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CASE REPORT

Efficacy of nintedanib in a patient with acute exacerbation of idiopathic pulmonary fibrosis

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Introduction

Acute exacerbation of idiopathic pulmonary fibrosis is a disease with a poor prognosis, and it is unclear whether antifibrotic therapy is effective for this condition. This article reports a patient with an acute exacerbation of idiopathic pulmonary fibrosis, who was successfully treated with nintedanib combined with corticosteroids and anticoagulant therapy.

Case report

A 69-year-old man developed fever, cough and progressive dyspnoea on 19 January 2019. A chest computed tomography scan showed subpleural and basal predominant ground-glass opacities (Figures 1a and b). He was diagnosed with pneumonia, and antibiotic treatment was attempted in his local hospital but failed. He was transferred to the authors' hospital 9 days later, and on examination, he had clubbing fingers and Velcro crackles. Past medical history included smoking for 40 years; he had no connective tissue diseases. Laboratory investigations showed the following results: white blood cell count was 4.6×10^9/litre, C-reactive protein level was 208.5 mg/litre, D-dimer level was 11 350 ug/litre. The results of blood and sputum cultures were negative. Ultrasound examination showed deep vein thrombosis. Computed tomography pulmonary angiography was normal. In addition, a high resolution computed tomography scan of the chest on admission to the authors' hospital showed a significant increase in lesion size (Figures 1c and d).

The patient was treated with methylprednisolone 40 mg and low molecular weight heparin. However, 8 days later, his symptoms and chest high resolution computed tomography had worsened (Figures 2a and b), so nintedanib 150 mg twice daily was prescribed for anti-fibrosis treatment 15 days after his initial admission to the authors' hospital. After 3 weeks, the patient's symptoms improved, with just some abdominal distension. Two months later, a follow-up chest computed tomography scan showed significant improvement (Figures 2c and d).

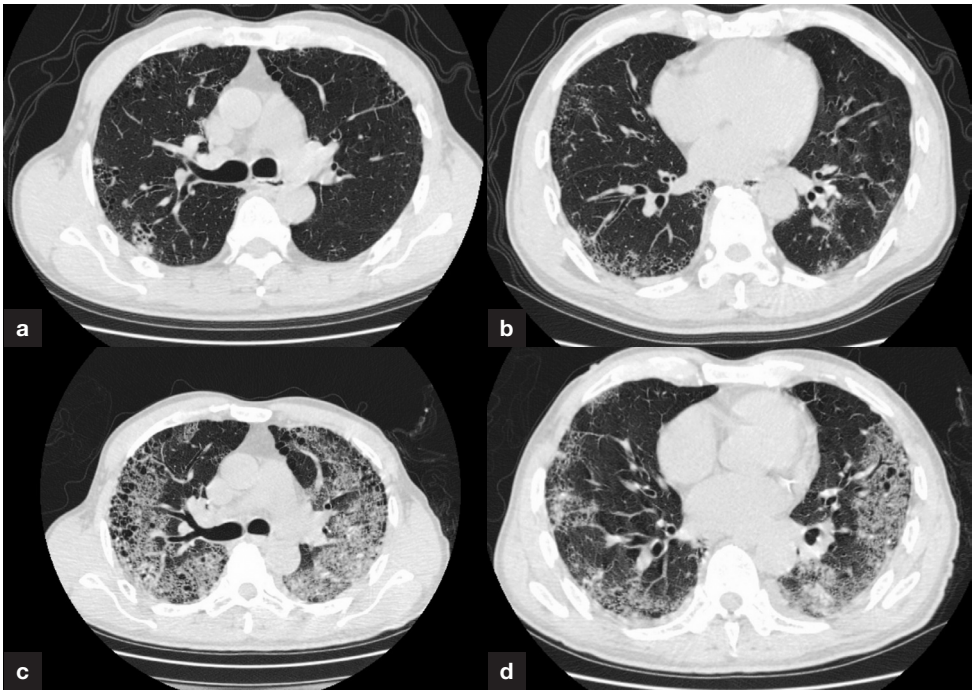

Figure 1. Chest high resolution computed tomography scan (a,b) on admission to the patient's local hospital, showing subpleural and basal predominant ground-glass opacities, and (c,d) 9 days later, showing aggravation of ground-glass opacities and diffuse mosaic sign.

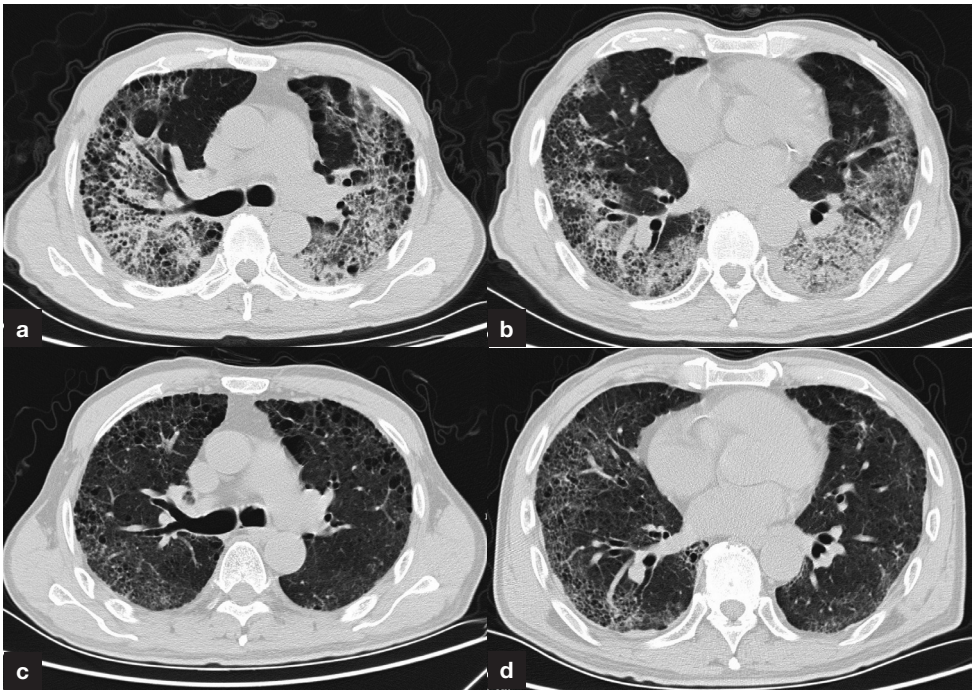

Figure 2. Chest high resolution computed tomography scan (a,b) showing apparent fibrosis and honeycomb pattern, 8 days after admission to the authors' hospital and (c,d) improvement after 2 months of nintedanib therapy.

Discussion

Nintedanib is recommended for the treatment of idiopathic pulmonary fibrosis (Raghu et al, 2015). The INPULSIS trial showed that it may improve quality of life for patients with an acute exacerbation of idiopathic pulmonary fibrosis, but these data are exploratory and need more clinical studies to confirm them (Collard et al, 2017).

Nakashima et al (2021) reported two cases of acute exacerbation of idiopathic pulmonary fibrosis treated with nintedanib after corticosteroid pulse therapy, and the symptoms were significantly improved. It should be noted that acute exacerbation of idiopathic pulmonary fibrosis is often accompanied by deep vein thrombosis. Anticoagulation therapy (recombinant human soluble thrombomodulin) has a potential impact on acute exacerbation of idiopathic pulmonary fibrosis (Tsushima et al, 2014). The authors' patient was promptly treated with corticosteroids and anticoagulant therapy (low molecular weight heparin), but his symptoms did not improve, in contrast to the improvements seen by Nakashima et al (2021), whereas the addition of nintedanib significantly improved this patient's symptoms.

Uzel et al (2020) reported a patient with acute exacerbation of idiopathic pulmonary fibrosis caused by COVID-19, who also improved after treatment with nintedanib, but the current patient's disease (admitted in January 2019) was not related to acute COVID-19 pneumonitis. Pirfenidone is another antifibrotic drug approved for idiopathic pulmonary fibrosis, and its use combined with corticosteroids and anticoagulants can improve the survival rate of patients with acute exacerbation of idiopathic pulmonary fibrosis (Furuya et al, 2017). The efficacy of nintedanib and pirfenidone in these patients requires further research.

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Learning points

- Nintedanib may be considered as an additional treatment for patients with acute exacerbation of idiopathic pulmonary fibrosis in whom corticosteroid treatment does not work.
- Nintedanib combined with corticosteroids and anticoagulant therapy improved this acute exacerbation of idiopathic pulmonary fibrosis.
- The efficacy of nintedanib in patients with acute exacerbation of idiopathic pulmonary fibrosis requires further research.

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