Giant cell arteritis (a.k.a., Horton’s temporal arteritis) belongs to the vasculitis group of diseases and affects the great vessels, especially branches of the carotid arteries in the head. Polymyalgia rheumatica (PMR) was formerly regarded as an independent disease. Nowadays, due to shared accumulations, it is also commonly assumed that these two diseases are caused by identical pathogeneses, those of inflammatory involvement of the proximal peripheral arteries. Often, both phenomena occur simultaneously. Hence, a joint overview of both diseases follows, belonging to some of the most common rheumatologic diseases.

Definition of Giant Cell Arteritis and Polymyalgia Rheumatica

Vasculitis as the foundation of giant cell arteritis and
Both **giant cell arteritis (GCA)** and **polymyalgia rheumatica (PMR)** arise from the basis of **granulomatous vasculitis** of medium-sized and large arteries. Both diseases share symptoms of localized pain and the pathognomonic appearance of giant cells.

**Giant cell arteritis**, or **temporal arteritis**, predominantly affects cranial arteries radiating from the carotid artery and causes headaches so severe that it leads to ocular involvement.

**Polymyalgia rheumatica** causes symmetrical muscle pain in the shoulder girdle through inflammation of the peripheral **arteries**, often the subclavian artery. Additional features of PMR are bursitis or synovitis of large joints.

### Epidemiology of Giant Cell Arteritis

**Giant cell arteritis – older women commonly affected**

Giant cell arteritis is the most common of all vasculitides, affecting mostly women of advanced age (this is true for about 75% of all cases). The incidence following the eighth decade of life is around 50/100,000.

**Note:** Unlike giant cell arteritis, which affects mostly older women, **Takayasu’s arteritis** often occurs in patients under 40 years of age.

### Etiology of Giant Cell Arteritis

**Cause of giant cell arteritis still unclear**

The etiology of giant cell arteritis remains unexplained to this day. The scientific community suspects a trigger, possibly through viral infections, which activates the disease in the presence of **genetic predisposition**.

### Pathology and Pathophysiology of Giant Cell Arteritis and Polymyalgia Rheumatica

**Giant cell arteritis and polymyalgia rheumatica are autoimmune diseases**
Giant cell arteritis and polymyalgia rheumatica are **T-cell-dependent autoimmune diseases**, which particularly affect medium-sized and large arteries as granulomatous polyarteritis. The pattern of involvement is indeed different, and the diseases can occur separately; however, in about 50 % of the cases, the two manifestations overlap.

Histologically, it can often be detected in the vessel wall through the presence of **giant cells** and infiltration of **lymphocytes**. Significant **luminal stenosis** frequently results due to reactive wall thickening in the inflamed areas. This is especially relevant in implication of the **ophthalmic artery** as hypoperfusion of the eye may lead to irreversible damage within few hours. In polymyalgia rheumatica, inflammatory infiltrates may also be found in the large joints.

### Symptoms of Giant Cell Arteritis and Polymyalgia Rheumatica

Both diseases are associated with **general symptoms** such as **fatigue**, unintentional **weight loss**, **night sweats** and possible **fever**. In addition, both GCA and PMR have classic symptoms and patterns of involvement, by which they can be distinguished from one another. It is important to remember that in about 50 % of GCA cases, PMR is also present; as such, these diseases are not always observed in isolation.

### Symptoms of giant cell arteritis

The majority of patients with present giant cell arteritis primarily complain of **throbbing and ‘drilling’ headaches**, with the sensations often occurring unilaterally and at the temples. If these are aggravated by chewing, the pain is referred to as **claudicatio masticatoria** (masseter pain).

Furthermore, about 40 % of GCA cases present with **ocular involvement** in the form of transient **visual disturbances** through hypoperfusion of the retinal arteries (**amaurosis fugax**), ocular pain and general vision impairments progressing to blindness. The time window for avoiding irreversible blindness in progressive symptoms may be less than an hour.

Clinically, patients with manifest GCA typically present with a tender prominent temporal artery and, in certain circumstances, a weakened pulse. Rarely, larger proximal vessels, such as the aorta or peripheral arteries, may also show inter-arm and inter-leg blood pressure differences and claudication.
Symptoms of polymyalgia rheumatica

Polymyalgia rheumatica has symmetrical, intense pain in the shoulder or hip regions as a leading symptom. The pain especially appears at night and is often accompanied with tenderness of the muscles in the affected areas. Usually, affected patients complain of a morning stiffness that typically ceases about an hour or more after waking up. Joint involvement may eventually lead to severe restriction of a patient’s movements.

Diagnostics

Inflammatory markers provide information about giant cell arteritis and polymyalgia rheumatica

For both diseases, the American College of Rheumatology has devised diagnostic criteria (ACR criteria), which are used for diagnosis and progression monitoring.

In both diseases, medical laboratory inflammatory parameters are paramount for establishing the diagnosis. A commonly observed phenomenon is an extremely elevated erythrocyte sedimentation rate. In the initial stage, however, the ESR may be normal. Another important inflammatory parameter is C-reactive protein (CRP), a liver-derived acute phase protein. Leukocytosis in patients with GCA or PMR is less pronounced.

Ultrasound is an important method of investigation. In the case of giant cell arteritis, a narrowing of the lumen and a hypoechoic ring (halo) around the vessel are observable through color-coded duplex ultrasound of the temporal arteries, presumably showing an inflammatory wall thickening. In the presence of typical signs, especially that of high-grade stenosis, one can probably do without biopsy of the temporal artery.
If PMR is suspected, ultrasound can be used to investigate proximal peripheral arteries; signs of bursitis of the large joints, especially bilateral bursitis, allows one to obtain additional confirmation.

ACR criteria of giant cell arteritis

ACR criteria of giant cell arteritis (temporal arteritis) require that at least 3 of the following 5 criteria are met for diagnosis to be made:

- Age > 50
- New-onset headaches
- Abnormal temporal arteries (absent pulse, tenderness, swelling)
- ESR > 50 mm in the first hour
- Abnormal temporal artery biopsy

ACR criteria of polymyalgia rheumatica

The ACR criteria of polymyalgia rheumatica presume the patient to be at least 50 years old, to complain of new-onset bilateral shoulder pain and to show increased CRP or elevated ESR in investigations. For this, the following criteria apply:

- Morning stiffness > 45 minutes (2pts)
- No evidence of RF/anti-CCP (2pts)
- Pelvic pain or stiffness (1pt)
- Absence of other joint manifestations (1pt)

Complemented by ultrasound:

- Shoulder joint and hip joint affected (1pt)
- Both shoulder joints affected (1pt)

PMR diagnosis is confirmed at a minimum of 4 points without ultrasound findings, or a minimum of 5 points when ultrasound findings are included.
**Note:** One can easily memorize the most important signs and symptoms of the disease using the ACR diagnostic criteria.

**Differential Diagnoses**

**Important distinction between giant cell arteritis and meningitis**

Important differential diagnoses of giant cell arteritis are other diseases that are associated with new-onset headaches. For instance, the distinguishing of meningitis is of utmost importance, as corticosteroid therapy could have disastrous effects. **Amaurosis fugax** can also arise from arterial occlusion, such as an **embolus** from the carotid artery.

**Exclusion of other rheumatic diseases in polymyalgia rheumatica**

One must especially distinguish polymyalgia rheumatica from other **rheumatic diseases**. Here, the absence of serum markers such as rheumatoid factor serves as crucial evidence. **Polymyositis** or **dermatomyositis**, which can have very similar characteristics, are accompanied by a greater increase in muscle enzymes than PMR is, and are especially associated with an increased CK-value.

**Therapeutics**

**Corticosteroids in treatment of giant cell arteritis and polymyalgia rheumatica**

The most important treatment of both diseases is the pharmacological management with **glucocorticosteroids**. However, differences exist in administration and dosage. After a few days, symptoms should improve significantly; if they don’t, other differential diagnoses should be considered.

Diagnosed **giant cell arteritis** is treated in different ways, depending on the acute symptoms. An initial **loading dose** of corticosteroids is gradually reduced to a
maintenance dose that is continued for at least 2 years, in order to prevent recurrence. If there is already ocular involvement or amaurosis fugax, it is considered an emergency, and one must immediately start intravenous high-dose therapy.

Polymyalgia rheumatica can usually be treated with a lower dose of corticosteroids. Through administering immunosuppressants one can reduce steroids; methotrexate, for instance, is suitable here.

Long-term corticosteroid therapy should be supplemented with bisphosphonates for osteoporosis prevention and with PPIs to protect the stomach lining.

**Note:** Swift action and intravenous corticosteroid therapy is of utmost importance, particularly in the case of imminent blindness!

**Prognosis of Giant Cell Arteritis and Polymyalgia Rheumatica**

**Progression of giant cell arteritis and polymyalgia rheumatica**

Untreated giant cell arteritis leads to blindness in about 30% of the cases. With appropriate treatment, the prognosis of both diseases is comparatively favorable. Complete remission can be expected after about 2 years of treatment. Recurrences present in approximately one in three GCA patients. After the treatment period of 2 years, one should start stepping down the dose of steroids as long-term corticosteroid therapy is associated with increased mortality.

**Popular Exam Questions on Giant Cell Arteritis and Polymyalgia Rheumatica**

The solutions are below the references.

**1. Which laboratory parameter is presumably greatly increased in polymyalgia rheumatica?**

   A. Rheumatoid factor (RF)
   B. Creatine kinase (CK)
   C. C-reactive protein (CRP)
   D. Hemoglobin value (Hb)
   E. Bilirubin value

**2. What is the most important acute complication of temporal arteritis?**

   A. Blindness
   B. Septic shock
   C. Hemorrhagic stroke
   D. Torn blood vessel
   E. Aphasia

**3. Which of the named body parts is generally most frequently affected by polymyalgia-rheumatica-related symptoms?**
A. Face and neck region
B. Lower legs and feet
C. Abdomen
D. Neck-shoulder-upper arm area
E. Forearm-hand region

References

ACR-Guidelines zur Polymyalgia rheumatic
Delank, Gehlen: Neurologie, 12. Auflage - Thieme Verlag
Oxford Handbook of Clinical Medicine, 9. Auflage - Oxford University Press
Herold, G. und MA: Innere Medizin, 2014 - Gerd Herold Verlag

Correct answers: 1C, 2A, 3D

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