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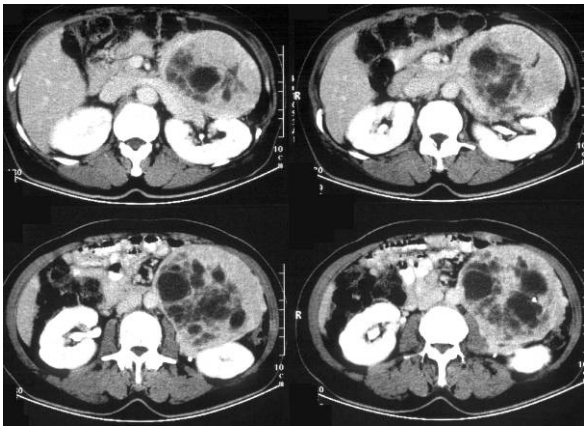
腹部巨大副神經節瘤（Paraganglioma)

曾秋德，曾譯諏

本案例爲一 48 歲女性，主訴左側腰部及左上腹部觸痛約有 1 個月，其它相關症狀方面，在左上腹部有摸到硬塊但體重沒有減輕，也沒有高血壓。追述其病史，病人於 1 個多月前健檢發現疑似胰臟腫瘤，在血液生化檢查方面大概都在正常範圍，CEA: 0.3ng/dl; CA19-9: 8.9ng/dl 也都在正常範圍內，腹部電腦斷層檢查在胰臟體部及尾部有一腫瘤(mixed solid and cystic)，R/O Carcinoma(如圖示)，於是幫病人安排手術治療，術中發現並非爲胰臟腫瘤而是一位於 celiac artery 附近的腹內大腫瘤(約 1130g)，病理報告爲一副神經節瘤，術後病人病情穩定，出院後於門診追蹤治療。



腹部電腦斷層掃描發現胰臟體部及尾部有一腫瘤。

討論

1. 副神經節瘤並不常見，發病尖峰期爲 30-60 歲。
2. 大部分副神經節瘤爲偶發的，且多爲單一性。
3. 副神經節瘤長於 Adrenal medullary 或其它 Chromaffin tissue 的腫瘤，90%發生於 medullary of Adrenal; 10% 發生於 aorticosympathetic paraganglia，organ of Zuckerkandl，carotid body，vagal body 及 jugulotympanic body。
4. 副神經節瘤誰然分界清楚，但是豐富的 blood supply 卻造成此腫瘤的切除困難。
5. 症狀: 因分泌過多的 catecholamine，因而引起頭痛，發汗，心悸，頻脈或高血壓。
6. 診斷: (a) 尿中血中的 catecholamine 值及尿中 vanilmandelic acid (VMA)，normetanephrine，metanephrine 值上升; (b)藥物檢查如 regitine test， histamine test; (c)放射線檢查如 sonography，CT，MRI 或 131I-MIBG。
7. 治療: 腫瘤切除.但術前需矯正血容積減少的病態，以防術中，術後休克.;對於不能切除的腫瘤，也應投於藥物控制血壓。
8. 病理: 顯微鏡下，可見副神經節瘤由聚成巢狀的神經內分泌細胞所組成，細胞巢則是富含血管的間質，神經內分泌細胞內尚可見到富含 catecholamine 的分泌性顆粒，可能導致 catecholamine 的過度分泌性。
10. 預後： 有 10-50%的副神經節瘤會在切除後復發，有 10%的副神經節瘤會轉移。

參考文獻

Abdominal and pelvic extra-adrenal paraganglioma: a review of literature and a report on 7 cases.
Ali-el-Dein B, el-Sobky E, el-Baz M, Shaaban AA.
Urology & Nephrology Center, Mansoura University, Mansoura, Egypt. balieldein@mum.mans.eun.eg
INTRODUCTION: Extra-adrenal paraganglioma (pheochromocytoma) is a rare tumor. Herein we describe the clinical and pathological findings in patients with paragangliomas of the urinary bladder, seminal vesicle and retroperitoneum. METHODS: Between January 1994 and January 2001, extra-adrenal paragangliomas were diagnosed in 7 patients: 3 males and 4 females. The mean age of our patients was 32 +/- 15.9 years. We reviewed the clinical data. Urinary metanephrines and vanillyl mandelic acid and blood catecholamine levels were estimated in 4 cases. CT scan and/or MRI were used in the imaging of all cases. 123I-MIBG was used in only 1 patient, who harbored multiple tumors. All the patients but one underwent surgical treatment. RESULTS: The definitive diagnosis was made by histopathological examination of the removed tumors and was confirmed in all cases by the immunohistochemical stains of chromogranin A and S100 protein. There was metastasis in the pelvic lymph nodes in 1 patient. Follow-up ranged from 3 to 82 months (mean = 37.9 +/- 25.8). The catecholamine level was elevated in 3 patients under basal conditions and during endoscopic resection of the tumor in a fourth patient. In all cases, the catecholamine level was normalized after surgery. There was no recurrence or metastasis in any case following surgery. CONCLUSION: Pre-operative diagnosis of nonfunctioning bladder paraganglioma is difficult, but the tumors should be suspected in patients who have hypertension, hematuria or mass effects due to the tumor growth in the pelvis and/or retroperitoneum. Six of the seven cases reported here were found in the usual locations: 3 in the urinary bladder, 2 in the renal hilum and 1 in the organ of Zuckerkandl. One patient had multiple tumors, including a paraganglioma of the seminal vesicles. Resection is the treatment of choice, and in the case of urinary bladder paraganglioma should include total cystectomy. In patients with unresectable multiple tumors, medical therapy may be used to control hypertension.
PMID: 12224134 [PubMed - in process]

Safe retroperitoneal endoscopic resection of pheochromocytomas.
Berends FJ, Harst EV, Giraudo G, Terkivatan T, Kazemier G, Bruining HA, De Herder WW, Bonjer HJ.
Department of Surgery, University Hospital Dijkzigt, Dr Molewaterplein 40, 3015 GD Rotterdam, The Netherlands.
Although endoscopic adrenalectomy is advocated for small adrenocortical tumors, questions remain about the safety of endoscopic retroperitoneal resection of pheochromocytomas. In this study we evaluated the outcome of retroperitoneal endoscopic adrenalectomy for pheochromocytoma. Between June 1995 and September 1999 we performed 18 retroperitoneal endoscopic adrenalectomies for a pheochromocytoma or paraganglioma. All patients received adequate alpha-adrenergic blockade. The adrenal vein was ligated at the end of the procedure. Operative blood pressure values were recorded and evaluated. Altogether 15 patients (11 women, 4 men; mean age 47.2 years) were operated on for 17 pheochromocytomas and 1 extraadrenal tumor (4 right, 11 left, 3 bilateral). One female patient was operated on at 13 weeks' gestation. Hypertensive episodes at operation were seen in 4 (26.7%) patients, and tachycardia occurred in 5 (33%). Hemodynamic changes could be corrected in all cases using simple measures without morbidity or detrimental effects. The mean operating time was 125 minutes (80-180 minutes), and the conversion rate was 5.6% (1/18). The median hospital stay was 5 days (3-28 days). Morbidity was 20% (3/15). Endoscopic retroperitoneal adrenalectomy for pheochromocytoma is safe and effective, and it is associated with limited morbidity.
PMID: 12098038 [PubMed - indexed for MEDLINE]

肝硬化併肝門靜脈瘤

陳明楨

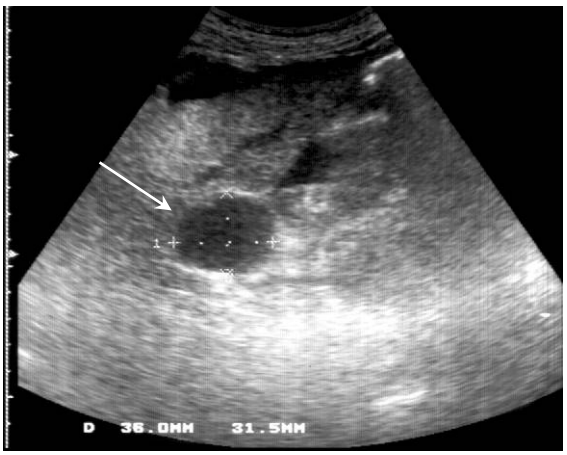
65 歲男性患者，因數週來體重減輕，全身倦怠且最近幾天有腹脹、茶色尿情形，故至本院就診。在此之前身體健康情形良好，否認有肝炎病史。

入院時理學檢查：身高 160 ㎝、體重 66kg、血壓 117/70 mmHg、呈慢性病容，有黃疸現象，腹部柔軟微脹，無肝脾種大，無下肢水腫。實驗室檢查：GOT：68 u/L、GPT：42 u/L、Alb：2.4 mg/dl、Bil(T/D)：0.72/4.79、HbsAg(+)、Anti-HCV(-)。

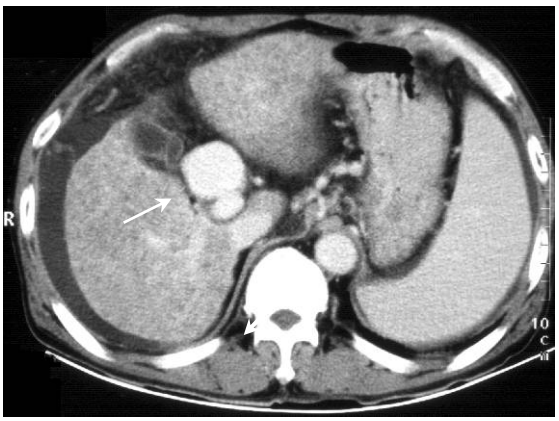
超音波檢查除有肝硬化併腹水外，在肝門附近有一 3.0 ㎝之低回音囊狀病灶。經電腦斷層攝影檢查，確定爲肝門靜脈瘤。

討論：

肝門靜脈瘤並不常見，可分爲肝內及肝外肝門靜脈瘤。大部份和慢性肝病及肝硬化有關。大多爲超音波檢查時意外發現且無症狀，腹部超音波、電腦斷層攝影、血管攝影檢查可作爲診斷工具。如果肝門靜脈瘤有變大或血栓形成可考慮外科治療。



腹部超音波檢查有肝硬化併腹水外，在肝門附近有一 3.0 ㎝之低回音囊狀病灶。



腹部電腦斷層攝影中確定爲肝門靜脈瘤。

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Portal vein aneurysm: report of six cases and review of the literature.

Ohnami Y, Ishida H, Konno K, Naganuma H, Hamashima Y, Zeniya A, Masamune O.

First Department of Internal Medicine, Akita University School of Medicine, Japan.

Portal vein aneurysm is very rare, and its relation to portal hypertension has been emphasized. We report six cases of portal vein aneurysm (five extrahepatic and one intrahepatic). All patients were asymptomatic and had no signs suggestive of portal hypertension; the lesion was incidentally detected by ultrasound. Color Doppler sonography showed a constant hepatopetal flow along the aneurysmal wall, which immediately led to the diagnosis. We stress the usefulness of color Doppler sonography for studying the hemodynamics of this vascular anomaly and briefly review the literature.

Publication Types: Review of Reported Cases

PMID: 9107651 [PubMed - indexed for MEDLINE]

Extrahepatic portal vein aneurysm: report of a case treated by thrombectomy and aneurysmorrhaphy.

Glazer S, Gaspar MR, Esposito V, Harrison L.

Department of Surgery, St. Mary Medical Center, Long Beach, California.

Extrahepatic portal vein aneurysm is a rare condition with only 15 cases before ours being reported in the English literature. The etiology is thought to be congenital, secondary to portal hypertension or associated with abnormal weakness of the vein wall. It often presents in conjunction with major gastrointestinal bleeding, but may occur with minimal or no symptoms. Diagnosis is made with color duplex ultrasound, computed tomographic scan, venous phase mesenteric angiography, magnetic resonance imaging, or splenoportography. Thrombosis, rupture, and pressure effects are the major complications of portal vein aneurysm. Shunting procedures are recommended in cases with portal hypertension secondary to liver disease. We report the first case treated by thrombectomy and aneurysmorrhaphy with a successful 10 year follow-up. This procedure should be considered to preserve portal vein flow when portal hypertension is absent or is secondary to the aneurysm itself.

Publication Types: Review of Reported Cases

PMID: 1390021 [PubMed - indexed for MEDLINE]