

BACKGROUND: Clinical presentations of pathologically confirmed corticobasal degeneration (CBD) vary, and the heterogeneity makes its clinical diagnosis difficult, especially when a patient lacks any motor disturbance in the early stage.

METHODS: We compared clinical and pathological features of four pathologically confirmed CBD cases that initially developed non-motor symptoms, including behavioural and psychiatric symptoms but without motor disturbance (CBD-NM), and five CBD cases that initially developed parkinsonism and/or falls (CBD-M). The age range at death for the CBD-NM and CBD-M subjects (58–85 years vs 45–67 years) and the range of disease duration (2–18 years vs 2–6 years) did not significantly differ between the groups.

RESULTS: Prominent symptoms in the early stage of CBD-NM cases included self-centred behaviours such as frontotemporal dementia (n = 1), apathy with and without auditory hallucination (n = 2), and aggressive behaviours with delusion and visual hallucination (n = 1). Among the four CBD-NM cases, only one developed asymmetric motor disturbance, and two could walk without support throughout the course. Final clinical diagnoses of the CBD-NM cases were frontotemporal dementia (n = 2), senile psychosis with delirium (n = 1), and schizophrenia (n = 1). Neuronal loss was significantly less severe in the subthalamic nucleus and substantia nigra in the CBD-NM cases than in the CBD-M cases. The severity of tau pathology in all regions examined was comparable in the two groups.

CONCLUSION: CBD cases that initially develop psychiatric and behavioural changes without motor symptoms may have less severe degenerative changes in the subthalamic nucleus and substantia nigra, and some CBD cases can lack motor disturbance not only in the early stage but also in the last stage of the course.

2. 【ALS の長期経過予後】 長期入院 ALS 患者の急変時の指示

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難病と在宅ケア(1880–9200) 2014.11; 20: 24–27

長期療養者が多い当院に入院中のALS(筋萎縮性側索硬化症)患者28名を対象に、呼吸器の有無とコミュニケーション状態、および急変時の指示の有無について検討した。その結果、28名中25名が呼吸器を使用しており、内訳はTPPV(気管切開下陽圧換気療法)が20名、NPPV(非侵襲的陽圧換気療法)が5名で、呼吸器を使用していない3名とNPPVの5名は、急変時での挿管などの処置を本人または家族が希望しておらず、全員が何らかの急変時の対応を指示していた。また、TPPVの患者では、コミュニケーションがとれない患者の方が、とれる患者よりも急変時の対応を決める傾向がみられた。
4. **Complete resolution of steroid-resistant organizing pneumonia associated with myelodysplastic syndrome following allogeneic hematopoietic cell transplantation.**

Pulmonary complications in patients with hematological malignancies are often caused by infection but are sometimes associated with an underlying disease such as organizing pneumonia (OP). Here, we report a case of life-threatening steroid-resistant OP associated with myelodysplastic syndrome (MDS) and successfully performed allogeneic hematopoietic cell transplantation (HSCT). A 33-year-old female with refractory anemia with excess blasts-1 that had progressed from refractory anemia with ringed sideroblasts and concomitant Sweet’s syndrome was admitted. Multiple pulmonary infiltrates were revealed on a chest computed tomography scan, which progressively worsened even after chemotherapy and corticosteroid therapy. No evidence of infection was observed in bronchoalveolar lavage fluid. A histological examination of a transbronchial lung biopsy specimen showed lymphocyte invasion with fibrosis, indicating that the pulmonary infiltrates were OP associated with MDS. Before transplantation, she suffered from respiratory failure and required oxygen supplementation. She developed idiopathic pneumonitis syndrome on day 61 that responded well to corticosteroid therapy, and the OP pulmonary infiltrates improved gradually after HSCT. She was discharged on day 104 and is well without recurrence of OP or MDS 2 years after HSCT.

Keywords: Organizing pneumonia; Myelodysplastic syndrome; Sweet’s syndrome; Allogeneic hematopoietic cell transplantation

5. **A case of systemic lupus erythematosus presenting with an acute abdomen: successful treatment with steroid.**
Fukatsu H, Ota S, Sugiyama K, Kasahara A, Matsumura T.

Abdominal pain continues to pose diagnostic challenges for emergency clinicians. A 56-year-old Japanese woman was referred to our hospital due to severe abdominal pain which presented as occasional epigastric pain five months before and intermittent abdominal pain. She had a past history of ileus twice, for both of which laparotomy was performed without an alimentary tract resection. The wall thickening with marked three-wall structure from terminal ileum to sigmoid colon was seen and bladder wall was irregularly thick and enhanced irregularly. Among the differential diagnosis of the acute abdomen, autoimmune diseases were suspected, especially lupus erythematosus and Henoch–Schönlein purpura. On the second day of admission, abdominal pain worsened. The results of examinations of antinuclear antibody, anti–double–stranded DNA antibody, ANCA, and the complements were not obtained at that...
time; however, we started 1-g steroid pulse treatment for three days with success. With the results obtained later, the patient was given a diagnosis of probable systemic lupus erythematosus (SLE). The present case shows that SLE can present with acute abdomen and should be included in the wide range of the differential diagnosis of acute abdomen.

6. 岡山県下のクリニック・診療所におけるリウマチ診療・病診連携の実態に関する研究
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岡山県下のクリニック・診療所を対象に関節リウマチ(RA)診療・病診連携に関するアンケート調査を実施し、実態について検討した。その結果、依頼した300施設のうち、1回の再依頼の結果150施設(50%)から協力可能との回答が得られ、最終的に100施設(33%)から回答を得た。医師の臨床経験年数は平均30年で、内科医が80%を占め、日本リウマチ学会専門医は1%、日本整形外科学会認定リウマチ医は5%であった。診療に関しては、何らかの形でRAに関わっている医師が83%を占め、岡山県下でRAと診断された患者の84%がガイドラインで推奨されている適切な治療を開始されていることが推察された。病診連携の66%の施設で行われており、担っている役割は主に「診察」「検査」「処方」「common diseaseへの対応」等であった。更なる病診連携の発展のためには、適切な情報提供やより質の高い連携体制を構築する必要があると考えられた。

7. 吸入療法の意義を理解しよう
谷本安, 宗田良

ApoTalk 2014.08; 38: 12-13

8. 小児/高齢者の吸入療法の注意点
木村五郎, 宗田良

ApoTalk 2014.10; 39: 12-13

＜呼吸器疾患＞


BACKGROUND: Although cisplatin-based chemotherapy combined with thoracic irradiation (TRT) is a standard treatment for unresectable, locally advanced non-small cell lung cancer (NSCLC), this treatment outcome has remained unsatisfactory. We had previously conducted a phase I trial of cisplatin plus S-1, an oral 5-fluorouracil derivative, and TRT, which were safe and effective.
METHODS: In this phase II trial, 48 patients with stage III NSCLC received cisplatin (40mg/m(2) on days 1, 8, 29 and 36) and S-1 (80mg/m(2) on days 1-14 and 29-42) and TRT (60Gy). The primary endpoint was the response rate.

RESULTS: A partial response was observed in 37 patients (77%; 95% confidence interval: 63–88%). At a median follow up of 54 months, the median progression-free survival and median survival time were 9.3 and 31.3 months, respectively. No difference in efficacy was observed when the patients were stratified by histology. Toxicities were generally mild except for grade 3 or worse febrile neutropenia and pneumonitis of 8% and 4%, respectively. No patient developed severe esophagitis. At the time of this analysis, 35 (73%) of the 48 patients recurred; 15 (31%) showed distant metastasis, 17 (35%) had loco-regional disease, and 2 (4%) showed both loco-regional disease and distant metastasis.

CONCLUSIONS: This chemoradiotherapy regimen yielded a relatively favorable efficacy with mild toxicities in patients with locally advanced NSCLC.

KEYWORDS: Chemotherapy; Cisplatin; Phase II trial; Radiation; S-1; Stage III

10. Invasive mucinous adenocarcinoma mimicking organizing pneumonia associated with Mycobacterium fortuitum infection.


We herein report the case of a 68-year-old man diagnosed with invasive mucinous adenocarcinoma of the lungs. Chest computed tomography showed subpleural ground-glass opacity and small nodules with cavitation. A culture of the bronchoalveolar lavage fluid resulted in the detection of Mycobacterium fortuitum. The patient’s lung consolidation rapidly progressed; however, repeated bronchoscopy showed no atypical cells, thus suggesting a diagnosis of organizing pneumonia associated with M. fortuitum infection. However, the surgical biopsy specimen was diagnostic for adenocarcinoma, with no mycobacterial infection. Invasive mucinous adenocarcinoma should not be excluded in the differential diagnosis of patients with clinical features of organizing pneumonia and nontuberculous mycobacterium infection, even if a transbronchial biopsy confirms the absence of malignancy.

11. 肺胞出血症候群の診断と治療

谷本安


12. 慢性呼吸器疾患（COPD, 気管支拡張症, 陳旧性肺結核等）の気道感染症治療ガイドライン

谷本安

岡山医学会雑誌 (0030-1558) 2014.08; 126: 151-153

＜重症心身障害児・者＞

13. 対応が難しい場合の家族ケアの進め方

遠部泰子

こどもケア 2014.06; 9(2): 87-90
14. 治療とケア 症例から考える 短い恐怖のエピソードを繰り返す女児
秋山麻里
Epilepsy (1882-1480) 2014.05; 8(1): 34-37

15. 【けいれん性疾患の最新の治療】 慢性期 新しい抗てんかん薬の適応と使い方
高橋幸利、長尾雅悦、遠山潤、渡邊宏雄、夫敬憲、井上美智子、馬場健至
小児科診療 (0386-9806) 2015.02; 78(2): 207-214

＜診療支援部＞

16. 胎内栄養環境と高血圧症 -成人病胎児期発症起源説の視点から考える-
福岡秀興、平野大志、向井伸二
血圧 (1340-4598) 2014.10; 21(10): 841-848

17. 25年度精度管理報告 生理機能検査部門 心臓・血管超音波検査
高松泉、宮本由紀子、井石充洋
岡山医学検査 (1349-1504) 2014.07; 51(2): 104-109