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肝內結石術後總膽管腫瘤

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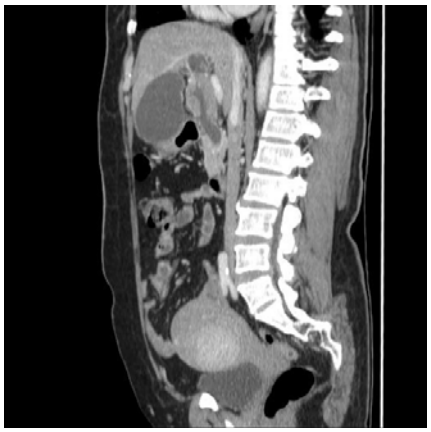
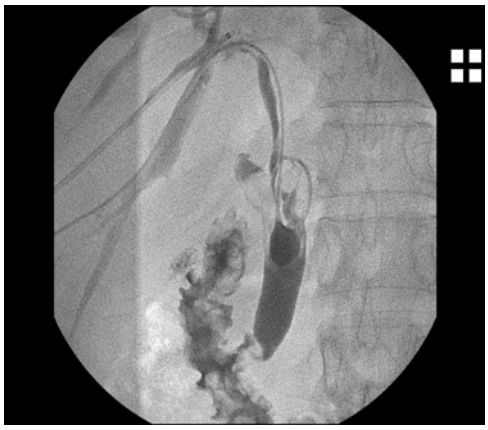
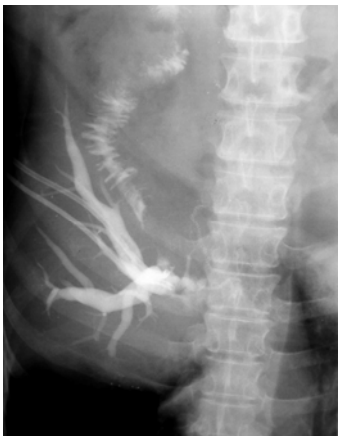
一位56歲女性患者,16年前因左肝內結石接受左外肝葉切除術,約1年前腹部超音波發現總膽管腫瘤.因臨床症狀僅偶爾上腹不適外,並無黃疸或急性膽管炎症狀.直至今年6月初出現黃疸症狀至某醫學中心求診,經腹部斷層掃描,經皮穿肝膽道造影及引流(PTCD)治療,懷疑總膽管惡性腫瘤,建議做膽道支架治療.後經家屬要求轉介至本院治療.

住院時,理學檢查發現已無黃疸症狀,腹部除PTCD傷口疼痛外並無不適.血液檢查報告:
WBC:6600/ul, Hb:12.7g/dl, Plt:380000/ul, GOT/GPT:46/63U/L, Alk-P:74U/L,
Bil.T/D:0.86/0.26mg/dl, BUN/Cr:12/0.7mg/dl, Na/K/Cl:138/4.2/103meq/L, CA199:48.4

腹部斷層掃描(圖一)及經皮穿肝膽道造影(圖二)可見近端總膽管因腫瘤造成管壁增厚併肝內膽管擴張.住院接受手術治療時,術中發現近端總膽管內多發性乳突狀腫瘤併黏液性膽汁分泌.冷凍切片報告為良性乳突瘤,手術術式為膽囊切除,總膽管切除及肝管空腸吻合(hepatico-jejunostomy)併Roux-en-Y吻合術.術後膽道造影(圖三)顯示肝管空腸吻合情況良好.目前門診追踪治療中.


腹部斷層掃描(圖一)


經皮穿肝膽道造影(圖二)


術後膽道造影(圖三)

討論:

1. 黏液性膽道乳突瘤患者臨床上常以反覆性腹痛,黃疸及急性膽管炎來表現.
2. 逆行性內視鏡膽道造影(ERCP)顯示出膽道管壁不規則增厚呈現多發性圓形或卵圓形病灶.腹部超音可見膽道內界線清楚腫瘤併近端膽道擴張.
3. 黏液性(mucin-producing BP)與非黏液性(nonmucin-producing BP)膽道乳突瘤1年存活率為69%與89%, 3年存活率為37%與57%, 5年存活率為19%與52%.
4. 膽道乳突瘤(biliary tract papillomatosis)病理雖為良性病灶(benign neoplasm),但臨床表現為癌前病變(premalignancy),高復發率且具高度惡性變化(high malignant transformation).治療原則視為惡性腫瘤且術後需密切追踪治療.

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1.Cancer. 2004 Feb 15;100(4):783-93.
Clinicopathologic review of 58 patients with biliary papillomatosis.
Lee SS, Kim MH, Lee SK, Jang SJ, Song MH, Kim KP, Kim HJ, Seo DW, Song DE, Yu E, Lee SG, Min YI.
Department of Internal Medicine, University of Ulsan College of Medicine, Asan Medical Center, Seoul, Korea
BACKGROUND: Biliary papillomatosis (BP) is a rare disease that is characterized by multiple numerous papillary adenomas in the biliary tree. The clinical features and outcome, however, are not well known. The authors retrospectively analyzed their clinicopathologic features and long-term follow-up results.

METHODS: Between March 1995 and January 2003, 58 patients were diagnosed with BP by cholangioscopic and histologic findings at a tertiary referral center, Asan Medical Center (University of Ulsan College of Medicine, Seoul, Korea). The authors retrospectively reviewed the medical records to obtain demographic, radiologic, cholangioscopic, and pathologic data.
RESULTS: The common clinical manifestations at the presentation of patients were repeated episodes of abdominal pain, jaundice, and acute cholangitis. Acute cholangitis was more common in patients with mucin-hypersecreting BP (MBP), whereas patients with nonmucin-producing BP (NMBP) were more asymptomatic ($P < 0.05$). Papillary adenocarcinoma and mucinous carcinoma were detected in 48 patients (83%) with papillary adenomas. Overall survival rates of NMBP and MBP were 89% and 69% at 1 year, 57% and 37% at 3 years, and 52% and 19% at 5 years, respectively. The mean survival period of NMBP and MBP was 52.27 +/- 6.72 months and 30.84 +/- 8.36 months, respectively.
CONCLUSIONS: BP should be regarded as a premalignant disease with high malignant potential. The pathogenesis of progression from benign to malignant disease may follow the adenomacarcinoma sequence. Although clinical presentations were somewhat different for patients with NMBP and MBP, the long-term survival rate was similar.

2.Endoscopy. 1998 Nov;30(9):763-7.
Biliary papillomatosis: clinical, cholangiographic and cholangioscopic findings.

Kim YS, Myung SJ, Kim SY, Kim HJ, Kim JS, Park ET, Lim BC, Seo DW, Lee SK, Kim MH, Min YI.
BACKGROUND AND STUDY AIMS: Biliary papillomatosis (BP) is a very rare disease and its clinical features and outcome are not well known. The aims of this study were to describe the characteristic cholangiographic findings and to define the role of cholangioscopic examination in the diagnosis and treatment of this disorder.
PATIENTS AND METHODS: Nine patients (six men and three women, mean age 57 years) were diagnosed as BP among 5361 cases of endoscopic retrograde cholangiography (ERC) from 1990 to 1997 in our institution. The cholangiographic and cholangioscopic findings as well as clinical features were retrospectively analyzed.
RESULTS: ERC findings showed multiple small, round-to-ovoid filling defects in the bile duct and ductal wall irregularity in all the patients. Seven out of nine patients underwent percutaneous transhepatic cholangioscopic (PTC) examination. Additional small papillary lesions in the intrahepatic bile duct were detected by cholangioscopic examination in four patients whose ERC findings only revealed the extrahepatic lesions. Two of these four patients underwent curative resection. In these two patients, the initial surgical plan was changed from Whipple's operation to hepatico-pancratico-duodenectomy after preoperative cholangioscopic examination due to the detection of new lesions in the intrahepatic bile duct.
CONCLUSIONS: ERC findings of BP were highly characteristic. When BP is suspected by conventional imaging including ERC, preoperative percutaneous transhepatic cholangioscopic examination is, however, strongly recommended. This procedure may be beneficial to precisely determine the ductal extension of the disease, hence to decide whether or not hepatic resection is needed as well as to confirm the histology.

惡性黑色素瘤併胃轉移

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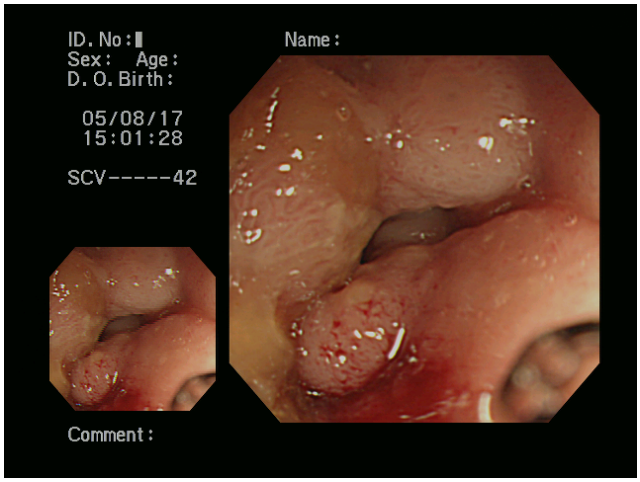
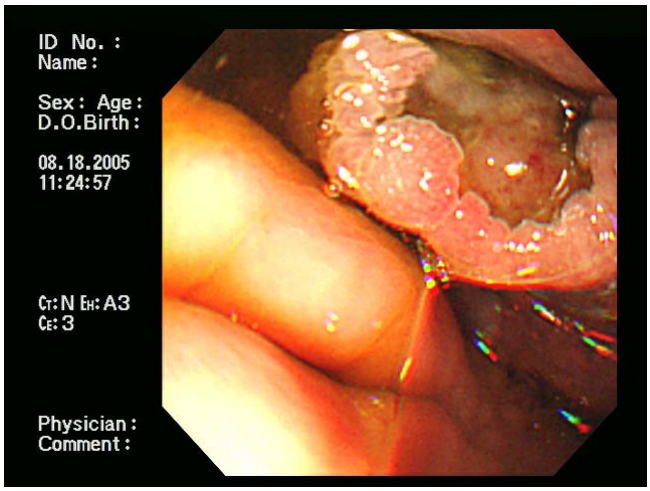
一個 78 歲的男性，因為咳嗽且痰中有血及呼吸困難，被家屬送到急診室求診.經初步檢查發現病人血氧濃度只有 81%，且胸部 X 光發現疑似肺炎的變化，於是收入胸腔科做進一步檢查及治療。

病人身高 162 公分，體重 43 公斤，過去並無全身性的疾病，只有一次因右腳底長雞眼，四年前在本院門診手術切除，之後並未回診。因病人在多年前發作疑似燥鬱症及老人失智症，經常離家失蹤，有時會自行返家，但多數需報警尋回，離家出走通常約數日至一兩個月之間，這一次也是病人離家數周後家人報警尋回，家人發現病人嚴重營養不良，又咳嗽且痰中有血及呼吸困難，家屬叫 119 送到本院急診室求診。

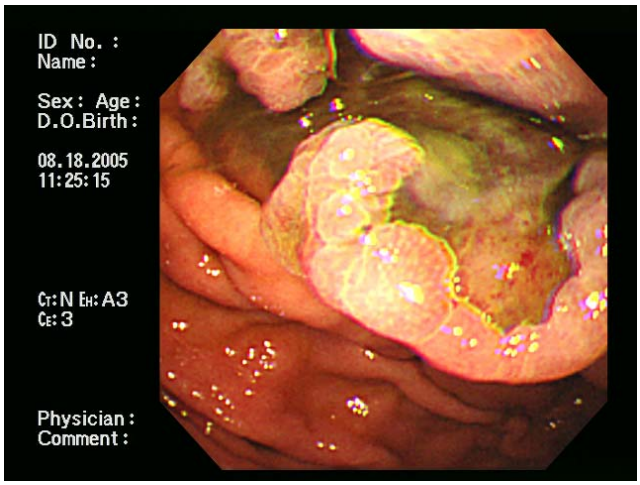
因病人抱怨下腹部疼痛，於是安排腹部超音波掃描，發現病人腹腔內有多顆腫瘤，疑似轉移的病灶。又在膀胱後方發現直腸壁變的非常厚，直腸管腔狹窄，初步診斷為直腸癌併腹腔內轉移。加上病人原先被認為是肺炎的病灶，發現可能是瀰漫性肺轉移病變，而咳血其實是吐血，於是病人轉到消化系內科作進一步檢查。安排大腸鏡檢查，發現在距離肛門口 15 公分處有一個黏膜病灶，造成整個直腸管腔狹窄，甚至整個阻塞，在做了切片檢查之後，因內視鏡無法再深入於是結束檢查。另外安排胃鏡檢查中，發現一個惡性病灶與一般胃癌不太相同病灶，呈現火山口樣的外觀，火山口周圍有一些色素較深的細胞，呈褐色或藍色，分佈在整個病灶表面，經切片檢查後，病理報告為轉移性惡性黑色素瘤。而大腸鏡切片並無惡性細胞只是直腸絨毛狀腺瘤。



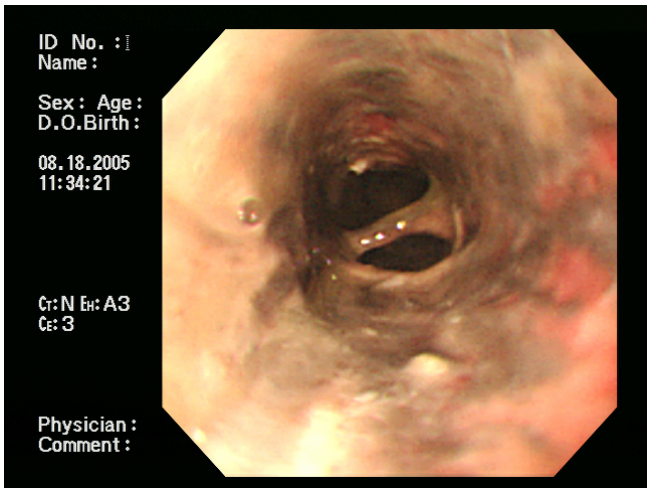
疑似大腸轉移病灶



轉移胃病灶呈現火山口樣病變及色素異常



轉移胃病灶呈現火山口樣病變及色素異常



食道色素異常病變也疑似轉移之病變

在查閱病人的過去病歷，發現四年前的雞眼手術，病理報告為惡性黑色素瘤，而病人沒有再回診治療，因此安排進一步檢查。

但住院後因病人呈現敗血症及惡病質(cachexia)，且血液及痰培養出多種細菌及黴菌(白色念珠菌)，儘管已使用強效的抗生素及全靜脈營養補給(TPN)，病人的病情依然急轉直下，很快的演變成敗血性休克併呼吸衰竭，於住院第八日死亡，而來不及作進一步肺部及腦部轉移的檢查。最後的診斷為肢端痣樣惡性黑色素瘤術後，併胃轉移及疑似直腸、腹腔內、肺部、腦部轉移。

惡性黑色素瘤是源自黑色素細胞(Melanocytes)，大多存在於表皮層中，有時也在真皮層中發現，在白人的發生率約每一萬人 1.5～1.8 例／每年，其他人種少一些，黑人最少，約只有白人的二十分之一，它可在各年齡層發現，最年輕的案例為十幾歲的青少年。近 40 年來發生率約增加三倍。若早期發現可經由外科手術切除完全根治。

惡性黑色素瘤的發病原因尚未完全明瞭，但與過度的陽光曝曬有關。約十分之一的病人有家族病史。若家中有 2 人以上得過惡性黑色素瘤、或是成年人身上有大於 50 個痣大於 2mm 以上或是有變大中的痣的人是高危險症族群，發生率高過正常族群 50 倍以上。白種人，家族中有得過惡性黑色素瘤，或本人先前得過但以根治者為中度危險族群，發生率高過於正常族群 10 倍以上。此外免疫不全(包括使用藥物抑制免疫系統的人，如:器官移植後)或過度曝曬陽光的人，發生發生率高過於正常族 2～4 倍。

惡性黑色素瘤的特徵表現為:痣的邊緣不規則、點狀並具有色素和顏色的改變。70%的早期患者都表現出病灶範圍增大。惡性黑色素瘤的晚期徵像是出血、潰瘍和疼痛。通常惡性黑色素瘤長在慢性日光曝曬的部位(如:顏面、頸部、手背部),但有一種肢端痣樣惡性黑色素瘤(Acral lentiginious melanoma)發生於手掌、腳底、指甲床及黏膜。在男性惡性黑色素瘤最常見於背部。女性則常見於膝至踝的小腿部位。

惡性黑色素瘤是一種相當惡性的腫瘤，轉移得非常快，但如果初期發現，治療效果很好治癒率高。若病灶厚度<0.76mm 即切除 5 年存活率高達 96%～99%，但是一旦轉移，存活率就大幅降低。一個淋巴結轉移者，5 年存活率為 50%，4 個以上淋巴結轉移，5 年存活率降為不到 5%。

惡性黑色素瘤是由淋巴系統或血行轉移常見的部位是肝臟、肺、骨和腦，在少數情況下也可能轉移到其他器官，如:腸、胃…..等等。本病例就是少數轉移至胃及直腸的案例。