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Wegener's granulomatosis – exacerbation with pulmonary manifestation

Wegener's granulomatosis disease of unknown cause (7), is generally considered a hypersensitivity disorder with granulomatosus inflammation and necrotizing small vessel vasculitis (3,10).. It is the most frequent pulmonary granulomatosis coupled with vasculitis. It was described by Kling in 1931 as *polyarthritis nodosa*, later as a separate disorder by Wegener in 1936.

CASE DESCRIPTION

We describe a case of a female patient who was admitted to the Tuberculosis Department of the Regional Hospital in Chełm. The patient was diagnosed with WG three years prior to admission due to nasolaryngeal ulcerations, fistula, and destruction of paranasal wall of the left orbit, confirmed with histopathological examination. She had been suffering from mild fever, productive of purulent cough, episodes of haemoptysis, dyspnoea and chest pain for two weeks prior to admission. On the day of admission she presented in poor general condition, with severe dyspnoea, haemoptysis, fever and chest pain. Laboratory tests revealed ESR 97/112, mild normochromic anaemia and low partial oxygen pressure in blood gases. Chest X-rays showed massive pulmonary manifestation (Fig. 1).

The patient underwent multidrug antibiotic therapy with only slight improvement of general condition, relief of dyspnoea and fever. There was no radiological improvement. The sputum examination proved negative for acid-resistant mycobaceria and atypical cells. The sputum culture was also negative. The therapy of oral cyclophosphamide (2 mg/kg/day) administered initially intravenously, followed by oral prednisolone (1 mg/kg/day) was commenced. The first two months of intensive treatment brought about the improvement of general status, regression of symptoms and partial radiological remission (Fig. 2).

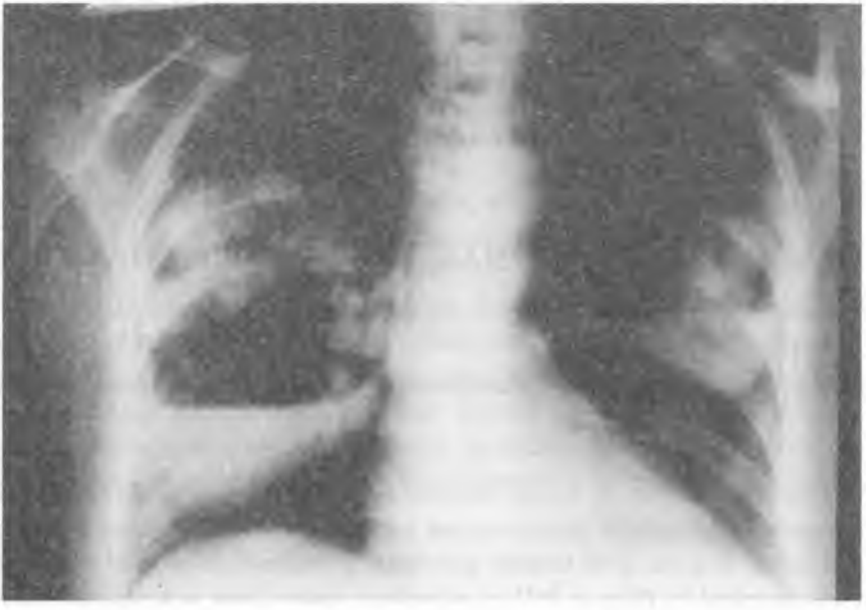

Fig. 1. An extensive cavity in the right lung, located from the 2nd to 5th rib, with a level of fluid and round opacities in the upper part. In the left lung there is a thin-walled cavity with a level of fluid and irregular opacity in the middle

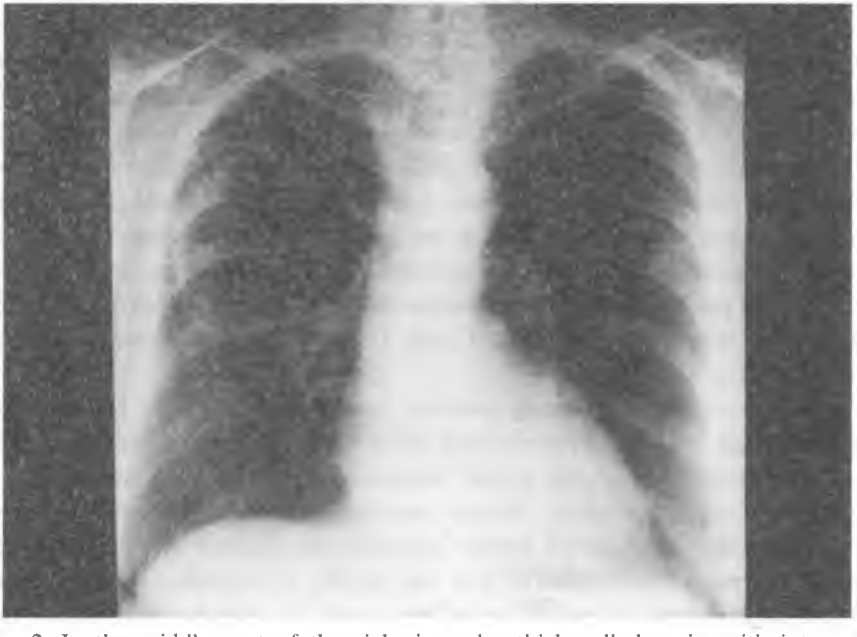

Fig. 2. In the middle part of the right irregular thick-walled cavity with intranasal septa. In the left lung there is a thin-walled cavity 4 cm in diameter

After the remission the patient continued maintenance treatment and was monitored closely also for other WG manifestations for 15 months. The patient is now well

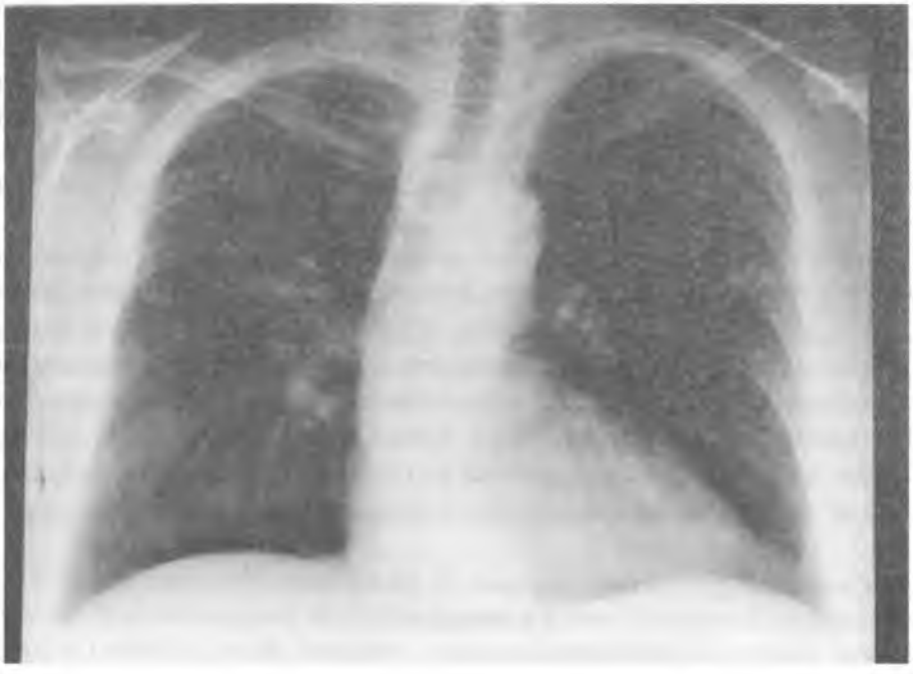

Fig. 3. Fascicular fibrous lesions in the middle parts of both lungs

and under observation on outpatient basis without any signs of progression of the disease. Chest X-ray is unremarkable (Fig. 3).

DISCUSSION

WG is characterized by three main symptoms: 1) Upper respiratory tract granulomatosis, 2) Focal nephrosis, 3) Pulmonary granulomatosis. It typically involves the upper and lower respiratory tracts, paranasal sinuses and kidneys, resulting in glomerulonephritis (9). Pulmonary involvement is present in 95% of cases, with cough, haemoptysis and dyspnoea being most common (1, 2). However, pulmonary involvement may by asymptomatic. The most common pulmonary abnormality consists of large cavitary nodules, that may by transient and recurrent. Cavitation implies necrosis with expulsion of the necrotic material via an airway. A nodule containing an air-filled necrotic region is termed cavitary. Cavitary nodules have thicker and more irregular walls than lung cyst or bullae (5). Pulmonary vasculitis with haemorrhage can also be seen (4). Involve-

ment of the trachea can result in airway narrowing and obstruction in as many as 15% of patients, which is associated with thickening and irregularity of the airway wall as shown on CT (6).

Clinically, morbidity and mortality is more often related to glomerulonephritis and renal insufficiency than to bronchopolmonary disease.

The main criteria for WG diagnosis are: 1) nasal and oral cavity ulceration with purulent and/or haemorrhagic discharge, 2) radiological findings in the chest examination, 3) urinary sedminent abnormalities (microhaematuria, erythrocyte casts), 4) histopathological changes: granulomatous inflation of arterial walls and/or perivascular tissue (8).

The diagnostic process of WG causes numerous problems as clinical symptoms and laboratory tests are not decisive and may mimic inflammatory or neoplasmatic disease.

The diagnosis is based on clinical symptoms, histopathological findings and the presence of ANCA antibodies (11). Laboratory tests are not pathognomonic – high ESR, normochromic anaemia, the presence of rheumatoid factor and higher than normal levels of serum IgE and IgA are the most frequent abnormalities of the diseases activity an early diagnosis of the recurrence.

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SUMMARY

There was described a case of a 56-year-old woman with lesions in chest X-ray. Three years before she was treated in the Department of Laryngology of Medical University of Lublin because of ulcerations of nasopharynx. There was established diagnosis of Wegener's granulomatosis.

Ziarniniakowatość Wegenera – przypadek nawrotu choroby z manifestacją płucną

Opisano przypadek 56-letniej kobiety ze zmianami w obrazie radiologicznym klatki piersiowej. Trzy lata wcześniej chora była leczona w Klinice Laryngologii Akademii Medycznej w Lublinie z powodu owrzodzeń w obrębie nosogardzieli. Rozpoznano wówczas ziarniniakowatość Wegenera.