1. Coexistence of TDP-43 and tau pathology in neurodegeneration with brain iron accumulation type 1 (NBIA-1, formerly Hallervorden–Spatz syndrome)


Neuropathology (1440-1789) 2011 Jan 30 [Epub ahead of print]

We report here an autopsy case of sporadic adult-onset Hallervorden–Spatz syndrome, also known as neurodegeneration with brain iron accumulation type 1 (NBIA1), without hereditary burden. A 49-year-old woman died after a 27-year disease course. At the age of 22, she suffered from akinesia, resting tremor, and rigidity. At the age of 28, she was admitted to our hospital because of worsening parkinsonism and dementia. Within several years, she developed akinetic mutism. At the age of 49, she died of bleeding from a tracheostomy. Autopsy revealed a severely atrophic brain weighing 460 g. Histologically, there were iron deposits in the globus pallidus and substantia nigra pars reticulata, and numerous axonal spheroids in the subthalamic nuclei. Neurofibrillary tangles were abundant in the hippocampus, cerebral neocortex, basal ganglia, and brain stem. Neuritic plaques and amyloid deposits were absent. Lewy bodies and Lewy neurites, which are immunolabeled by anti-α-synuclein, were absent. We also observed the presence of TDP-43-positive neuronal perinuclear cytoplasmic inclusions, with variable frequency in the dentate gyrus granular cells, frontal and temporal cortices, and basal ganglia. TDP-43-positive glial cytoplasmic inclusions were also found with variable frequency in the frontal and temporal lobes and basal ganglia. The present case was diagnosed with adult-onset NBIA-1 with typical histological findings in the basal ganglia and brainstem. However, in this case, tau and TDP-43 pathology was exceedingly more abundant than α-synuclein pathology. This case contributes to the increasing evidence for the heterogeneity of NBIA-1.

2. TPPV施行のALS患者の直接死因と予後

信園圭吾, 田邊康之

難病と在宅ケア (1880-9200) 16 (1), 55-57, 2010.04

TPPV (侵襲的陽圧換気療法)施行のALS (筋萎縮性側索硬化症)患者の直接死因と予後を明らかにすることを目的に，1983～2007年の当該患者で死亡が確認されている27例を分析した。その結果，VAP（人工呼吸器関連肺炎）を直接死因とするALS症例は減少しているが，敗血症や腎不全，心筋梗塞を死因とする例は依然として多いことが分かった。

3. 歩行障害で発症し，認知症を呈し，剖検にてアルツハイマー病変，ビンスワンガー病変とレビー小体病変を認めた1例

岸本由紀, 石津秀樹, 武田直也, 吉田英純, 大島悦子, 本田肇, 石原武士, 原口俊, 山本真, 中島唯夫, 中島良彦, 寺田整司, 黒田重利
歩行障害で発症し、認知症を呈し、剖検にてアルツハイマー病変、ビンスワンガー病変とレビー小体病変という複数病理の併存が明らかとなった認知症の1例（死亡時91歳女性）について報告した。現在わが国の高齢者、特に認知症を呈する高齢者の中には、認知症+歩行障害という組み合わせは多く、今回はその1例として、18年という長期経過中に複数病理を合併し、認知機能障害の進行が加速し複雑な病像を呈した症例を経験した。本例のように、複合した病理学的成因により生前の確定診断に苦慮する例が潜在的にかなりの割合で存在するものと思われる。

4. 在宅TPPV施行ALS患者における感染予防対策

信國圭吾

難病と在宅ケア（1880-9200）16(9), 13-16, 2010.12

＜免疫疾患＞

5. Nationwide cross-sectional population-based study on the prevalences of asthma and asthma symptoms among Japanese adults.


Int Arch Allergy Immunol 153 (3), 280-287, 2010  Epub 2010 May 20

BACKGROUND: Asthma is a common respiratory disease worldwide. However, few reports are available on the prevalences of asthma and asthma symptoms among Asian subjects. METHODS: To determine the prevalences of asthma and asthma symptoms among Japanese subjects, we performed a nationwide cross-sectional, population-based study on Japanese adults aged 20-79 years. Ten areas spread throughout the country were randomly selected. Door-to-door or postal surveys were performed using a translated version of the European Community Respiratory Health Survey questionnaire. RESULTS: The survey was completed by 23,483 participants. The overall response rate was 70.6%. The prevalences of wheeze and current asthma among all participants aged 20-79 years were 10.1% (95% CI: 9.7-10.5%) and 4.2% (95% CI: 4.0-4.5%), respectively. The prevalences among young adults aged 20-44 years were 9.3% (95% CI: 8.7-9.9%) and 5.3% (95% CI: 4.8-5.8%), respectively. The prevalence of current asthma was highest in females aged 30-39 years in comparison with the other gender and age groups. CONCLUSIONS: This nationwide study determined the prevalences of asthma and asthma symptoms among Japanese adults. The results provide fundamental information on the respiratory health of Japanese adults.

6. 喘息自己管理実行のための携帯電話を用いたe-ダイアリー記録システム（E-Diary Recording System Using Cellular Phones for Asthma Self-Control Practices）（英語）

Okada C, Sumiyoshi T, Mandai T, Suko M, Tomioka H

Quality of Life Journal 11 (1), 25-32, 2010.09

従来、喘息自己管理のために紙による日誌が使用されてきたが、次世代の日誌として携帯電話を用いたe-
ダイアリーシステムを開発した。自分用の携帯電話を所有する例の喘息患者を対象として、毎日20時にeメールを送信し、患者にe-ダイアリーへのアクセスを忘れないようにさせた。e-ダイアリーはピークフロー（PF）値、服用薬、主観的状況、QOL質問用紙から構成されている。3966件のeメールを送信し、3635件の毎日の記録を得た（反応率91.7%）。e-ダイアリーは特にPFやQOLが低い患者で有効であった。本システムは個別の薬物治療や個人のQOL改善に利用できると考えられた。

7. 有病率と喘息死の推移と現状（成人）
谷本安, 髙桥清
The 30th Rokko Conference, 秋山一男・森川昭廣・他 編,ライフサイエンス出版(株), 東京, p.14-20, 2010

8. 追悼 木村郁郎先生を偲んで
髙橋清
アレルギー 60 (2), 214-216, 2011

＜呼吸器疾患＞

9. Non-localized Mycobacterium avium Lung Disease Successfully Treated with Lobectomy and Chemotherapy
Hamada N, Kawata N, Shibayama T, Makihara S, Tada A, Soda R, Takahashi K
Intern Med (0918-2918) 49 (12), 1233-1236, 2010 Epub 2010 Jun 15

A 17-year-old boy presented with a large cavity and bilateral nodular opacities on his chest roentgenogram. Mycobacterium avium was identified in his sputum. According to the recommendations of the American Thoracic Society, he was not strongly recommended to undergo surgery because of non-localized lesions. But since cavities can provide a means for disease to spread to other lobes, we decided to perform a lobectomy including the cavity combined with chemotherapy. Now he has been well for 4 years without exacerbation. There is a possibility of long-term remission with this combination treatment in cases a destructive lesion of airway such as a cavity which is localized to one lobe, even if other lesions such as nodular opacities exist in many other lobes.

10. Phase III trial comparing docetaxel and cisplatin combination chemotherapy with mitomycin, vindesine, and cisplatin combination chemotherapy with concurrent thoracic radiotherapy in locally advanced non-small-cell lung cancer: OLCSG 0007
J Clin Oncol 28 (20), 3299-3306, 2010.06 Epub 2010 Jun 7

PURPOSE: To demonstrate the efficacy of docetaxel and cisplatin (DP) chemotherapy with concurrent thoracic radiotherapy (TRT) for patients with locally advanced non–small–cell lung cancer (LA–NSCLC). PATIENTS AND METHODS: Patients age 75 years or younger with LA–NSCLC, stratified by performance status, stage, and institution, were randomly assigned to two arms consisting of DP (docetaxel 40 mg/m(2) and cisplatin 40 mg/m(2) on
days 1, 8, 29, and 36) or mitomycin, vindesine, and cisplatin (MVP) chemotherapy with concurrent TRT.  RESULTS: Between July 2000 and July 2005, 200 patients were allocated into either the DP or MVP arm. The survival time at 2 years, a primary end point, was favorable to the DP arm (P = .059 by a stratified log-rank test as a planned analysis and P = .044 by an early-period, weighted log-rank as an unplanned analysis). There was a trend toward improved response rate, 2-year survival rate, median progression-free time, and median survival in the DP arm (78.8%, 60.3%, 13.4 months, and 26.8 months, respectively) compared with the MVP arm (70.3%, 48.1%, 10.5 months, and 23.7 months, respectively), which was not statistically significant (P > .05). Grade 3 febrile neutropenia occurred more often in the MVP arm than in the DP arm (39% v 22%, respectively; P = .012), and grade 3 to 4 radiation esophagitis was likely to be more common in the DP arm than in the MVP arm (14% v 6%, P = .056).  CONCLUSION: DP chemotherapy combined with concurrent TRT is an alternative to MVP chemotherapy for patients with LA-NSCLC.


Lung Cancer 2011 Feb 17 [Epub ahead of print]

BACKGROUNDs: Chemotherapy is a mainstay in the treatment of extensive-disease small-cell lung cancer (ED-SCLC), although the survival benefit remains modest. We conducted a phase II trial of amrubicin (a topoisomerase II inhibitor) and topotecan (a topoisomerase I inhibitor) in chemotherapy-naïve and relapsed SCLC patients.
METHODS: Amrubicin (35mg/m(2)) and topotecan (0.75mg/m(2)) were administered on days 3–5 and 1–5, respectively. The objective response rate (ORR) was set as the primary endpoint, which was assessed separately in chemotherapy-naïve and relapsed cases.
RESULTS: Fifty-nine patients were enrolled (chemotherapy-naïve 31, relapsed 28). The ORRs were 74% and 43% in the chemotherapy-naïve and relapsed cases, respectively. Survival data were also promising, with a median progression-free survival time and median survival time of 5.3 and 14.9 months and 4.7 and 10.2 months in the chemotherapy-naïve and relapsed cases, respectively. Even refractory-relapsed cases responded to the treatment favorably (27% ORR). The primary toxicity was myelosuppression with grades 3 or 4 neutropenia in 97% of the patients, which led to grades 3 or 4 febrile neutropenia in 41% of the patients and two toxic deaths.
CONCLUSION: This phase II study showed the favorable efficacy and moderate safety profiles of a topotecan and amrubicin two-drug combination especially in relapsed patients with ED-SCLC.

12. 非結核性抗酸菌症と薬剤感受性試験

河田典子

非結核性抗酸菌症の臨床 ISBN:978-4-88002-709-8, 佐々木結花・小川賢二 編, pp.75-78, (株)新興医学出版社, 東京, 2010.10

＜血液疾患＞

13. イマチニブに対してLate Suboptimal Responseを示した慢性期CML症例に対する治療選択

松村到, 今村賢一, 杉本正, 坂本一樹, 藤川一輝, 竹内義信, 杉山齋子, 村田美智子, 小野寺晴, 松本一, 坂本美里, 岡部雅弘, 植木直樹, 今村昌也, 足立陽子,
European Leukemia Netの判定基準は慢性期の慢性骨髄性白血病をイマチニブ（IM）で治療した際の予後推定に有効であるが、18ヵ月以降のLate Suboptimal Response（CCgR達成、MMR未達成：LSR）の評価には異論がある。本研究では国内のLSR16例を解析した。IMの投与期間中央値は62（25-87）ヵ月、開始量中央値は400mg/日、10例で種々の理由により600-800mg/日に増量された。LSRに対してIMが増量、継続された4例中2例はMMRを達成、残り2例も分子遺伝学的な改善を示した。IMを増量しなかった6例では4例のみがその後MMRを達成した。今回の16例では病期進行はなかった。以上より、LSR例のMMR達成にはIM増量が有効と考えられた。ただし、ニロチニブなどの第二世代TKIは、より早期にMMRを達成し病期進行のリスクを軽減することも期待される。

＜耳鼻科＞

14. Differential expression of glycogenes in tonsillar B lymphocytes in association with proteinuria and renal dysfunction in IgA nephropathy


Aberrant O-glycosylation of serum and tonsillar IgA1 is one of the main pathogeneses of IgA nephropathy (IgAN). However, the synthesis of underglycosylated IgA1 in tonsils has not yet been characterized. This study examined tonsillar B lymphocytes of IgAN (n=34) using tonsils derived from patients with chronic tonsillitis (n=24) and sleep apnea syndrome (n=14) as a control. Gene expression of β1,3-galactosyltransferase (β3GalT), and the core 1β3GalT-specific molecular chaperone, Cosmc, UDP-N-acetyl-α-D-galactosamine: polypeptide N-acetylgalactosaminyl-transferase 2, were significantly decreased in tonsillar CD19-positive B lymphocytes from IgAN patients compared to control tonsillar tissues as determined by real-time RT-PCR. Tonsillar B cell β3GalT gene expression significantly correlated with estimated GFR and negatively correlated with proteinuria and histological injury score. Western blotting showed the protein expression of β3GalT in the tonsils to significantly decrease in IgAN in comparison to the controls. These data suggest the downregulation of β3GalT in tonsillar B lymphocytes to be closely associated with the clinical characteristics of IgAN.

15. 移植腎IgAN腎症例への扁摘・パルス療法

土井彰, 田村耕三, 島本久美子, 小川愛由, 松岡孝至, 土山芳徳, 渋谷祐一, 赤木博文

口腔・咽頭科 (0917-5105) 23 (2), 129-132, 2010.06

Recurrent IgA nephropathy is associated with a better prognosis than primary IgA nephropathy. Given previous cases of recurrent IgA nephropathy soon leading to renal dysfunction and the lack of reports on long-term prognosis, the actual recurrent IgA nephropathy prognosis may be worse than suggested. We conducted tonsillectomy with steroid pulse therapy in 5 cases of IgA nephropathy associated with kidney transplantation. Of these 4 had idiopathic renal failure, and thus were considered recurrent IgA nephropathy. Although steroids are continuously used after
renal transplant to prevent graft refection, IgA nephropathy still recurs, suggesting that even steroid therapy cannot completely prevent onset or progression. Given subjects’ ages and results at renal biopsy, we expect that they will eventually develop renal dysfunction and decided to conduct surgery as soon as possible. Our findings suggest that this combination is at least initially effective in such subjects because no microhematuria or recurrence was seen within 3 months.

16. 中毒疹を発症した化学物質過敏症症例
内田浩志, 土井彰, 赤木博文
口腔・咽頭科 (0917-5105) 23 (2), 157-162, 2010.06

A 51-year-old woman admitted for a cervical phlegmon following odontogenic infection had a history of multiple chemical sensitivity. During antibiotic therapy, she developed a rash on the palms and lower extremities, diagnosed as toxicoderma by a dermatologist. Possible causes included drug-induced, infection, and multiple chemical sensitivity. A change in antibiotics and corticosteroid use did not improve her rash. On day 8 after admission, she sensed a stimulating smell at the newly built outpatient clinic and her rash worsened a few hours later. After the cervical phlegmon resolved, she was transferred to the dermatology department. Although she continued to be treated, she was discharged without complete rash disappearance and her rash disappeared and after she was discharged her rash soon disappeared. Suggesting that her skin lesion was due to multiple chemical sensitivity.

17. IgA腎症に対する扁桃摘出術のEBMとは？—耳鼻咽喉科の立場から—. 扁桃病巣疾患
赤木博文

18. 耳鼻咽喉科学用語解説集
赤木博文（分担執筆）
耳鼻咽喉科学用語解説集 (ISBN978-4-7653-1433-6), 日本耳鼻咽喉科学会 編, (株)金芳堂, 京都, 2010.10
＜整形外科＞

19. Measurement of serum hepcidin-25 levels as a potential test for diagnosing hemochromatosis and related disorders
J Gastroenterol 45 (11), 1163-1171, 2010 Epub 2010 Jun 9

BACKGROUND: Iron overload syndromes include a wide spectrum of genetic and acquired conditions. Recent studies suggest suppressed hepcidin synthesis in the liver to be the molecular basis of hemochromatosis. However, a liver with acquired iron overload synthesizes an adequate amount of hepcidin. Thus, hepcidin could function as a biochemical marker for differential diagnosis of iron overload syndromes. METHODS: We measured serum iron
parameters and hepcidin-25 levels followed by sequencing HFE, HJV, HAMP, TFR2, and SLC40A1 genes in 13 Japanese patients with iron overload syndromes. In addition, we performed direct measurement of serum hepcidin-25 levels using liquid chromatography–tandem mass spectrometry in 3 Japanese patients with aceruloplasminemia and 4 Italians with HFE hemochromatosis. RESULTS: One patient with HJV hemochromatosis, 2 with TFR2 hemochromatosis, and 3 with ferroportin disease were found among the 13 Japanese patients. The remaining 7 Japanese patients showed no evidence for genetic basis of iron overload syndrome. As far as the serum hepcidin-25 was concerned, seven patients with hemochromatosis and 3 with aceruloplasminemia showed markedly decreased serum hepcidin-25 levels. In contrast, 3 patients with ferroportin disease and 7 with secondary iron overload syndromes showed serum hepcidin levels parallel to their hyperferritinemia. Patients with iron overload syndromes were divided into 2 phenotypes presenting as low and high hepcidinemia. These were then associated with their genotypes. CONCLUSION: Determining serum hepcidin-25 levels may aid differential diagnosis of iron overload syndromes prior to genetic analysis.


To assess the effects of service dogs on health–related quality of life (HRQOL), we conducted a survey of 10 service dog owners using SF–36v2 (Medical Outcomes Study 36 Item Short–Form Health Survey Version 2.0) and compared it with a matched control group of people with physical disabilities who did not have service dogs but were eligible for one. The scores for mental health and role emotional of service dog owners were relatively high, and their mental component summary was higher than the general population norm. These results indicate that service dogs affect the mentality of their owners. The comparison with the control group indicated that service dogs alleviate the mental burden of daily activities, and subjectively improved the physical functioning of their owners. This study showed that service dogs have positive functional and mental effects on their disabled owners.

<外科>
21. 低肺機能高齢者に対して１年間に３回の開腹手術を施行した１例
奥谷大介, 永井太士, 小谷一敏, 牧原重喜, 奥谷珠美
Journal of Clinical Rehabilitation (0918–5259) 20 (1), 99–102, 2011.01

86歳男。呼吸不全にて呼吸器内科通院中、数回の血便を認めた。下部消化管内視鏡検査にてS状結腸がんと診断された。S状結腸切除、上行結腸部分切除術(D1郭清)を施行した。呼吸器合併症も認めず術後16日目に退院した。以降経過良好であったが、大量の下血のためショック状態で受診した。下部消化管内視鏡検査、造影検査にてS状結腸憩室出血と診断された。また、同下部消化管内視鏡検査において上行結腸にも腫瘍変を認め、組織生検によって高分化型腺がんと診断された。S状結腸切除術、上行結腸部分切除術(D1郭清)を施行した。術後16日に黑色便が出現し、上部内視鏡検査にて十二指腸潰瘍と診断された。絶飲絶食、PPI投与にて軽快した。術後35日目に退院した。発熱と上腹部痛が出現し、総胆管、胆嚢内に結石を伴う急性胆囊炎と診断された。胆囊摘出術、総胆管切開術を施行した。術後33日目に退院した。呼吸・全身状態は
良好である。

＜麻酔科＞
22. 麻酔科領域におけるScalable Vector Graphicsの利用
齋藤智彦
麻酔・集中治療とテクノロジー 2008, 31-33, 2010.05

23. 肥満体型の胸部X線写真の特徴
齋藤智彦
呼吸器&循環器ケア 10 (5), 53-57, 2010.12

＜看護部＞
24. リンクナースを手指衛生のオピニオンリーダーに育てる
形山優子
INFECTION CONTROL (0919-1011) 19 (6), 550-553, 2010.06

25. DOTSの取り組み 独立行政法人国立病院機構南岡山医療センター
川本奈保, 橋本数江
保健師・看護師の結核展望 (1347-4553) 48(1) (2010 前期号), 60-66, 2010.08
保健師・看護師の結核展望 (1347-4553)通巻 95, 1-7, 2010.08

26. 結核患者の服薬に対する思いについて 退院後の面接調査を行って見えてきたもの
野坂章子, 寒川啓子, 原知江, 藤本みどり, 藤田笑子
中国四国地区国立病院機構・国立療養所看護研究学会誌 (1880-6619) 6, 219-221, 2011.01

自己管理で抗結核薬の服用を継続している患者の思いについて明らかにするため、退院後2週間〜1ヵ月経過した4名(男3名、女1名、20歳代〜70歳代)を対象に面接調査を行った。面接内容を分析しカテゴリー化した結果、18サブカテゴリー、「内服を継続するための工夫」「疾患、内服に対する理解、知識」「疾患治療、内服に対する不安、困ったこと」「継続内服する上での日常生活」「継続内服の危うさ」の5カテゴリーが抽出された。退院した患者は、疾患や内服については理解できていたが、退院後1ヵ月の間でも、日常生活に戻り規則性を失うことが薬の飲み忘れにつながり、危うい内服状況を形成していることがわかった。

27. 神経・筋難病棟の腰痛を持つ職員の思いの分析
西田聖子, 藤原典子, 平良早代, 加藤元樹, 宮本敏子
中国四国地区国立病院機構・国立療養所看護研究学会誌 (1880-6619) 6, 84-87, 2011.01

神經・筋難病棟に勤務する腰痛を持つ職員の思いを明らかにするため、看護師2名、療養介助員1名を対象にインタビューを実施し、内容分析にてカテゴリー化を行った。334コード、17サブカテゴリー、「腰痛は我慢」「特殊な看護・介護業務」「自分で工夫」「悪影響の恐れ」「業務支障の自責」「大事な腰」「他職員の腰痛の有無」の7カテゴリーが抽出された。腰痛を持つ職員は、腰痛は我慢していることが分かった。また、
他職員の腰痛の有無を気にしたり、業務支障の自責を感じたり、腰痛は精神的にも影響してくることが分かった。腰痛の負担が軽くなるような介護機器の導入により、患者の安全と安楽を確保すると共に、看護師・療養介助員自身を腰痛から守ることが重要である。

28. 化学療法を受けた男性患者の脱毛に対する意識調査
佐藤章子, 越智紘子, 三谷順子, 池上美智子
中国四国地区国立病院機構・国立療養所看護研究学会誌 (1880-6619) 6, 5-8, 2011.01

化学療法後の男性の脱毛に対する意識を明らかにするため、男性患者4名(20〜30歳代3名、60歳代1名)を対象に聞き取り調査を行い、内容分析の手法を参考にカテゴリ化した。「変化の拒否」「脱毛の知識不足」「過剰な自意識」「周囲の理解」「習慣の性差」「治療の利点」の6カテゴリーが抽出された。前回の研究で、女性は容姿の変化による精神的ショックが大きいことが分かったが、今回の研究では、男性も抗癌剤で初めての脱毛を経験し、容姿の変化に抵抗があり、ショックを受けていることが分かった。脱毛が始まったとき、患者の気持ちに寄り添い、コミュニケーションをとってゆく必要があると考えられた。