoma and blue nevi. He also had a papilloma removed from the mouth. At age 15 he had the onset of TIA's followed by emboli to his legs. Two years later he suffered a major cerebral vascular accident on the left side. An atrial myxoma was removed the same year. He remained well for nine years except for slight residual right hemiparesis. At this time his sister developed overt Cushing's syndrome. A family screening revealed that the patient had biochemical Cushing's syndrome as well. However, there was no evidence of diabetes or increased blood pressure. At age 26 he underwent a bilateral adrenalectomy and has continued since with replacement cortisone therapy. One year later, he was found to have a testicular tumor which was resected and diagnosed as Leydig's cell tumor.

Family history revealed the father to have a cutaneous myxoma a gastric myxoma, and biochemical Cushing's syndrome; and his sister had a breast myxoma, Cushing's syndrome, and cutaneous lesions.

Physical examination shows a pleasant and intelligent young man who was normotensive. There were no skin lesions and neurological assessment revealed a right hemiparesis and dysesthesia of right leg.

Laminectomies: (x 2) for resection extradural mass right L4-5 abutting on nerve root; and resection extradural mass left T4-5 were performed approximately 1 month apart.

Material submitted: One H&E stained section (either block 3A or 3B)

Points for discussion:

1. What is the differential diagnosis?
2. What other lab data would help in establishing correct diagnosis?
3. What can be said about the family history?