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CASE REPORT

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An autopsy case of acute fibrinous and organizing pneumonia with periorbital MRSA cellulitis and rheumatoid arthritis

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Abstract

This case report details a 78-year-old male with periorbital Methicillin-resistant Staphylococcus aureus (MRSA) cellulitis whose condition rapidly deteriorated despite treatment. An autopsy confirmed acute fibrinous and organizing pneumonia (AFOP), revealing fibrin ball formation and organizing pneumonia. While both idiopathic and secondary AFOP cases often exhibit bilateral consolidation on CT, our patient presented with ground-glass opacities, which are frequently associated with secondary AFOP. Notably, secondary AFOP, linked to higher mortality, can result from various factors. In this case, well-controlled rheumatoid arthritis and prolonged oral medication use suggest bilateral periorbital MRSA cellulitis as a significant factor. The study underscores AFOP's diagnostic challenges and the necessity for further research on effective treatments.

KEYWORDS
acute fibrinous and organizing pneumonia, ground-glass opacities, MRSA cellulitis

INTRODUCTION

Acute fibrinous and organizing pneumonia (AFOP) was originally described in 2002 by Beasley et al. as a histologic pattern associated with an acute or subacute disease onset that did not meet the histologic criteria of diffuse alveolar damage (DAD), acute eosinophilic pneumonia, or organizing pneumonia, but rather consisted of organizing intra-alveolar fibrin balls without hyaline membrane formation, significant neutrophils, eosinophils, or macrophages.[1] The overall mortality rate was similar to DAD. Over half of the cases were associated with an underlying aetiology such as collagen vascular disease, occupational exposure, drug reaction, hematologic disease, or infection.[2,3] Recently cases of SARS-CoV-2 infection were reported.[4] Immunosuppressants have been used for treatment, but therapeutic responses are variable and there is no established treatment. Here, we describe an autopsy case of AFOP.

CASE REPORT

A 78-year-old male with a history of type 2 diabetes and rheumatoid arthritis, without a smoking history, presented with bilateral periorbital edema for 1 week and fever with dyspnea for 3 days. His vital signs at admission revealed a temperature of 40.3°C and a demand for nasal oxygen of 3 L/min. For rheumatoid arthritis, he was taking methotrexate, iguratimod, and prednisone orally. Physical examination revealed prominent swelling of both eyelids with blood-tinged discharge on the conjunctiva. Superficial lymph nodes in the head and neck were not palpable. Respiratory sounds were clear, and crackles were not heard. No joint deformities, rashes, or lower leg edema were observed. Other abnormalities were not identified on physical examination. Contrast-enhanced computed tomography (CT) of the head showed thickening and enhanced signals of both conjunctivae, suggesting findings of blepharoconjunctivitis and cellulitis (Figure 1A,B). Chest x-ray revealed diffuse ground-glass

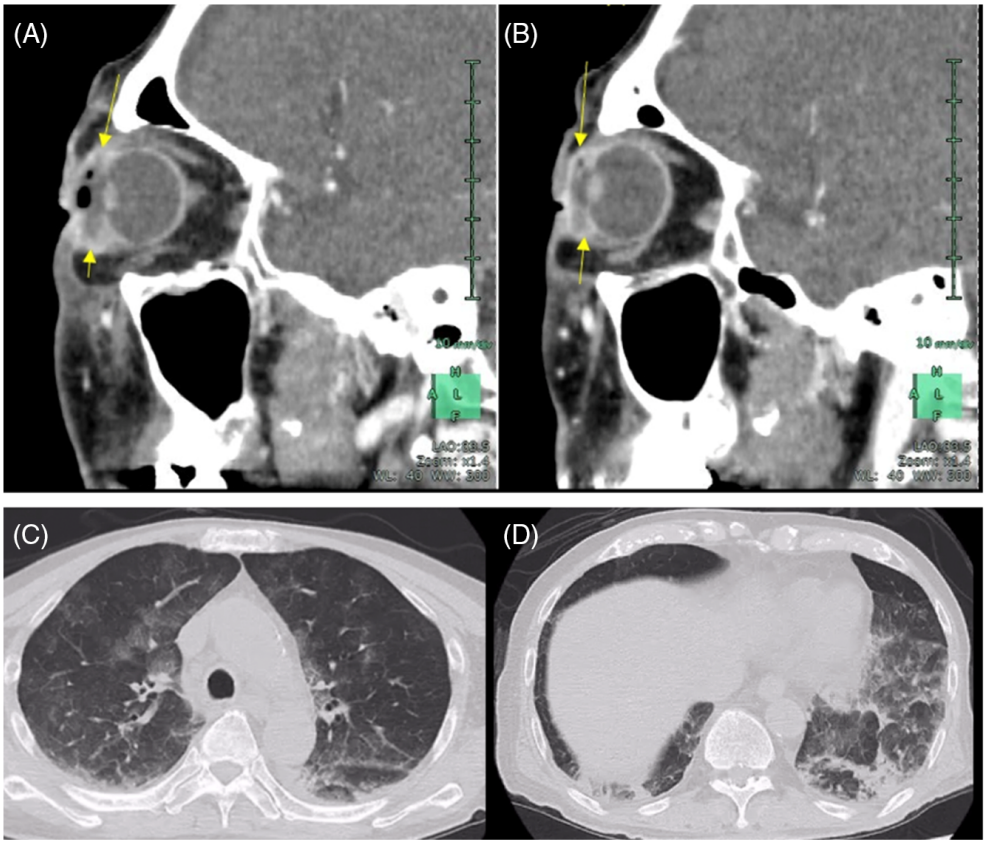

FIGURE 1 In both (A) left and (B) right eye sockets, contrast-enhanced CT revealed thickening of the conjunctiva of the eyeballs with enhanced contrast effect (arrows), suggesting bilateral periorbital conjunctivitis and cellulitis. (C), (D) Non-contrast chest computed tomography (CT) showed extensive ground-glass opacities (GGO) bilaterally with partial infiltration shadows in the left lower lobe.

opacities (GGO) in both lungs, and non-contrast chest CT showed extensive ground-glass opacities bilaterally with partial infiltration shadows in the left lower lobe (Figure 1C,D). Urinalysis revealed 2+ occult blood and proteinuria. The complete blood count demonstrated a white blood cell count of 12,100/μL (Neut, 72.5%); haemoglobin level, 11.9 g/dL; and platelet count, 161,000/μL. Biochemical examinations indicated an aspartate aminotransferase (AST) level of 44 IU/L (normal range, 13–30 IU/L), alanine aminotransferase (ALT) level of 23 IU/L (normal range, 6–27 IU/L), lactate dehydrogenase level of 391 IU/L (normal range, 124–222 IU/L), and C-reactive protein (CRP) level of 12.2 mg/dL (normal range, 0–0.3 mg/dL). Elevated levels of KL-6 (713 U/mL, normal range: 0–500 U/mL) and MMP-3 (374.9 ng/mL, normal range: 37–121 ng/mL) were observed. However, specific markers for connective tissue diseases were within normal limits: antinuclear antibodies were less than 40 times (normal range, <40 times), rheumatoid factor (RF) quantification was 5.9 IU/mL (normal range, <15 IU/mL), and levels of anti-CCP antibodies, anti-SS-A antibodies, anti-SS-B antibodies, anti-Sm antibodies, anti-dsDNA IgG antibodies, anti-RNAPIII antibodies, anti-Scl-70 antibodies, anti-Jo-1 antibodies, anti-GBM antibodies, anti-ARS antibodies, anti-Mi-2 antibodies, anti-MDA5 antibodies, PR3-ANCA, and MPO-ANCA were all within their respective normal ranges. Infection markers were negative, including Legionella, Streptococcus pneumoniae, viruses, and fungi. However, Methicillin-resistant Staphylococcus aureus (MRSA) was detected in conjunctival swab and sputum cultures. The differential diagnoses included rheumatoid lung, contagious pneumonia, and drug-induced pneumonia. Treatment was initiated by discontinuing suspected drugs, such as methotrexate, and administering broad-spectrum antibiotics, including meropenem, levofloxacin, and vancomycin, in combination with high-dose methylprednisolone. After the 9th day of hospitalization, the patient showed signs of improvement, temporarily able to be weaned off oxygen, and bilateral periorbital cellulitis also improved. However, the patient's oxygenation rapidly deteriorated on the 13th day of hospitalization, leading to his demise. Autopsy revealed the presence of fibrin balls and organizing pneumonia within the alveolar spaces (Figure 2A,B). No hyaline membrane formation, lymphoid follicles, or other abnormalities were observed. The diagnosis was confirmed as AFOP. As for findings other than the lungs, bilateral MRSA cellulitis, diabetes mellitus, and old myocardial infarction were observed. However, there were no findings related to the cause of death other than AFOP.

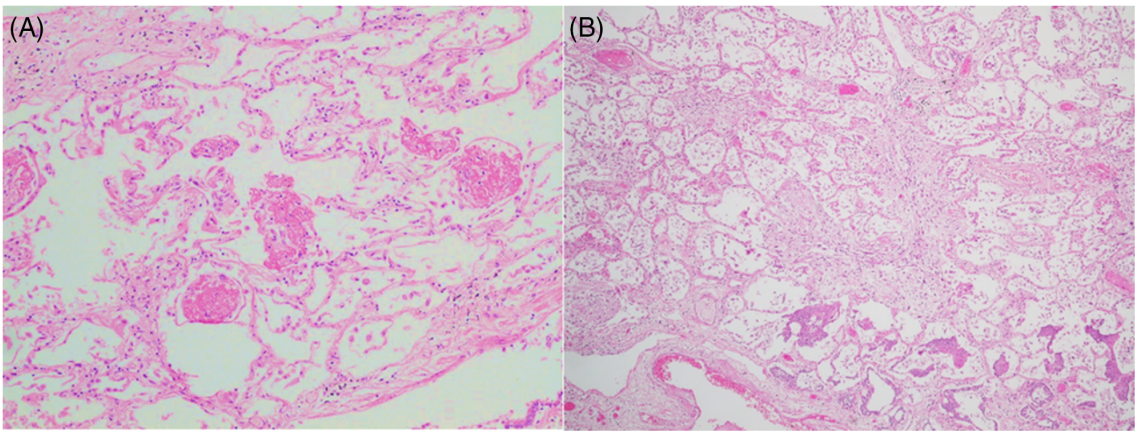

FIGURE 2 Histology of the lungs revealed the presence of fibrin balls (A) and organizing pneumonia (B) within the alveolar spaces. No hyaline membrane formation, lymphoid follicles, or other abnormalities were observed.

DISCUSSION

We reported a case in which, despite interventions involving steroid and antibiotic treatments, the patient rapidly deteriorated and died due to respiratory failure. Autopsy confirmed the diagnosis of AFOP. This case exhibited an acute course, as outlined by Beasley et al., who described two distinct AFOP progression patterns: an acute form characterized by rapid progression leading to death and a subacute form associated with eventual recovery.[1] A comprehensive literature review of 29 AFOP cases revealed that 19 presented with the subacute form, while the acute form demonstrated greater severity.[5] Notably, Beasley et al. reported that, out of 17 AFOP cases, 9 succumbed to the disease, with the duration from symptom onset to death ranging from 6 to 36 days, averaging 29 days.[1] In this case, the patient succumbed within 20 days from the onset of bilateral periorbital edema.

While the exact mechanisms are not well understood, AFOP development may be idiopathic or associated with an underlying aetiology such as collagen vascular disease, occupational exposure, drug reaction, hematologic disease, or infection.[2,3] In a paper reviewing 150 cases from 81 articles reported as AFOP between 2002 and 2019, the most common CT pattern in both idiopathic and secondary AFOP was bilateral consolidation.[2] However, secondary AFOP, in comparison to idiopathic cases, exhibited a higher frequency of GGO and was associated with a reported higher incidence of poor prognosis.[2] In this case, the CT findings also revealed bilateral GGO and the onset of AFOP is believed to be associated with multiple factors. Considering that the underlying rheumatoid arthritis was well-controlled and medications, including methotrexate, had been taken for over a year, it is plausible to consider that bilateral periorbital MRSA cellulitis, which was present concurrently with pneumonia onset, might have been more involved in the development of AFOP than rheumatoid arthritis or medications.

In conclusion, we reported a case of acute-type AFOP following periorbital MRSA cellulitis in a patient with underlying rheumatoid arthritis. Cases presenting rapidly progressing ground-glass and infiltrative opacities warrant the consideration of AFOP in the differential diagnosis, and further research is needed to explore the effective treatments.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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