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K.O. Lutsyshyn[1], A.I. Zhemela[2], L.A. Hryshchuk[2], I.Ya. Hospodarskyy[2], T.V. Boyko[2], O.M. Slyzka[1], S.O. Bilyk[1], M.O. Vynnychuk[1]

[1] Ternopil Regional Phthisiopulmonological Medical Center, Ternopil, Ukraine

[2] I. Horbachevsky Ternopil National Medical University, Ternopil, Ukraine

Pulmonary Langerhans Cell Histiocytosis (Clinical Case)

We describe a clinical case of pulmonary histiocytosis from Langerhans cells.

Patient P. is 57 years old, lives in a village. The disease began in October 2022 with complaints of morning cough, shortness of breath during physical activity. Complaints persisted for about 6 months and increased in dynamics. Over the past 3 months, he has lost 3 kg. He sought medical care from his family doctor, where on 07.04.2023 a computed tomography scan of the chest was performed and severe pathological changes in the lungs were detected. The patient was referred to a pulmonologist for consultation, and after further examination was hospitalized in the pulmonology department of the Ternopil Regional Phthisiopulmonology Medical Center. His medical history revealed that he had been in contact with pigeons for about 50 years. He was engaged in repair work and did not use respiratory protection.

Additional examination methods were performed. Computed tomography of the chest cavity dated 07.04.2023, conclusion — diffuse interstitial changes in the lung parenchyma. Bilateral multisegmental pulmonary nodules and thin-walled pulmonary cysts. Bronchoscopy and pathological examination were performed. The patient was consulted by a cardiologist. The clinical diagnosis was «Pulmonary Langerhans cell histiocytosis, active phase, first detected. «Honeycomb lungs». Bilateral diffuse pneumofibrosis. Chronic pulmonary heart disease. Metabolic cardiomyopathy. Transient extrasystole. Heart failure, stage I. Incomplete blockade of the right leg of the bundle of his». Treatment was prescribed according to the clinical guidelines. On 12.05.2023 he was discharged for outpatient treatment with improvement of his general condition.

In December 2023, the patient completed the course of treatment and began to notice an increase in shortness of breath with minimal physical activity, dry cough, and therefore on January 16, 2024, the patient consulted a pulmonologist, where, after further examination, he was hospitalized in the pulmonology department. After the examination the patient was diagnosed with: Chronic Obstructive Pulmonary Disease (COPD) stage II, group E, fase of infectious exacerbation. Pulmonary Langerhans cell histiocytosis, sluggishly progressive course, active phase. «Honeycomb lungs». Bilateral diffuse pneumofibrosis, coronary artery disease. Cardiosclerosis. Aortosclerosis. Incomplete blockade of the right pedicle of the bundle of His. Chronic pulmonary heart disease. Heart failure, stage I, functional class I.

It has been established that pulmonary Langerhans cell histiocytosis is a systemic pathology that in most cases leads to chronicity of the process, damage to the bronchopulmonary system (in this case, the onset of COPD); cardiovascular system (development of coronary heart disease), which requires constant pharmacological treatment and medical monitoring. In the case of a typical CT picture (widespread cystic lung disease with cysts of varying sizes and shapes with relative sparing of the lung bases and small solid nodules), a patient should be guided by a general practitioner (family doctor) to the pulmonologist to confirm or exclude pulmonary Langerhans cell histiocytosis. This allows for early diagnosis and treatment.

Keywords

Pulmonary Langerhans cell histiocytosis, adult.

Langerhans cell histiocytosis (LCH) is a rare systemic disorder characterized by the accumulation of CD1a+/Langerin+ LCH cells and wide-ranging organ involvement. Langerhans cell histiocytosis was formerly referred to as histiocytosis X, until it was renamed in 1987. Langerhans cell histiocytosis β was named for its morphological similarity to skin Langerhans cells. Studies have shown that LCH cells originate from myeloid dendritic cells rather than skin Langerhans cells. There has been significant debate regarding whether LCH should be defined as an immune disorder or a neoplasm. A breakthrough in understanding the pathogenesis of LCH occurred in 2010 when a gain-of-function mutation in BRAF (V600E) was identified in more than half of LCH patient samples. Studies have since reported that 100 % of LCH cases show ERK phosphorylation, indicating that LCH is likely to be a clonally expanding myeloid neoplasm. Langerhans cell histiocytosis is now defined as an inflammatory myeloid neoplasm in the revised 2016 Histiocyte Society classification. Randomized trials and novel approaches have led to improved outcomes for pediatric patients, but no well-defined treatments for adult patients have been developed to date. Although LCH is not fatal in all cases, delayed diagnosis or treatment can result in serious impairment of organ function and decreased quality of life. This study summarizes recent advances in the pathophysiology and treatment of adult LCH, to raise awareness of this «orphan disease» [3, 9, 16, 17, 19].

The authors found that pulmonary Langerhans cell (LC) histiocytosis (PLCH) has an unknown cause and is a rare neoplastic disorder characterized by the infiltration of the lungs and various organs by bone marrow-derived Langerhans cells with an accompanying strong inflammatory response. These cells carry somatic mutations of BRAF gene and/or NRAS, KRAS, and MAP2K1 genes, which cause activation of the mitogen-activated protein kinase (MAPK)/extracellular signal-regulated kinase (ERK) signaling pathway. PLCH occurs predominantly in young smokers, without gender predominance. Lungs might be involved as an isolated organ or as part of a multiorgan disease. High-resolution computed chest tomography plays an outstanding role in PLCH diagnosis. The typical radiological picture of PLCH is the presence of small intralobular nodules, «tree-in-bud» opacities, cavitated nodules, and thin- and thick-walled cysts, frequently confluent. Histological examination of the lesion and demonstration of characteristic eosinophilic granulomas with the presence of LCs that display antigen CD1a or CD207 in immunohistochemistry are required for definite diagnosis. Smoking cessation is the most important recommendation for PLCH patients, but treatment of progressive PLCH and multisystem disease is based on chemotherapy. Recently, new targeted therapies have been implemented [12, 13].

Several articles report the cases that presented as progressive PLCH and multisystem disease following COVID-19 infection, EBV- and HCV-infections. The COVID-19 pandemic has brought the state of impossibility in the management due to little knowledge about its etiopathogenesis; therefore, the diagnosis holds the utmost importance as management differs in both these conditions [2, 12, 14].

Some authors consider that granulomatous lung diseases are a heterogeneous group of disorders that have a wide spectrum of pathologies with variable clinical manifestations and outcomes. Precise clinical evaluation, laboratory testing, pulmonary function testing, radiological imaging including high-resolution computed tomography and often histopathological assessment contribute to making a confident diagnosis of granulomatous lung diseases. Differential diagnosis is challenging, and includes both infectious (mycobacteria and fungi) and non-infectious lung diseases (sarcoidosis, necrotising sarcoid granulomatosis, hypersensitivity pneumonitis, hot tub lung, berylliosis, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, rheumatoid nodules, talc granulomatosis, Langerhans cell histiocytosis and bronchocentric granulomatosis). Bronchoalveolar lavage, endobronchial ultrasound-guided transbronchial needle aspiration, transbronchial cryobiopsy, positron emission tomography and genetic evaluation are potential candidates to improve the diagnostic accuracy for granulomatous lung diseases. As granuloma alone is a non-specific histopathological finding, a multidisciplinary approach is important for a confident diagnosis [10].

It was established that LCH can affect children and adults with a wide variety of clinical manifestations, including unifocal, single-system multifocal, single-system pulmonary (smoking-associated), or multisystem disease. The existing paradigms in the management of LCH in adults are mostly derived from the pediatric literature. Over the last decade, the discovery of clonality and MAPK-ERK pathway mutations in most cases has led to the recognition of LCH as a hematopoietic neoplasm, opening the doors for treatment with targeted therapies. These advances have necessitated an update of the existing recommendations for the diagnosis and treatment of LCH in adults. This document presents consensus recommendations that resulted from the discussions at the annual Histiocyte Society meeting in 2019, encompassing clinical features, classifica-

tion, diagnostic criteria, treatment algorithm, and response assessment for adults with LCH.

The final diagnosis is based on the clinical picture and the detection of Langerhans cells in the examined material. The probable diagnosis is based on the clinical and X-ray image. A CT scan of the lungs may be indicated for patients with abnormal chest X-rays or pulmonary symptoms. High-resolution CT scans may show evidence of pulmonary LCH when the chest X-ray is normal.

The recommendations favor the use of 18F-Fluorodeoxyglucose positron emission tomography-based imaging for staging and response assessment in the majority of cases. Most adults with unifocal disease may be cured by local therapies, while the first-line treatment for single-system pulmonary LCH remains smoking cessation. Among patients not amenable or unresponsive to these treatments and/or those with multifocal and multisystem disease, systemic treatments are recommended. Preferred systemic treatments in adults with LCH include cladribine or cytarabine, with the emerging role of targeted (BRAF and MEK inhibitor) therapies. Despite documented responses to treatments, many patients struggle with a high symptom burden from pain, fatigue, and mood disorders that should be acknowledged and managed appropriately [1, 4, 5, 11].

A number of clinical cases of pulmonary Langerhans cell histiocytosis were presented in the available literature.

A case of Langerhans cell histiocytosis presenting as an obstructing tracheal lesion in a 55-year-old woman was described. Following complete resection of the lesion via flexible bronchoscopy, full recovery was achieved. This case represents a unique cause of tracheal obstruction, as well as an unreported manifestation of pulmonary Langerhans cell histiocytosis [20].

During a thoracic computed tomography (CT) scan, a 36-year-old male was diagnosed with a solitary oval pulmonary mixed ground-glass nodule in the right upper lobe of the lung. The edge of the nodule was well-defined, and its largest axial size was approximately 1.1 × 0.9 cm. This nodule was slightly lobulated, but not obviously spiculated. Solid components, micro-cystic lucency shadow, small high-density rings and tiny vascular branches were all visible in the nodule. During hospitalization, a technetium 99 m methylene diphosphonate (Tc-99 m MDP) bone scan was performed, which showed a skeletal focus with abnormal uptake in the left iliac. A pulmonary lobectomy of the right upper lobe of the lung by video-assisted thoracoscopy was performed. In post-operative pathological photomicrographs, proliferative Langerhans' cells, eosinophils and lymphocytes were found. Immunohistochemistry showed that the expression of S-100 protein, CD1a, and CD68 antigen all stained positive. Since LCH that is also associated with isolated mixed ground-glass nodules is relatively rare, such a multi-systemic LCH case as identified herein, is reported [18].

Solitary pulmonary nodules are an uncommon manifestation of pulmonary PLCH. A case of a 45-year-old male cigarette smoker who presented with an asymptomatic solitary pulmonary nodule that showed histologic and immunophenotypic characteristics of PLCH was. Twenty-one years after excision of the nodule, at the age of 66 years, he is asymptomatic with a new contralateral lung nodule but no evidence of interstitial disease. The new nodule has remained unchanged after 36 months of observation. This case affirms that PLCH can occasionally cause solitary lesions, which should not be interpreted as a harbinger of interstitial lung disease. Isolated PLCH should be included in the differential diagnosis of unusual solitary pulmonary nodules [6].

The authors encountered two rare cases of pulmonary eosinophilic granuloma with multiple nodular shadows in both lungs. The patient in case 1 was a 54-year-old man complaining of dry cough and chest pain. He had smoked 20 cigarettes a day for 36 years. The patient in case 2 was a 37-year-old woman complaining of dry cough. She had smoked 15 cigarettes a day for 20 years. Chest radiography and CT revealed multiple nodular shadows in both lungs. Diagnosis was made by open lung biopsy in case 1 and by percutaneous lung biopsy in case 2. After smoking cessation, symptoms improved markedly and the shadows in the chest radiographs and CT disappeared. In cases of bilateral multiple nodular shadows, other than metastatic lung tumor cases, pulmonary eosinophilic granuloma should be considered [8].

A retrospective analysis of the clinical and follow-up data of 15 hospitalized PLCH cases from September 2012 to June 2021 at the Second Xiangya Hospital of Central South University was performed. The following results were obtained. The age of 15 patients (9 men and 6 women, with a sex ratio of 3 to 2) was 21—52 (median 33) years. Among them, 8 had a history of smoking and 5 experienced spontaneous pneumothorax during disease course. There were 3 patients with single system PLCH and 12 patients with multi-system PLCH, including 7 patients with pituitary involvement, 7 patients with lymph node involvement, 6 patients with bone involvement, 5 patients with liver involvement, 2 patients with skin involvement, 2 patients with thyroid involvement, and 1 patient with thymus involvement. The clinical manifestations were varied

but non-specific. Respiratory symptoms mainly included dry cough, sputum expectoration, chest pain, etc. Constitutional symptoms included fever and weight loss. Patients with multi-system involvement experienced symptoms such as polyuria-polydipsia, bone pain, and skin rash. All patients were confirmed by pathology, including 6 by lung biopsy, 3 by bone biopsy, 2 by lymph node biopsy, and 4 by liver, skin, suprasternal fossa tumor, or pituitary stalk biopsy. The most common CT findings from this cohort of patients were nodules and/or cysts and nodular and cystic shadows were found in 7 patients. Three patients presented simple multiple cystic shadows, 3 patients presented multiple nodules, and 2 patients presented with single nodules and mass shadows. Pulmonary function tests were performed in 4 patients, ventilation dysfunction was observed in 2 patients at the first visit. Pulmonary diffusion function tests were performed in 4 patients and showed a decrease in 3 patients. Smoking cessation was recommended to PLCH patients with smoking history. Ten patients received chemotherapy, while 2 patients received oral glucocorticoid therapy. Among the 11 patients with the long-term follow-up, 9 were in stable condition.

So, PLCH is a neoplastic disease closely related to smoking. The clinical manifestations and laboratory examination are not specific. Pneumothorax could be the first symptom which is highly suggestive of the disease. Definitive diagnosis relies on histology. There is no unified treatment plan for PLCH, and individualized treatment should be carried out according to organ involvement. Early smoking cessation is essential. Chemotherapy is the main treatment for rapidly progressing PLCH involving multiple organs. All diagnosed patients should be considered for the detection of BRAFV600E gene and relevant targeted therapies have been implemented recently [7].

Objective — to familiarize practitioners and scientists with a clinical case of pulmonary Langerhans cell histiocytosis.

Clinical Case

Patient P. is 57 years old, lives in a village. The disease began in October 2022 with complaints of morning cough, shortness of breath during physical activity. The complaints persisted for about 6 months and progressively worsened.

Over the past 3 months, he has lost 3 kg. He sought medical help from his family doctor, where on 07.04.2023 a CT scan of the chest was performed, revealing severe pathological changes in the lungs. The patient was referred to a pulmonologist for consultation, and after further examination, he was hospitalized in the pulmonology department of the Ternopil Regional Phthisiopulmonology Medical Centre.

He denies contact with tuberculosis patients. His medical history shows that he has been in contact with pigeons for about 50 years. He was engaged in repair work and did not use respiratory protection. He denies contact with people who came from other countries and patients with coronavirus disease, a rapid test for COVID-19 (25.04.2023) was negative.

Complaints at the time of hospitalization: cough in the morning, shortness of breath during physical activity, weight loss. Objectively: the general condition is relatively satisfactory. Body temperature is 36.8 °C. Conscious, communicative, adequate. Correct build, satisfactory nutrition. The skin and visible mucous membranes are pale, clean. The tongue is dry and covered with white fur. The pharynx is clean, granular. Peripheral lymph nodes are not palpable. The thyroid gland is without induration. Respiratory rate 28/min, SpO_2 97 % when breathing atmospheric air. Percussion over the lungs is a clear pulmonary sound, shortened in the lower parts. Auscultation is rigid breathing. Cardiac activity is rhythmic, tones are weakened. Pulse 100 per minute (tachycardia) is rhythmic, satisfactory filling and tension. Blood pressure 145/100 mm Hg. The abdomen is soft, not painful on palpation. The liver is at the level of the right rib. The spleen is not palpable. Pasternatsky's symptom «—» on both sides. Edema on the feet. Physiological discharges are normal.

Bronchoscopy dated 28.04.2023.

Under general anesthesia, a bronchoscope was inserted into the trachea through an intubation tube. The carina is acute, slightly deformed. The bifurcation angle is not widened (Fig. 1). The bronchial tree is slightly deformed, more so in the segmental and subsegmental bronchi. The mucosa is thin, pale, sometimes with areas of hyperemia, with the presence of easily expressed pigment spots. Bronchial elasticity is preserved (Fig. 2—4). Lavage was taken for PCR, MBT and antibiogram. Biopsies were taken (no stiffness). Smears for acid-resistant bacteria and cytology were taken from the right B9. Conclusion: diffuse bilateral deforming atrophic pigmented bronchitis.

Pathological examination of 05.05.2023.

There is no epithelium in the provided material. There are connective tissue elements and single muscle fragments, mucus. There is a marked inflammatory infiltration everywhere, represented mainly by lymphocytes and macrophages. Fibrosis is observed. No signs of tumor process, markers of specific inflammation were found. Conclusion: chronic inflammatory process of nonspecific etiology.

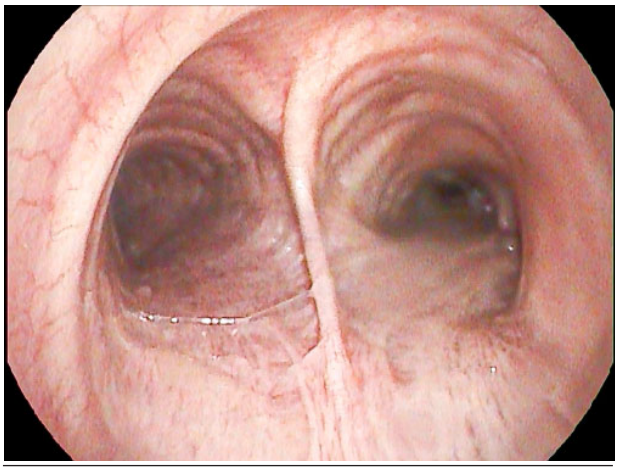

Fig. 1. **Bronchoscopy. Trachea, tracheal bifurcation**

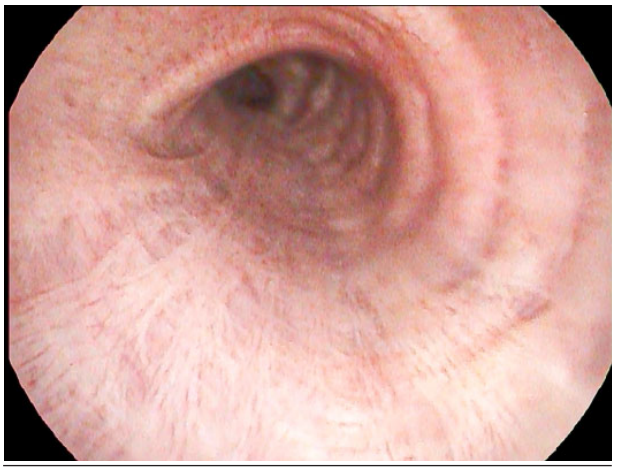

Fig. 2. **Bronchoscopy. Left main bronchus, proximal part**

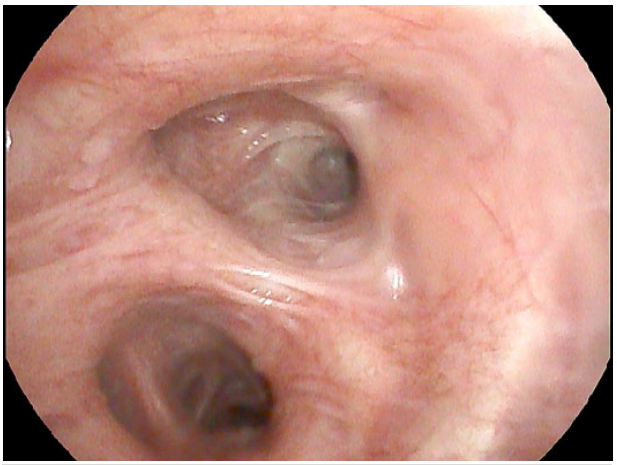

Fig. 3. **Bronchoscopy. Left main bronchus, distal part**

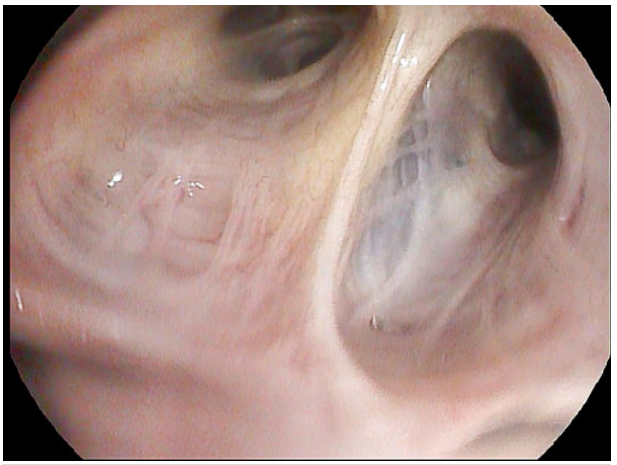

Fig. 4. **Bronchoscopy. Right upper lobe bronchus**

CT scan of the chest dated 07.04.2023 (Fig. 5). Conclusion: diffuse interstitial changes in the lung parenchyma. Bilateral polysegmental pulmonary nodules. Bilateral polysegmental thin-walled pulmonary cysts.

02.05.2023. Consultation of a cardiologist: Chronic pulmonary heart disease. Metabolic cardiomyopathy. Transient extrasystole. Heart failure, stage I. Incomplete blockade of the right leg of the bundle branch of His.

The patient was diagnosed with: Pulmonary Langerhans cells histiocytosis, active phase, first detected. «Honeycomb lungs». Bilateral diffuse pneumofibrosis. Chronic pulmonary heart disease. Metabolic cardiomyopathy. Transient extrasystole. Heart failure, stage I. Incomplete blockade of the right leg of the bundle of his.

The diagnosis has been confirmed based on clinical and CT findings. Treatment is prescribed according to clinical guidelines: Histiocyte Society Evaluation and Treatment Guidelines, 2012 [5].

The patient with improved general condition (i.e. respiratory rate 19/min, SpO_2 99 %) was discharged to continue treatment on an outpatient basis under the supervision of a family doctor, pulmonologist at the place of residence. The patient was recommended to undergo a CT scan in a month to assess the effectiveness of the prescribed treatment. The patient attended a follow-up visit on 06.06.2023. A positive clinical picture of improvement in the patient's general condition was noted. The follow-up examination revealed positive dynamics.

CT scan of the chest dated 06.06.2023 (1 month after discharge, Fig. 6). Conclusion: CT picture of chronic fibrosing lung disease. Positive dynamics of resorption.

The patient was recommended to continue the prescribed treatment and come for follow-up in 3 months, for which he applied on 04.08.2023. Clinically positive dynamics, CT scan of the chest cavity showed stabilization of the pathological process. Respiratory rate 18/min, SpO_2 99 %.

CT scan of the chest cavity dated 04.08.2023 (3 months after discharge, Fig. 7). Conclusion: CT picture of chronic fibrosing lung disease — histiocytosis, stabilization of the process.

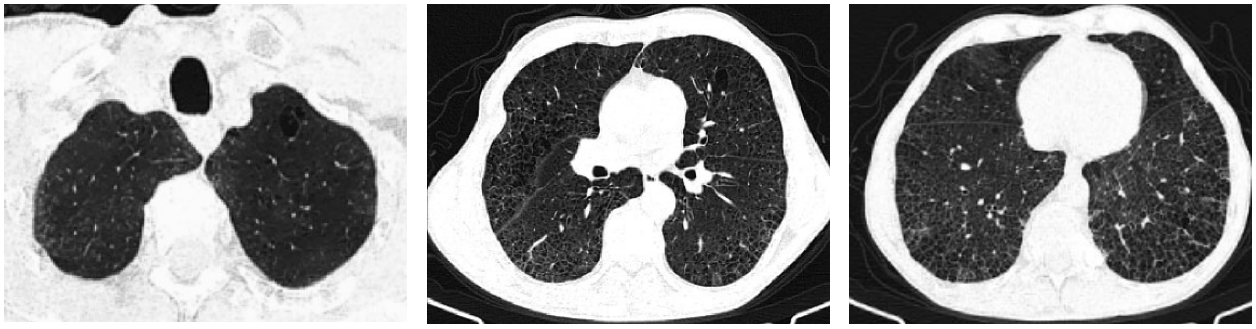

Fig. 5. **Computed tomography of the chest cavity dated 07.04.2023**

Bilaterally diffusely altered pneumatization of the lung parenchyma due to a reticular pattern of the «honeycomb lungs» type. Thin-walled pulmonary cysts up to 12 mm in size are seen bilaterally in a polysegmental manner. On the right in S3, S9, S10, on the left in S3, S10, there are rounded pulmonary nodules with clear contours up to 4 mm in size of solid structure, some with signs of calcification. Fibrous cords are seen bilaterally in the basal regions.

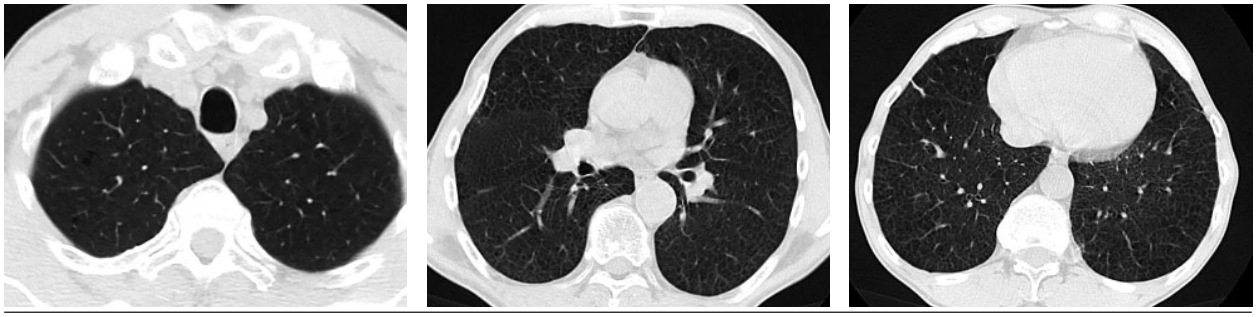

Fig. 6. **Computed tomography of the chest cavity dated 06.06.2023 (1 month after discharge)**

Pulmonary parenchyma of reduced pneumatization, fibrous-mesh deformation throughout, accentuated, compacted interlobular septa, peribronchial couplings, multiple small cystic lucencies — centrilobular emphysema, fibrous cords at the apices, frosted glass consolidation foci, several small calcifications on the right. The bronchi are patent, free, the walls are thickened. Enlarged bronchopulmonary lymph nodes are not detected. Mediastinal organs are located medially. Enlarged mediastinal lymph nodes are not detected.

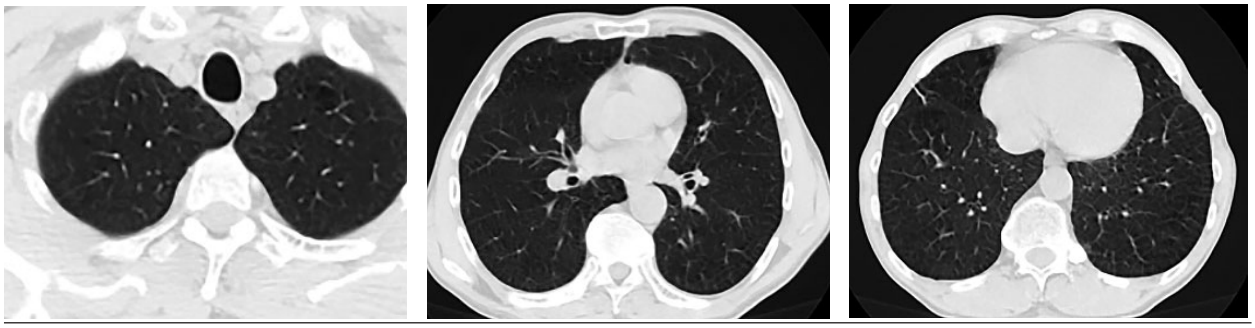

Fig. 7. **Computed tomography of the chest cavity dated 04.08.2023 (3 months after discharge)**

Pulmonary parenchyma of reduced pneumatization, fibrous-mesh deformation throughout, accentuated, compacted interlobular septa, peribronchial couplings, multiple small cystic lucencies — centrilobular emphysema (cobblestone symptom), fibrous cords at the apices, several small calcifications on the right. The bronchi are patent, free, and the walls are thickened. Enlarged bronchopulmonary lymph nodes are not detected. Mediastinal organs are located medially. Enlarged mediastinal lymph nodes are not detected.

The treatment was adjusted, and it was recommended to continue treatment on an outpatient basis according to a tapering regimen until complete discontinuation in December 2023.

In December 2023, the patient completed the course of treatment and began to notice an increase in shortness of breath with minimal physical activity, cough; therefore, on January 16, 2024, the patient consulted a pulmonologist, where, after a further examination, he was hospitalized in the pulmonology department.

CT scan of the chest from January 16, 2024 (Fig. 8). Conclusion: CT picture of chronic fibrosing lung disease — histiocytosis, stabilization of the process.

Spirometry dated January 23, 2024.

Conclusion: obstructive and restrictive changes of mild severity.

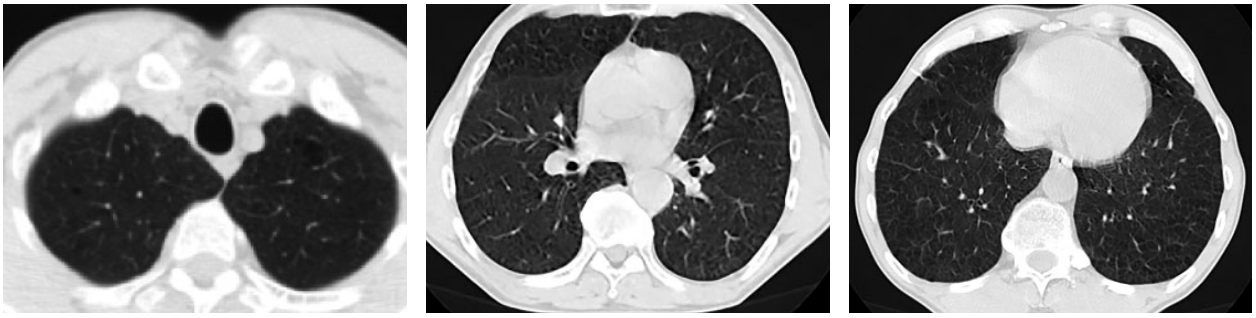

Fig. 8. **Computed tomography of the chest cavity dated January 16, 2024**

The lung parenchyma is emphysematous. Throughout, fibrotic-mesh deformation of the lung pattern due to mixed pneumosclerosis, peribronchial couplings, small cystic lucencies. Small intense foci at the apices. The bronchi are patent, free, and the walls are thickened. Enlarged bronchopulmonary lymph nodes are not detected. Mediastinal organs are located medially. Enlarged mediastinal lymph nodes are not detected.

The patient was re-consulted by a cardiologist. Treatment was corrected.

The patient was diagnosed with: COPD stage II, group E, phase of infectious exacerbation. Pulmonary Langerhans cell histiocytosis, sluggishly progressive course, active phase. «Honeycomb lungs». Bilateral diffuse pneumofibrosis, coronary artery disease. Cardiosclerosis. Aortosclerosis. Incomplete blockade of the right bundle branch of His. Chronic pulmonary heart disease. Heart failure, stage I, functional class I.

The patient was prescribed treatment for COPD according to clinical guidelines and treatment of histiocytosis from Langerhans cells with glucocorticosteroids in minimal doses, which the patient has been receiving up to now. Positive clinical dynamics and stabilization of the radiological process were noted during treatment.

Conclusions

It has been established that pulmonary Langerhans cells histiocytosis is a systemic pathology that in most cases leads to chronicity of the process, damage to the bronchopulmonary system (in this case, the onset of COPD); cardiovascular system (development of coronary heart disease), which requires constant pharmacological treatment and medical monitoring.

In the case of a typical CT picture (widespread cystic lung disease with cysts of varying sizes and shapes with relative sparing of the lung bases and small solid nodules), a patient should be referred by a general practitioner (family doctor) to the pulmonologist to confirm or exclude pulmonary Langerhans cells histiocytosis. This allows for early diagnosis and treatment.

There is no conflict of interest.
Participation of authors: concept and design of the study — K.O. Lutsyshyn, A.I. Zhemela, L.A. Hryshchuk; collection and processing of the material — K.O. Lutsyshyn, A.I. Zhemela, L.A. Hryshchuk, I.Ya. Hospodarskyy, T.V. Boyko, O.M. Slyzka, S.O. Bilyk, M.O. Vynnychuk; writing and editing — A.I. Zhemela, L.A. Hryshchuk.

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К.О. Луцишин[1], А.І. Жемела[2], Л.А. Грищук[2], І.Я. Господарський[2], Т.В. Бойко[2], О.М. Слизька[1], С.О. Білик[1], М.О. Вінничук[1]

[1] Тернопільський обласний фтизіопульмонологічний медичний центр, Тернопіль
[2] Тернопільський національний медичний університет імені І.Я. Горбачевського, Тернопіль

Легеневий гістіоцитоз клітин Лангерганса (клінічний випадок)

Описано клінічний випадок легеневого гістіоцитозу клітин Лангерганса.

Пацієнт П., 57 років, мешкає в селі. Захворювання розпочалося в жовтні 2022 р. зі скарг на кашель уранці, задишку при фізичному навантаженні. Скарги спостерігалися близько 6 міс і наростали в динаміці. За останніх 3 міс схуд на 3 кг. Звернувся по медичну допомогу до сімейного лікаря. 07.04.2023 р. проведено комп'ютерну томографію органів грудної порожнини. Виявлено виразні патологічні зміни в легенях. Пацієнта направили на консультацію до пульмонолога, а після додаткового обстеження госпіталізували в пульмонологічне відділення Тернопільського обласного фтизіопульмонологічного медичного центру. З анамнезу відомо, що пацієнт близько 50 років контактував із голубами. Він займався ремонтними роботами та не використовував засоби захисту органів дихання.

Проведено додаткові обстеження. Комп'ютерна томографія органів грудної порожнини (07.04.2023): дифузні інтерстиціальні зміни в легеневій паренхімі, двобічні мультисегментарні легеневі вузлики та тонкостінні легеневі кісти. Виконано бронхоскопію та патологоанатомічне дослідження. Пацієнт був проконсультований кардіологом. Клінічний діагноз: Легеневий гістіоцитоз клітин Лангерганса, активна фаза, вперше виявлений. «Стільникові легені». Двосторонній дифузний пневмофіброз. Хронічне легеневе серце. Метаболічна кардіоміопатія. Транзиторна екстрасистолія. Серцева недостатність I стадії. Неповна блокада правої ніжки пучка Гіса. Лікування призначено згідно з клінічними рекомендаціями. 12.05.2023 р. виписаний на амбулаторне лікування з поліпшенням загального стану.

У грудні 2023 р. пацієнт завершив курс лікування і почав відзначати підсилення задишки при мінімальних фізичних навантаженнях, сухий кашель. З цього приводу 16 січня 2024 р. пацієнт звернувся до пульмонолога. Після дообстеження був госпіталізований у пульмонологічне відділення. Після обстеження пацієнту встановлено діагноз: хронічне обструктивне захворювання легень, II стадія, група Е, фаза інфекційного загострення. Легеневий гістіоцитоз клітин Лангерганса, перебіг, що в'яло прогресує, активна фаза. «Стільникові легені». Двосторонній дифузний пневмофіброз, ішемічна хвороба серця. Кардіосклероз. Аортосклероз. Неповна блокада правої ніжки пучка Гіса. Хронічне легеневе серце. Серцева недостатність I стадії, I функціональний клас.

Таким чином, легеневий гістіоцитоз клітин Лангерганса є системною патологією, яка в більшості випадків призводить до хронізації процесу, ураження бронхолегеневої системи (в описаному випадку приєднання хронічного обструктивного захворювання легень), серцево-судинної системи (розвиток ішемічної хвороби серця), що потребує постійного фармакологічного лікування та спостереження лікуючих лікарів. У разі типової комп'ютерно-томографічної картини (поширене кістозне ураження легень із кістами різного розміру та форми з відносним збереженням основи легень і невеликими солідними вузликами) лікар загальної практики (сімейний лікар) має направити пацієнта до пульмонолога для обстеження, щоб підтвердити або заперечити легеневий гістіоцитоз клітин Лангерганса. Це дає змогу провести ранню діагностику та лікування.

Ключові слова: легеневий гістіоцитоз клітин Лангерганса, дорослий.

***Контактна інформація** / Corresponding author*

Грищук Леонід Андрійович, д. мед. н., проф. кафедри пропедевтики внутрішніх хвороб та фтизіатрії
https://orcid.org/0000-0002-2528-8843
46001, м. Тернопіль, вул. Енергетична, 25
E-mail: hrischuk@ukr.net

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