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Neurological Manifestations of Wegener's Granulomatosis - Case Report

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Abstract

BACKGROUND: Granulomatosis with polyangiitis is an inflammatory process that affects the endothelium of small and medium blood vessels. It is an autoimmune disease, which is systemic and attacks multiple organ systems. It is also called Wegener's granulomatosis (Wegener Granulomatosis), according to the German pathologist Wegener who described the first cases of granulomatous inflammation of small and medium blood vessels.

CASE PRESENTATION: This paper presents a clinical case report of a middle-aged female patient with Wegener's granulomatosis, treated at the Department of Neurology at City General Hospital 8th September. The patient's initial symptoms began nine years ago. The course of her disease is chronic with a progressive nature, followed by relapses and exacerbations. The patient was promptly diagnosed and treated with appropriate therapy, which she tolerates well. The definitive diagnosis is established by biopsy of the affected tissue and pathohistological analysis, where the presence of granulomas is detected.

CONCLUSION: This form of polyangiitis is significant for the presence of anti-neutrophil cytoplasmic antibodies (ANCA) in the patient's blood. But their absence does not exclude the disease. In terms of therapy, drugs are used that regulate the patient's immune system. We must be aware of the risk of complications and worsening of the course of the disease. This condition requires a multidisciplinary approach to treatment.

Introduction

Wegener's Granulomatosis, also known as Granulomatosis with polyangiitis, is a form of necrotizing vasculitis of small and medium-sized blood vessels. It is a rare autoimmune and multisystem disease, accompanied by granulomatous inflammation [1]. This condition is a component of a wide spectrum of disorders called anti-neutrophil cytoplasmic antibody (ANCA)-related vasculitides [2].

The classification of the modern nomenclature of systemic vasculitides was established at the 2012 Chapel Hill consensus conference [3]. Wegener's granulomatosis is characterized by a pneumo-renal syndrome associated with otorhinolaryngological and neurological manifestations.

The first case was described by a German medical student, Heinz Klinger, in 1931 [4]. Five years later, in 1936, German pathologist Friedrich Wegener described three cases of an unusual vasculitis with granulomatous inflammation involving small and medium-sized blood vessels [5]. He thus identified this disorder as a distinct and specific form of vasculitis. In 1954, Godman and Churg published a review of 22 cases, and the disease was now universally known as Wegener's Granulomatosis [6].

Neurological involvement is not uncommon in Wegener's granulomatosis, with reported frequencies ranging from 22% to 54% [7]. In most cases, neurological signs represent peripheral disease, while central nervous system (CNS) involvement associated with Wegener's granulomatosis is less common. According to the studies conducted until now, CNS involvement has been reported in 7% to 11% of patients. Slightly higher rates of CNS involvement, ranging from 8% to 18%, have been found when isolated cranial nerve palsies and paresis were also included in the spectrum of CNS-associated symptoms.

Case Presentation

A clinical case report is made of a middle-aged patient with Polyangiitis cum granulomatosis-Wegener, who was admitted, examined and treated at the Department of Neurology - 8th September City General Hospital in Skopje. Additional analyses and examinations of the patient were also performed at the Public Health Institution - University Clinic for Rheumatology due to worsening of the condition.

The patient has shown progression of the condition with relapses and exacerbations. She responds well to the prescribed therapy. The main emphasis in the paper is placed on the course of Wegener's Granulomatosis with overall symptomatology, outcome and consequences. The onset of the condition dates back to February 2015.

Personal medical history:

A 52 years old female patient, married, mother of two children and unemployed. She is a graphic designer by profession, dominantly right-handed. She is a long-term smoker, over 20 years old, smoking 2 packs a day. Does not consume alcohol. Denies allergies to food and medications.

Chief complaints:

The main difficulties that lead to a medical examination are diffuse pain, most expressed in the small joints of the hands and feet, muscle weakness, changes in skin color and constitutional symptoms such as general weakness, prostration, weight loss and localized hyperhidrosis.

Present illness:

Duration of symptomatology is 3 months, with intense pain in the hands during sleep. Two weeks after the onset of the initial symptoms, similar pain appear in the feet, which persist throughout the entire period. At the same time, fevers occur, in the absence of elevated body temperature, increased sweating (in the neck and chest area), accompanied by diffuse pain throughout the body, general weakness and prostration.

Some time ago, the patient has been noticing muscle weakness in her hands (dropping objects from her hand, fine finger manipulation, difficulty using personal hygiene items). The onset of the above-mentioned symptoms was not preceded by an infectious episode or fever. The patient has not used any therapy. Data on weight loss (10-15 kg) over a period of 3 months have been reported. One month before the current examination, the patient noticed changes in coloration (discoloration) of the skin in her palms and feet. At first, the changes were with intensive redness, then bluishness and finally pallor, accompanied by the so-called occasional "mottling" of the skin.

In the interim period, muscle weakness is emphasized, which significantly compromises the patient's daily functioning. In addition, the condition immediately before admission to hospital treatment has worsened, accompanied by pain and paresis in the left half of the face, most expressed in the upper jaw area, which is preceded by severe pain in the left ear.

Radiographic examinations of the hands and cervical spine were performed. The X-ray characteristics of the hands showed initial degenerative changes, and of the cervical spine, discopathy at the level of fourth to sixth cervical vertebra (C4-C6), with advanced spondylosis. Outpatient laboratory examinations were performed, with the finding of CRP 64 mg/L (c-reactive protein) and RF 712 IU/ml (rheumatic factor).

Family history: The patient denies diseases of interest to her condition.

Psychomotor development: The patient has a regular early psychomotor development. Fifth child of five children.

Past diseases: Depressive syndrome, Lumbago.

Allergology history: The patient denies allergies to food and drugs.

Socio-epidemiological status: Information was obtained that the patient was in contact with a child, in whom Escherichia coli was isolated by a swab from nose and throat.

Pharmacological history: The patient is on tablet therapy that includes:

- Tbl. Naclofen a 75 mg 1x1, as needed 2x1
- Tbl. Asentra a 100 mg 1x1 in the evening
- Tbl. Helex a 0.5 mg 1x1

Status Praesens

General status: On examination, the patient is conscious and afebrile. She assumes an active position in bed. On the skin of anterior abdominal wall, chest and dorsal side of the hands, hyperpigmented oval changes are present, with a diameter of about 5-6 mm, which are in regression phase. In area of the right elbow, a change with a diameter of about 3 cm is present. Abdomen at the level of the chest, palpatory soft and painless, without signs of organomegaly, with preserved peristalsis on auscultation.

Chest: On auscultation of the lungs, sharp vesicular breathing is noted without associated pathological murmur.

Cardiovascular system: The heart action is rhythmic, with well audible tones, blood pressure

120/70 mm/Hg, pulse 70 beats/minute.

Extremities: The extremities are without visible deformities, with swelling of the wrist and ankle bilaterally.

Mental status: The patient is conscious, properly oriented in all directions (time, space, people), with a suppressed basic mood.

Neurological status: Cranial nerves are well innervated. Generalized, predominantly distal, hypotonic, hypotrophic muscle weakness of moderate-severe degree of the upper extremities and moderate degree of the lower extremities is noted. MTR (muscle-tendon reflexes) are obtained with regular and symmetrical responses, pathological reflexes are not present. Hypoesthesia and paresthesias of peripheral type of distribution, on the hands to the level of the palms, on the legs to the level of the feet. Stretch tests of the peripheral nerves - Lasseque et Bickeless - are bilaterally negative. Gait is paraparetic with abduction of the right leg. Sphincters are well controlled.

One month later, the patient returned for a medical examination with a Neurologist due to worsening of her condition. Main complaints are severe pain in the hands and feet, as well as the shoulder. Initially, the patient was diagnosed with sensorimotor polyneuropathy. The etiological thinking is focused on a vasculitic or paraneoplastic condition. BAB test for Brucellosis was performed, which was negative. The result from the Institute of Pulmonary Diseases and Tuberculosis from analyzed sputum is negative. The gynecological examination is unremarkable. Serological results indicating vasculitis are: RF 32.9; CRP 58.2; AST 35.5 IU/ml. The antibodies: anti CCP>268; anti-dsDNA, anti SSA, anti SSB, ACA, anti Jo- are negative. The patient was examined by a rheumatologist, because rheumatological complaints could be part of a paraneoplastic syndrome. Therapy with Decortin 10 mg in the morning was prescribed. Severe pain in the joints, hands and feet persisted throughout, with significant functional compromise, which resulted in repeated admission to neurological department for therapeutic purposes.

From the new general status of the patient, it was concluded that she is conscious and afebrile, but this time with a passive position in bed, with impression of a moderately-severe patient. On the skin of the anterior abdominal wall, chest and dorsal side of the hands, hyperpigmented oval changes are still present, which are in the regression phase (Figure 1 and Figure 2). The livid change on the right elbow measuring 3 cm in diameter is still present (Figure 3). Abdomen at the level of the chest, palpatory soft and painless, without signs of organomegaly, with preserved peristalsis on auscultation. In terms of mental status, the patient is conscious, contactable, properly autopsychically and allopsychically oriented, with suppressed basic mood and reduced volitional-instinctive dynamisms. Regarding the neurological status, there is generalized, predominantly distal, hypotonic and hypotrophic muscle weakness of the upper and lower extremities. MTRs are obtained with regular responses, pathological reflexes - bilateral “drop feet”. Hypoesthesia and paresthesias of peripheral type of distribution, on the arms to the level of the hands, on the legs to the level of the feet. The gait is paraparetic, difficult on the heels and toes. Sphincters are properly controlled.

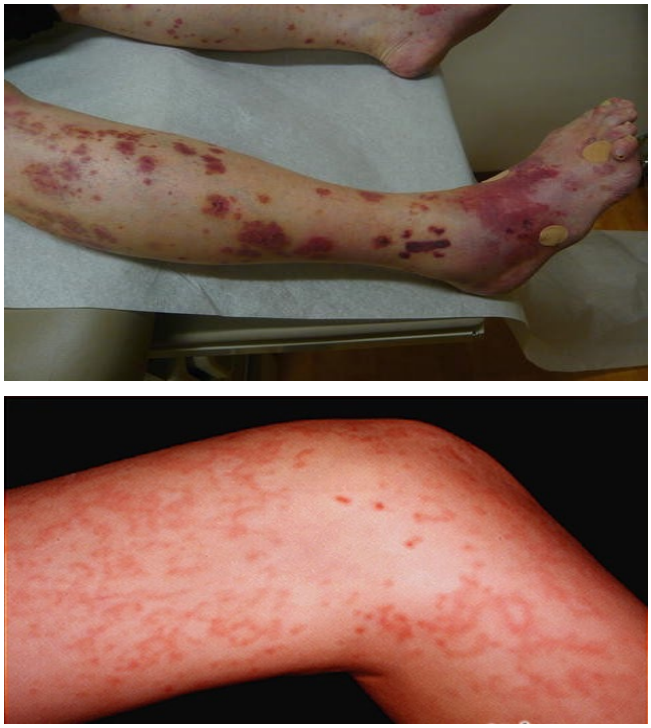

Figure 1: Skin changes

After the examinations, analyses and tests, the patient was discharged from Neurology Department at the 8th September General Hospital with the finding of Polyneuropathia senso-motoria obs Wegener granulomatosis. The patient needs to undergo additional tests for antibodies c-ANCA, p-ANCA, sedimentation rate (SE), complete blood count (CBC), urine, CRP and lung X-ray in one month (with regular controls after 3 months, 6 months and one year). A control with a neurologist and a pulmonologist is scheduled after one month of discharge, as well as a consultation with a hematologist. The patient has been prescribed the following therapy:

-Tbl. Prednisone a 50 mg 1x1 in the morning, after a meal, between 8 and 9 am- 10 days, then 40 mg for 1 month, then 30 mg for 2 months, then 20 mg 1x1 for one year.

- Caps. Omeprazole a 20 mg 1x1 in the morning

- Tabl. Evitol a 400 mg 1x1

- Tabl. Trimethoprim/sulphametoxasole a 480 mg 2x1 tablet every 3 days for 6 months.

- Sol. Vigantol (Vit. D3) 5 drops per day

- Tabl. Enalapril a 5 mg 2x1

- Tabl. Sertraline a 50 mg 1x1 in the morning

- Tabl. Alprazolam a 0.25 mg 3x1

A dietary regime with restriction of salt and sugar was prescribed.

The patient's pathology continued one year later, at the PHI Clinical Center for Rheumatology. In 2016, the patient was admitted to a daily hospital at the Rheumatology Clinic with the previously established diagnosis M31.3, i.e. Wegener Granulomatosis, and an acute new diagnosis M15.9, i.e. Polyosteoarthritis, unspecified. During the hospitalization at the Rheumatology Clinic, the patient was administered Cyclophosphamide according to the protocol, which she tolerated well. The treatment was continued with monthly applications of Cyclophosphamide for a period of 6 months and a high dose of oral corticosteroids. A recommendation was made for a neuromuscular biopsy in consultation with a primary neurologist, regular control of blood pressure and glycemia, and rheumatology control after one month. Home therapy was prescribed, which includes: Tbl. Decortin H20, Tbl. Decortin H5, Tbl. Furosemide, Tbl. CaCO3 + Sol. Vigantol, Tbl. Paracetamol (for pain) and Caps. Lansoprazole.

One year later, in 2017, the patient was hospitalized again at the Neurology Department of the 8th September General Hospital. This was the third hospitalization of the patient with defined sensorimotor polyneuropathy as part of Wegener Granulomatosis, which was diagnosed during the two previous hospitalizations at the neurological department in 2015 based on the overall clinical symptomatology, paraclinical findings (elevated erythrocyte sedimentation rate, high values of CRP, RF and anti CCP antibodies, leukocytosis with neutrophilia), skin changes, finding of nodular shadows in the lungs and polyneuropathy. The symptomatology began 2-3 years ago, presenting with intense pain in the hands and feet, accompanied by muscle weakness in them, as well as changes in skin color, general weakness, malaise, increased sweating, weight loss (10-15 kg). In the interim period, with the persistence of discomfort, she was regularly monitored on an outpatient basis by a neurologist and rheumatologist. She was also treated with regular immunosuppressive therapy with Imuran and Decortin, and symptomatic therapy.

The last few months before the third admission to hospital treatment, she has been experiencing increased joint pain (due to which a series of X-ray investigations were performed - X-rays of the knees, shoulder joint, pelvis with hips), as well as weakness especially in the right arm and leg, with significant functional compromise. Third admission was performed for therapeutic purposes. The patient is on therapy with:

- Caps Pregabalin (Lyrica) a 75 mg 2x1

- Tbl. Sertraline a 100 mg 1x1

- Tbl. Demetrin 10 mg 1/2+1/2

- Tbl. Azatioprin a 150 mg 1x1

- Tbl. Decortin a 5 mg 1x1.

The patient's general status is settled, she is conscious and afebrile. She takes an active position in bed. Heart action is rhythmic, with well audible tones, blood pressure on admission 130/70 mm/Hg, heart rate 94/min. Auscultatory finding of lungs with vesicular breathing without accompanying pathological murmurs. Abdomen at the level of the chest, palpatory soft and painless, without signs of organomegaly, with preserved peristalsis on auscultation. Regarding the neuro-psychic status, the patient is conscious, properly oriented with suppressed basic mood and proper innervation of the cranial nerves. There is generalized, predominantly distal, hypotonic, hypotrophic muscle weakness of the upper and lower extremities. MTR are obtained with proper responses, pathological reflexes - bilateral drop foot. Hypoesthesia and paresthesias of peripheral type of distribution, on the hands to the level of the hands, on the legs to the level of the feet. Walking is difficult on the heels and toes, it is paraparetic. Sphincters are properly controlled. In addition to the existing diagnoses of Polyneuropathia senso-motoria vasculitica and Polyangiitis cum granulomatosis-Wegener, the patient has also been diagnosed with Chronic Obstructive Pulmonary Disease (COPD) and Steatosis hepatis (fatty liver). A control with the attending rheumatologist and neurologist is scheduled after 1 month, as well as control of glycemia (fasting and postprandial, or with OGTT). Recommended therapy:

1. Caps. Pregabalin (Lyrica) a 75 mg 2x1 (for neuropathic pain control)

2. Tbl. Sertraline a 100 mg 1x1 in the morning

3. Tbl. Demetrin a 10 mg 1/2+1/2

4. Tbl. Azatioprin a 50 mg 2x1

5. Tbl. Prednisolon (Decortin H) a 5 mg 1x1 in the morning, after a meal best around 8 to 9 am

6. Tbl. Ranitidine a 150 mg 2x1

7. Tbl. Enalapril a 10 mg 2x1

8. Tbl. Amlodipin a 5 mg 1x1

9. Tbl. Aminophylline R 2x1/2

10. Spray Flexotide 3x2

11. Spray Ventolin as needed

12. Gut. Vigantol 1x15 drops/week

13. Caps. Sylimarin 3x1

14. Tbl. Naproxenum a 550 mg 2x1 after meals for pain

Local therapy:

1. Ung. Bethametasone S. 2x1 on areas where the skin is dry.

2. Bioderma Atoderm Intensive/ Pantexol balm on areas where the skin is dry, intensively and continuously in the morning and evening.

3. Tbl. Loratadine S 1x1 as needed with meals.

Over a period of six years, the course of the disease was properly controlled, but in December 2023, the clinical manifestation deteriorated. The patient presented with quadriparesis, somnolence and hypoxic encephalopathy, in addition to existing and previously established diagnoses. The patient was admitted as an emergency case to the Neurology department due to drowsiness, confusion, disorientation, shortness of breath, decreased oxygen saturation and clinical parameters for hypoxic encephalopathy. She was with regular immunosuppressive therapy. The patient's Wegener's Granulomatosis progressed, given that in addition to rheumatological symptoms, she also exhibited symptoms and signs of peripheral nervous system involvement (sensorimotor neuropathy), as well as respiratory symptoms.

Deterioration of the condition was noted one week before the emergency admission, when the patient became drowsy, difficult to communicate, dyspnoeic. An urgent CT scan was performed, which was without signs of acute focal changes (right temporal cortical, difficult assessment of the parenchyma). Initial cortical atrophic changes were noted on the left side. X-ray findings of the lungs indicate reduced transparency in the basal parts of confluent shadows, consolidated more to the right; expanded mediastinal shadow; voluminous hilus; FC (phrenico-costal) sinuses are not seen; aortic heart.

Somatic status: Afebrile, obese, with generalized edema, face with a Cushingoid appearance, with an aspect of severe illness. On auscultation of the lungs, there is weakened vesicular breathing with the finding of rare scraping and moist rales on both sides. Cardiac action is tachycardic, without accompanying murmurs, tones are clear, BP 130/70 mm/Hg. Extremities with mild testicular swelling bilaterally pretibially.

Neurological status: Isochoric pupils, moderately miotic, reactive to light. Generalized proximal and distal hypotonic, hypotrophic muscle weakness (with inability to raise the arms to a horizontal level), with hypotrophy of the small muscles of the hands, thenar, hypothenar, forearms, lower legs and feet; the same of moderate-severe degree in the upper extremities and severe degree in the lower extremities. Generalized abolished myotatic reflexes; pathological reflexes are not established. Paresthesias and dysesthesias at the level of the lower legs, feet, forearms and hands.

Psychiatric status: The patient is in a somnolence-sopor state, confused, disoriented, with probable visual hallucinations.

Consultative examination by a pulmonologist: Gas analysis was performed with oxygen support with a flow of 2L/min - normoxemia, normosaturation, moderate hypercapnia ($PaCO_2$ = 64 mm/Hg - 9.1 kPa) with a proper acid-base balance. Objectively, on auscultation of the lungs, breathing is audible in front, weakened above the basal parts (inability to hear due to obesity and horizontal position of the patient). The pulmonologist gave recommendations for clinical monitoring.

Control examination by a pulmonologist: Control gas analyses - compensated global respiratory insufficiency. Recommendations for therapy given.

Consultative examination by a cardiologist: to continue with regular cardiological therapy. The examination is unremarkable.

Consultative examination by a psychiatrist: At the time of examination, the patient is conscious and uncertainly oriented in time and space. Psychomotor intensity. At times she gives adequate answers, at times inadequate answers. She complains of nervousness and tension. Recommendations for therapy given.

Consultative examination by a physiatrist: Early rehabilitation will include respiratory gymnastics in the supine and semi-recumbent positions and exercises with sitting in a bed at an angle of 90 degrees.

Decursus morbi: The patient was admitted as an emergency case due to drowsiness, confusion, disorientation, difficulty breathing, decreased oxygen saturation and clinical parameters for hypoxemic encephalopathy. During hospitalization, she was treated with corticosteroid therapy, bronchodilators, low molecular weight heparin, fast-acting insulin (depending on serum glycemia), broad-spectrum antibiotics, antihypertensives, oxygen on a mask with a flow of 2L/min. Early physical rehabilitation was also initiated. After prescribed therapy, a slight improvement in consciousness was observed, however, respiratory deterioration indicates progression of the primary autoimmune systemic disease. The overall symptomatology with dominant qualitative and quantitative changes in consciousness, indicate hypoxic encephalopathy.

Further recommendation for consultation with rheumatologist and pulmonologist and referral for third-party care was given due to the inability to care for herself.

Prescribed therapy:

1. Tbl. Rivaroxaban a 20 mg 1x1 in the evening around 8 pm

2. Tbl. Perindopril a 4 mg 1x1 in the morning

3. Tbl. Furosemid a 40 mg 1x1

4. Tbl. Spironolactone a 25 mg 1x1

5. Tbl. Mepinto a 47.5 mg 1x1

6. Tbl. Cordipin R a 20 mg 2x1

7. Tbl. Paroxetin a 30 mg 1x1 in the morning

8. Tbl. Prazepam a 10 mg 2x1/2 as needed for anxiety

9. Tbl. Decortin a 20 mg 1x1 in the morning, after a meal, best between 8-9 am

10. Tbl. Pantoprazole a 40 mg 1x1

11. Tbl. Methotrexate a 2.5 mg 6 tablets at once

12. Tbl. Metformin a 850 mg 1x1 at noon

13. Tbl, Reodon a 1 mg 3x1

14. Airflusal Forspiro Salmeterol/Fluticasone propionate a 50/250 mcg 2x1 inhalation, rinsing the mouth with baking soda after inhalation

15. Respimat Tiotropium Bromide a 2.5 mcg 1x2 inhalations at noon

16. Tbl Aminophyllin R 2x1/2

17. Recommendation for oxygen therapy at home for 16 hours a day, with an oxygen flow of 1.5 L/min.

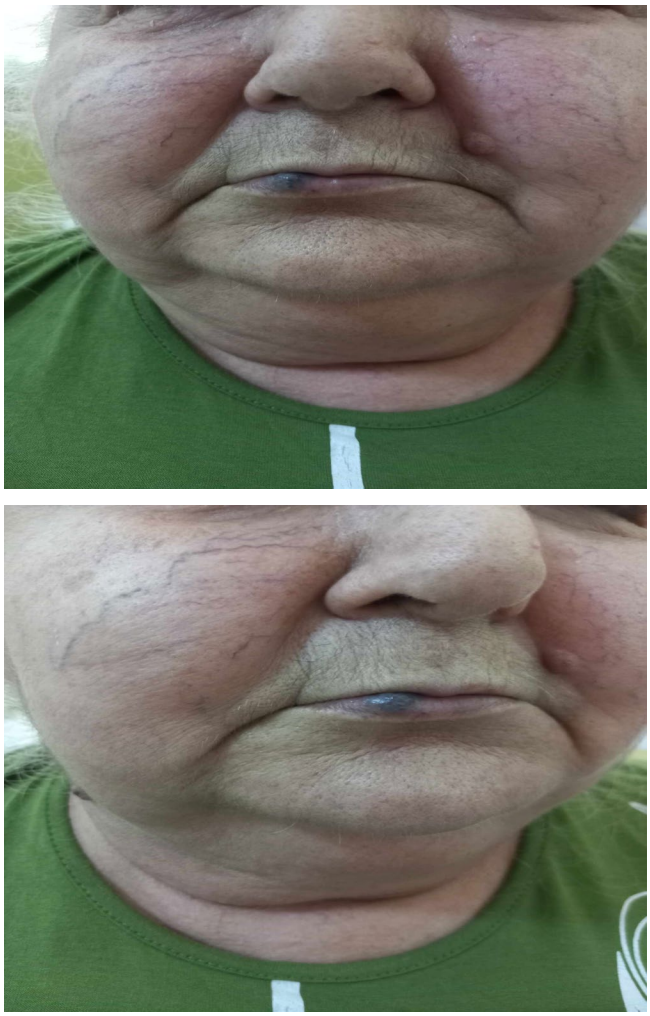

Figure 2: Changes on the face at the last clinical examination

At the last clinical check-up examination in May 2024, the patient came in a wheelchair due to expressed difficulties in walking. According to the heteroanamnesis from the family, she is sleepy all the time, however, without perceptual delusions, but with data for sleep disturbance, such as vocalization during sleep. Breathing is difficult, partly due to obesity with data on probable obstructive sleep apnea. Skin changes are present, most pronounced in the elbows, with a Cushingoid appearance on the face. Soft and pitting edema of the lower legs are present, as well as around the knees.

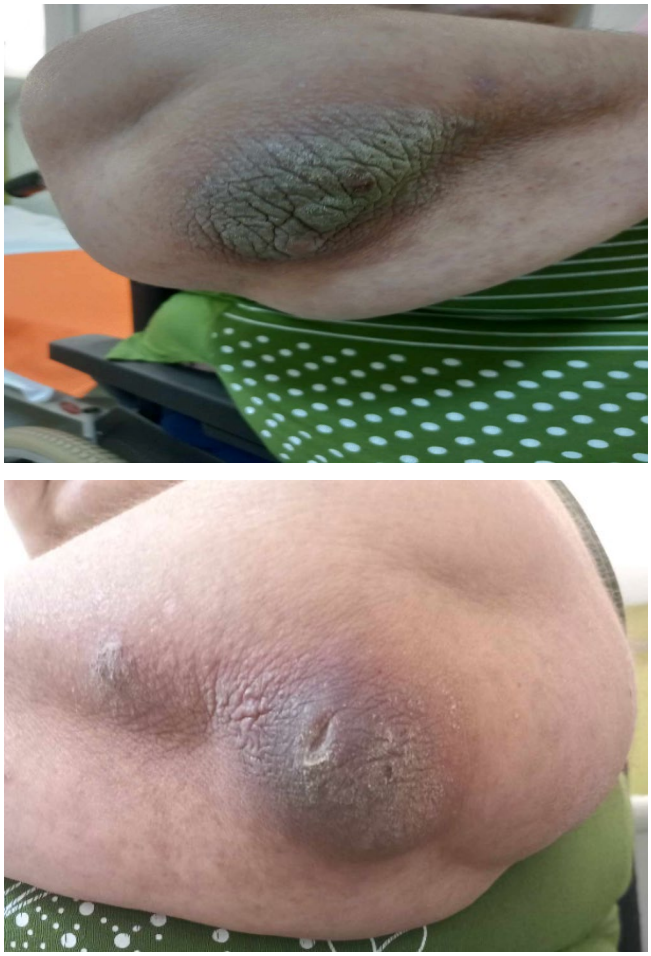

Figure 3: Changes on the elbows at the last clinical examination

Neurologically, signs of sensorimotor neuropathy and encephalopathy of a, probably hypoxic nature, dominate. She is currently using Methotrexate, Decortin, Pregabalin, Paroxetine and Rivaroxaban.

Discussion

Wegener's Granulomatosis is a systemic autoimmune disease, with a progressive character and a chronic course [8], [9]. The pathological process attacks small and medium-sized blood vessels. It causes necrotizing vasculitis of the blood vessel endothelium in most organ systems, most often the respiratory, renal and nervous systems, as well as the locomotor. The pathohistological substrate consists granulomas, which disrupt normal function of the affected tissue [10]. The form of the disease can be

generalized and localized. In addition to the presence of autoantibodies, certain drugs, as well as infectious agents, play a role in development of the disease. The clinical manifestation is characterized by a multitude of general symptoms and signs from the affected organs. The condition can suddenly worsen or have a gradual development of a severe clinical form. It is important to make diagnosis quickly and in a timely manner, in order to start timely treatment. When determining the diagnosis, it is mandatory to take into account the clinical manifestation of the patient's symptoms, the course and development of clinical signs, and the ACR criteria [1].

Definitive diagnosis is made by biopsy of the affected tissue and pathohistological analysis. Laboratory parameters and imaging techniques must not be omitted at all. From the laboratory analyses, the presence of anti-neutrophil cytoplasmic antibodies (ANCA) is dominant, although their absence does not exclude the possibility of developing granulomatosis with polyangiitis. In addition to these antibodies, there is an elevated sedimentation rate, CRP and leukocytosis with neutrophilia [11]. For detailed observation of inflammatory changes, Computed Tomography (CT) and Magnetic Resonance (MR) are most often used.

In this specific clinical case report, a progressive course of the condition of almost 10 years was observed. The initial symptoms that appeared in the patient had neurological origin. Also, the data from the searched literature refer to clinical cases where neurological symptoms were the initial clinical manifestation. For this reason, it is important and necessary to perform a detailed neurological examination. Biological preparations, corticosteroids and immunosuppressive drugs are used in treatment of this disease. They have the ability to control the patient's immune system and prevent excessive immune reaction. Main goal of the treatment is to achieve remission and prevent relapse. It is necessary to take into consideration the side effects of the therapy and to avoid complications. In the described patient, the treatment is carried out with a combination of corticosteroids and Cyclophosphamide, which the patient tolerates well, without any noted complications [12], [13].

Wegener's Granulomatosis is a very exhausting disease for patient and his family and a challenge for doctors. Life expectancy is variable, with active therapy it is estimated at 10 years. An active multidisciplinary approach is needed so the patient could be treated properly, with a better quality of life and improved daily functioning.

Conclusion

This form of polyangiitis is significant for the presence of anti-neutrophil cytoplasmic antibodies (ANCA) in the patient's blood. But their absence does not exclude the disease. In terms of therapy, drugs are used that regulate the patient's immune system. We must be aware of the risk of complications and worsening of the course of the disease. This condition requires a multidisciplinary approach to treatment.

This case report is unique, with a very thorough elaboration of a not so common disease that deserves attention.

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