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遺傳性出血性毛細血管擴張症

Hereditary hemorrhagic telangiectasia ; Osler Weber Rendu disease

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病例報告

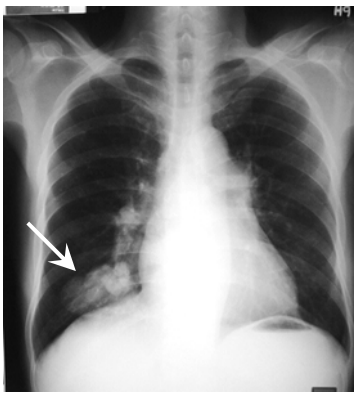
賴先生， 51 歲男性貨車司機， 過去常有流鼻血的病史。 這次入院是因爲頭昏，呼吸困難有 3 至 4 個月，同時陸續偶有解黑便的情況發生。 病患過去無糖尿病或高血壓等疾病，也無特別家族病史。住院之後理學檢查發現病人結膜蒼白，臉頰，舌頭，左右手掌及腳掌分布有毛細血管擴張（telangiectasia）的情形發生（圖一）。聽診發現右下肺野有雜音（bruit）。

圖一:手掌分布有毛細血管擴張（telangiectasia）的情形發生。

住院之後的實驗室檢查結果如下：

CBC: WBC 6.9k/uL, Hb: 3.1g/dL, Plt: 416k/uL, SMA: ALT/AST: 29/35 u/L, Bil(T): 0.65 mg/dL, BUN/Cr: 15/0.8 mg/dL, Stool OB: 4+, Ferritin:7.0 ng/mL

胸部 X 光檢查發現右下肺野圓形陰影（round opacity）（圖二）。



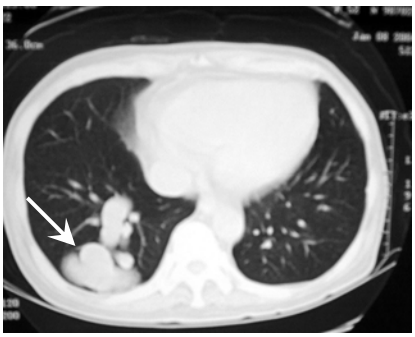
圖二: 胸部 X 光檢查發現右下肺野圓形陰影（round opacity）。

胸部 CT 可發現爲一動靜脈畸形（圖三）。

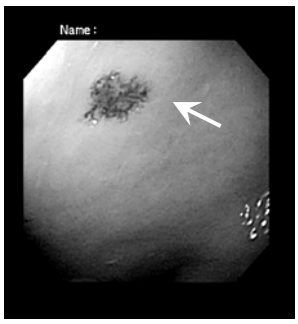
胃鏡檢查發現在胃竇（antrum），胃體部（body）廣泛分布毛細血管擴張及滲血的狀況。（圖四）

腹部超音波檢查爲肝實質病變，無明顯血管異常。

此病人住院診斷爲遺傳性出血性毛細血管擴張症（hereditary hemorrhagic telangiectasia ; Osler Weber Rendu disease）。



圖三: 胸部 CT 可發現爲一動靜脈畸形。



圖四： 胃鏡檢查發現在胃竇（antrum），胃體部（body）廣泛分布毛細血管擴張及滲血的狀況。

討論

早在 1864 年，Sutton 便提出了遺傳性出血性毛細血管擴張症的報告。1896 年，Rendu 首先提出遺傳性，易流鼻血及毛細血管擴張的特徵組合，並把此病與血友病做區分。隨後，Osler 及 Weber 發表了更多的病例報告。到了 1909 年，Hanes 則將其定名爲遺傳性出血性毛細血管擴張症（hereditary hemorrhagic telangiectasia
）。簡稱爲 HHT

HHT 的臨床症狀包括（1）流鼻血，（2）黏膜及皮膚毛細血管擴張（mucocutaneous telangiextasia），（3）臟器的動靜脈畸形（AV malformation）：30%病人有肺動靜脈畸形。大部分出現於下肺部。以及（4）遺傳家族史。以上臨床症狀若有三項吻合則屬於確診病例。25%病人會有腸胃道出血的症狀。反覆腸胃道出血的症狀通常在病人五六十歲時發生。腸胃道內視鏡可發現在胃，十二指腸，小腸，及大腸黏膜有許多大小均一的毛細血管擴張。形狀與口腔及鼻黏膜之毛細血管擴張類似。病人因缺鐵性貧血需要經常輸血及補充鐵劑。

藥物治療包括 estrogen, aminocaproic acid。 內視鏡燒灼止血術包括 BICAP, Heater probe，及 argon plasma coagulation（APC）。APC 原本是用在外科手術如肝葉切除或膽囊切除的止血，1994 年首度運用於內視鏡燒灼止血。APC 可以在短時間內處理較大面積的滲血，器械本身不需與病灶接觸便能達到燒灼止血的效果。對於如 gastric antral vascular ectasia 這類散在分布，多發性的血管異常出血能較有效率的處理。研究顯示內視鏡止血比起單純藥物治療可以增加 Hematocrit, 減少輸血的次數.以及因失血而住院的機會。不過即使內視鏡止血治療成功，有一半的機會在 14 個月內仍會再發出血。

參考資料:

1. Guttmacher AE, Marchuk DA, White RI Jr. Hereditary hemorrhagic telangiectasia**.** N Engl J Med. 1995;333:918-24.
2. Kitamura T, Tanabe S, Koizumi W, Ohida M, Saigengi K, Mitomi H. Rendu-Osler-Weber disease successfully treated by argon plasma coagulation. Gastrointest Endosc. 2001;54:525-7.

巨大肝細胞癌的手術

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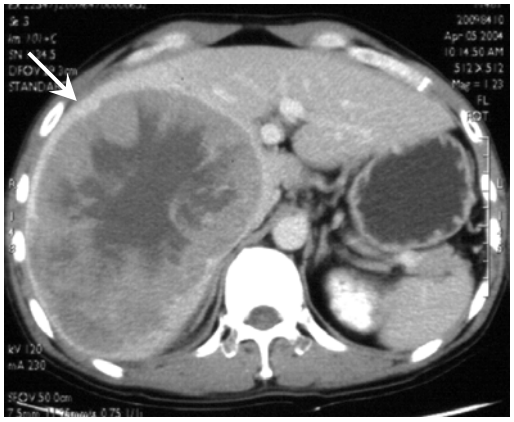
病患爲一 45 歲男性,有 B 型肝炎帶原. 因腹脹,腹部不適,噁心及體重減輕到台南某醫學中心求診,檢查後發現右肝有十幾公分大腫瘤,經轉介到阮綜合醫院作進一步診斷及治療.病患腹部電腦斷層: 右肝巨大肝細胞癌併中心壞死及下腔靜脈壓迫.;腹部超音波: 和腹部電腦斷層相符合,肝硬化及膽結石;生化檢查: AFP:2490 ; HbsAg (+); GOT/GPT=203/33 ; Bilirubin (T/D):0.85/0.15 Albumin:3.6 ; BUN/Cr:20/1.3 ;雖然病患的腫瘤很大且剩餘正常肝容量不高,但殘存肝機能試驗(Reserved Liver Function Test) ICG 值只有 6.2 %.表示病患的肝功能相當好,且又考慮病患還很年輕.在爲病患做其他心肺腎功能詳細評估以及確定沒有明顯其他部位轉移後,我們爲病患進行了擴大右肝切除及膽囊切除手術. 切除的肝標本爲 22.5 x 19 x 6 公分, 腫瘤大小爲 18.5 x 17 x 6 公分.病患術後恢復輕快,於術後 12 天出院,目前門診追蹤治療中.



腹部電腦斷層: 右肝巨大肝細胞癌併中心壞死及下腔靜脈壓迫。

討論

I. 肝臟的惡性腫瘤是死亡的主要原因之一,原發性的肝癌更是台灣十大死亡原因的第一位,近年來每年都有超過五千位病人死於肝癌.而無論是原發性肝癌,或罹患大腸直腸癌併肝臟轉移應當儘量接受手術切除,否則也應儘量想辦法處理肝臟裡的病灶.對肝癌而言,治癒性切除存活率第一年可達 80%,五年後有還有 50%.肝癌的手術,尤其是大範圍切肝手術,它的危險性較高且併發症的發生率也較一般手術來得高.但是醫學的進步,包括:謹慎地手術前的全身檢查,手術中優秀的麻醉醫術和外科技術以及手術後的醫護照顧,使得手術成功的機會大爲提高.以往手術死亡率由 30%以上降至今天的 10%以下,而併發症的發生率也由 50%以上降至 20%以下.至於手術的方法(即切肝要切多大範圍),通常主治醫師會依病人的全身健康情形,(例如有無心、肺、腎、腦等功等異常),腫瘤大小,腫瘤的位置,肝功能好壞等來決定.

參考資料:

[Treatment of hepatocellular carcinoma in the cirrhotic liver]
Hourmand-Ollivier I, Chiche L.
Service d'Hepatogastroenterologie, CHU Cote de Nacre - Caen.

The incidence of hepatocellular carcinoma (HCC) in cirrhotic patients is increasing. Despite advances in imaging and laboratory screening which allow earlier diagnosis, the surgeon is all too often confronted with an HCC of advanced stage or arising in the setting of severe

cirrhosis; this severely limits the treatment possibilities. Treatment options are constrained not only by the characteristics of the tumor but also by hepatocellular reserve, severity of portal hypertension, and the general condition of the host. "Curative treatments" envisage the complete eradication of the malignancy; they include liver transplantation, resection, or tumor destruction by radiofrequency or alcohol ablation. They are most effective in the early stages of HCC. Total hepatectomy and transplantation, by far the most complex surgical therapy, also has the best results avoiding the all-too-frequent local recurrence of HCC in the residual liver. Other medical and interventional treatments (chemo-embolization, radiotherapy with lipiodol) can only slow the progress of the HCC. Goals for the future include more precise and directed screening of the population at risk, and better chemopreventive and chemotherapeutic treatments.

Hepatectomy for hepatocellular carcinoma: patient selection and postoperative outcome.

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Hepatic resection and liver transplantation are considered the only curative treatments for hepatocellular carcinoma (HCC). Liver transplantation for HCCs < or =5 cm in diameter has been shown to produce favorable survival results, but its application is limited by the lack of donors. Hepatic resection remains the treatment of choice for patients who are not transplantation candidates because of large tumor, macroscopic vascular invasion, or advanced age. For small HCCs associated with Child's A cirrhosis, hepatic resection should still be considered the first-line treatment, but salvage transplantation for intrahepatic recurrence may be a feasible strategy. Recent improvement in surgical techniques and perioperative care has increased the safety and expanded the indication of hepatic resection for HCC to include large tumors that require extended hepatectomy in cirrhotic patients. Selection of appropriate candidates for hepatectomy depends on careful assessment of the tumor status and liver function reserve. Evaluation of the general fitness of patients is also critical because comorbid illness is an important cause of postoperative mortality, even if the patients have good liver function reserve. With careful patient selection and surgical expertise, the current operative mortality of hepatectomy for HCC is about 5% or less in major centers. Improved long-term survival results after resection of HCC have also been reported recently, with an overall 5-year survival rate of about 50%. The improved perioperative and long-term survival results have strengthened the role of hepatectomy as the mainstay of treatment for HCC despite the availability of a number of other treatment options for localized HCC.