Dural ectasia accompanying a case of Multipl Endocrine Neoplasia Type 2B

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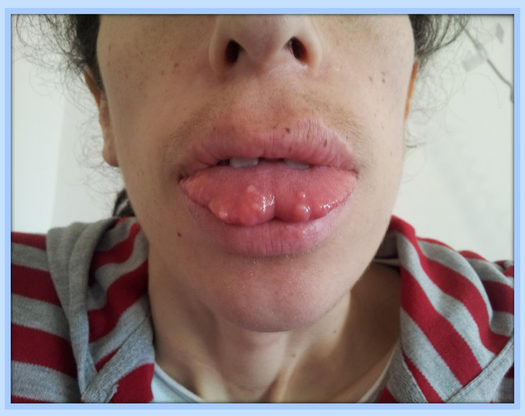
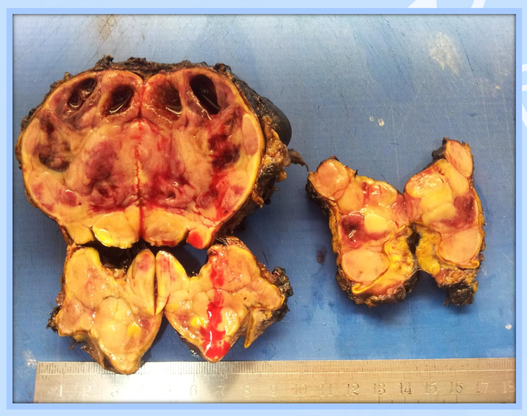
INTRODUCTION

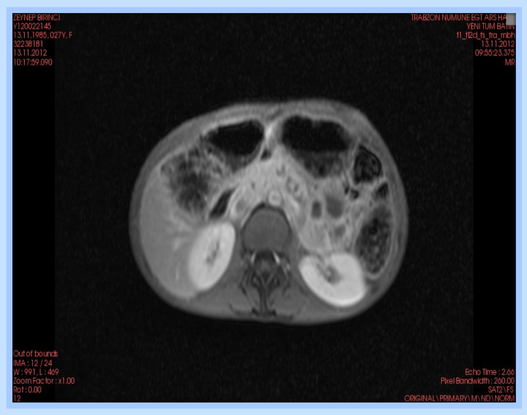
Manifestations of MEN2B include medullary carcinoma of thyroid (MCT), pheochromocytoma, and a number of somatic mutations like marfanoid habitus, mucosal neuromas, ganglioneuromatosis of the bowel. Dural ectasia results from enlargement of the spinal canal, was identified in 63-92% of patients with Marfan syndrome, however, it was not previously described in MEN2B. We detected dural ectasia in our patient with MEN2B.

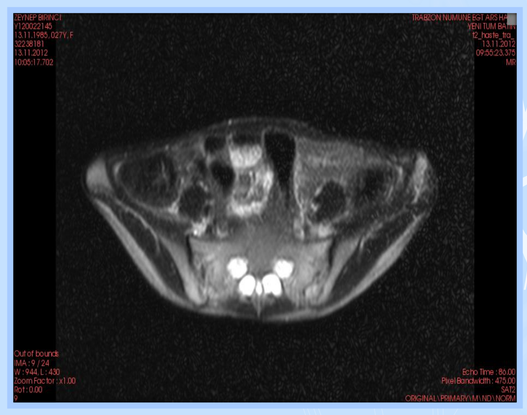
CASE REPORT

A 28 year-old woman was admitted with abdominal discomfort. Bilateral adrena masses were detected by ultrasonography. She had been operated for nodular disease and took a diagnosis of MCT before 13 years. At that time, she had not have pheochromocytoma and her adrenal glands were detected as normal. At physical examination, multiple neuromas on the lips and tongue, marfanoid facies were detected. A decreased upper/lower body ratio was present. Laboratory examination revealed elevated levels of urinary catecholamine metabolites (Normetanephrine:12789 µg/day, Metanephrine:9650 µg/day). Magnetic resonance imaging showed bilateral adrenal masses (40x18 mm left, 50x25 mm right) compatible with pheochromocytoma. Dural ectasia was detected incidentally at sacro-iliac region. Calcitonin level was detected to be normal, her neck ultrasonography was negative for recurrence of MCT. After bilateral adrenalectomy, she was started on hydrocortison and her urinary catecholamine metabolites were detected as normal.









DISCUSSION

Patients with MEN2B have development abnormalities, a decreased upper/lower body ratio, skeletal deformations, joint laxity, Marfanoid habitus, and myelinated corneal nerves. Disturbances of colonic function are common, including chronic constipation and megacolon. Dural ectasia can be seen in anchylosing spondylitis, achondroplasia, Loeys-Dietz syndrome and in the vascular form of Ehlers-Danlos syndrome besides Marfan syndrome. This abnormality results from enlargement of the spinal canal owing to progressive ectasia of dura and neural foramina and to erosion of vertebral bone, it involves lumbosacral spine. To our knowledge, this is the first case of coexistence of MEN2B and dural ectasia.