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Vasculitis of the Central Nervous System – A Rare Cause of Stroke

Małgorzata Wiszniewska² and Anna Członkowska¹

¹*Department of Neurology Institute of Psychiatry and Neurology Warsaw*

²*Specialist Hospital, Neurological Department, Piła
Poland*

1. Introduction

The term „vasculitis” encompasses a heterogenous group of multisystemic disorders characterized pathologically by inflammation and necrosis of the blood vessel wall [1]. The sequelae of vascular inflammation depend on the number, site and size of involved blood vessels. In vasculitis a disturbances of the blood vesels wall are focal. The segmental lesions lead, most common, to stenosis, occlusion and infarction. The lesions can also produce aneurysms, vessel wall rupture and haemorrhage. Almost all forms of vasculitis can involve the vessels feeding the brain parenchyma and cause stroke-like episodes [2, 3]. The clinical presentation of involvement the central nervous system (CNS) is very variable. Headaches and encephalopathy are most typical. Meningeal signs, psychiatric syndromes, dementia, cranial nerve palsies, epileptic seizures alone or in combination with neuropathy/ polyneuropathy or muscle damage , multiorgan involvement and non-specific systemiec symptoms (fever, weight loss) are also well known symptoms. Vasculitis is an infrequent disorder: the overall annual incidence (excluding giant cell arteritis) is estimated to be 31-47 cases/milion, but exist very rare types of vasculitis such as Churg-Strauss where annual incidence amounts ony 1-5 cases/milion [1, 4]. For polyarteritis nodosa (PAN) an annual incidence of 1.6 per milion has been described [5]. Vasculitis is a rare cause of stroke, even in the young age groups: only 1 case among 515 consecutive intracerebral haemorrhage, 13 among 1904 consecutive transient ischaemic attacks (TIA), 8 out of 254 (3%) stroke patients aged under 50 years [6] and 10 out of 215 (5%) patients aged under 45 years [7]. Neurological symtoms and stroke may be the first manifestation of vasculitis or may complicate the clinical course of previously diagnosed case.

2. Classification of vasculitis

There have been several proposals for the classification of vasculitis [1, 8-11], but all have limitation. The simple division of vasculitis usually consider as primary and secondary [12]. According to the Chapel Hill Consensus Conferences (CHCC) the primary systemic vasculitides are classified into three main groups: the affecting predominantly large-sized vessels, medium- and small-sized vessels, respectively (table 1) [13].

Vessel size	Granulomatous	Nongranulomatous
Large	Giant cell arteritis Cranial arteritis Takayasu arteritis	
Medium		Polyarteritis nodosa Kawasaki disease Microscopic polyarteritis
Small (with ANCA)	Wegener granulomatosis Churg-Strauss syndrome	
Small (with immune complexes)		Cryoglobulinemic vasculitis Behcet syndrome

Table 1. Classification of primary vasculitides (according Berlitz)

<p>Blood examinations: Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), blood cell differentiation, liver enzymes, creatinine, glomerular filtration rate, coagulation tests with search for coagulopathies [especially anticardiolipin-antibodies (ACA), lupus anticoagulant (LA)], electrophoresis, creatinine kinase, lactate dehydrogenase (LDH), haptoglobin, ferritin, angiotensin converting enzyme, immunofixation, quantitative evaluation of immunoglobulins, search for cryoglobulins, TSH, thyroid antibodies, rheuma factor, antinuclear antibodies (ANA), ds-DNA antibodies, histon antibodies, complement, anti-Ro (SS-A), anti-La (SS-B) antibodies, c- and p ANCA/MPO [myeloperoxidase [MPO]], anti-endothelial antibodies, drug screening, blood culture Serologic tests for syphilis, borreliosis, hepatitis B and C, HIV</p> <p>Urine examination including electrophoresis</p> <p>Cerebrospinal fluid (CSF): cell differentiation, isoelectric focussing, cultures, antigens, PCR [viruses, bacteria, mycosis]</p>

Table 2. Laboratory tests which are performed in suspected vasculitis (according Berlitz)

3. Diagnosis

If the diagnosis of vasculitis is suspected, a comprehensive medical history and physical examination should be very carefully carry out. Then appropriate laboratory tests and other ancillary procedures must be performed to asses organ damage [12, 14]. Laboratory findings suggestive of a systemic vasculitis include an acute inflammatory response like: raised erythrocyte sedimentation rate (ESR) and increased values of C-reactive protein (CRP). Anemia, thrombocytosis, elevated liver enzymes and low complement are frequent associated findings. Specific antibodies in the blood are evaluated in second order of laboratory tests in specialistic laboratories. It ought to exclude diseases which can also cause the lesions in the blood vesels mimic vasculitis – that's why should carry out serologic tests

for syphilis, borreliosis, hepatitis B and C as well as HIV in such case. In primary angiitis of central nervous system (PACNS) serum findings usually are normal, but cerebrospinal fluid (CSF) studies reveal inflammatory findings – a mild lymphomonocytic pleocytosis or protein elevation. The disorders in CSF are present in more than 90% of patients, but they are non specific [15]. Table 2 contains laboratory tests which are carry out in patients with suspicion of vasculitis.

Imaging radiological techniques which demonstrate arterial vessels play significant role in securing the diagnosis of vasculitis, and also demonstrate the cerebral involvement [12, 14, 16, 17, 18]. Computed tomography (CT) and magnetic resonance imaging (MRI) detect multiple CNS vascular lesions when only one is clinically apparent. MRI is more sensitive than CT and good detect the lesions in the cerebral trunk which is not visible in CT. Multiple infarcts in different vascular territories and of different ages are suggestive of vasculitis [17] Normal MRI rather excludes intracranial vasculitis. On digital subtraction cerebral angiography (DSA), angio- CT, MR angiography (MRA), areas of stenosis, dilatation, occlusion, microaneurysm, and narrowing in multiple-vessel tributaries, collateral flow, and regionally prolonged circulation time are suggestive of vasculitis [16, 17, 18, 19]. MRA offers the possibility of directly evaluating small vessel vasculitis. Small vessel vasculitis is currently best demonstrated by changes seen in brain parenchyma on MRI, but high field strength (7T) [16, 17]. These findings are, however, non specific, as they can be seen in other conditions such as vasospasm related to subarachnoid haemorrhage, drugs or severe hypertension, diabetes with intracranial atheroma, embolus recanalization, radiation, infectious or carcinomatous leptomeningitis, sickle-cell disease, reversible angiopathies, dissection. Therefore it is very important to analyse the radiological findings in clinical context. MRI performed with and without contrast medium is the good investigation not only for detection but also for monitoring cerebral involvements [20]. Noteworthy is 18-fluorodeoxyglucose positron emission tomography as well as certain MRI techniques which allow the visualization of vessel wall inflammation when the lumen is still unaffected on angiography, but these are methods high specialistics and not commonly used [16, 17, 21].

Confirmatory evidence can be obtained from tissue biopsy of occupied organ but this is invasive method not always perform. Moreover the lesion in vasculitis is segmental, and it can not encounter on involvement vessel [12, 14]. That's why a normal tissue biopsy does not exclude vasculitis.

4. Treatment of vasculitis

There are few controlled studies on the treatment of vasculitis. Usually a combination of steroids and cytotoxic drugs is used, but there is considerable variation between centres on current therapeutic regimens [12]. Cytotoxics drugs are usually used if the response to steroids is poor, if there are too much steroid side effects, in diseases that respond better to cytotoxic agents and in severe cases [12, 18, 22].

We present pathological and clinical aspects of most important primary vasculitides.

5. Primary vasculitis

5.1 Primary, large vessel vasculitis

Giant cell (temporal) arteritis (GCA)

GCA (Horton disease) is the most common of all the vasculitides. It is a chronic, granulomatous vasculitis of large- and medium sized arteries. Women are affected more

frequently than men (3:1 to 5:1). Mean age at the beginning of the disorder is 65 years or more. The American College of Rheumatology (ACR) diagnostic criteria [23] include at least three of: 1. Age 50 years or more, 2. New developed headache, 3. Tenderness of the superficial temporal artery, 4. Elevated sedimentation rate, at least 55 mm/h, 5. Giant cell arteritis in a biopsy specimen from the temporal artery. The definitive diagnosis of GCA requires the pathologic demonstration of biopsy with the typical mononuclear, granulomatous cell infiltration of all mural layers, usually with multinucleated giant cells (fig.1). The lesion of vessels involves most often superficial temporal artery, vertebral, ophthalmic and posterior ciliary arteries, seldom external and internal carotid arteries, while intracranial arteries are almost never involved [24]. The biopsy is the cornerstone of diagnosis and often remains positive for two to six weeks after the commencement of treatment but the best delivery is performance of biopsy as quick as possible but this procedure can not delay treatment. Besides biopsy may be negative in some patients with GCA, due to the presence of skip lesions or to sub-optimal biopsy. Therefore, patients with negative biopsy should be managed as having GCA if there is typical clinical and laboratory picture [25]. Duplex ultrasonography can detect the characteristic appearance of a hypoechoic "halo", occlusion and stenosis, but requires a high level of experience and training. Positron emission tomography scanning can pick up aortic and large vessel disease, and 3-tesla MRI has shown mural enhancement in inflamed temporal and others arteries. These new techniques can be useful for diagnosis and monitoring the disease, but currently are too expensive, not easily accessible, and do not replace the temporal artery biopsy. Clinicians should remember that jaw and tongue claudication, visual symptoms, constitutional symptoms (fever, weight loss and tiredness) and acute phase response may go unrecognised especially if not accompanied by headache [26]. Abrupt and irreversible loss of vision due to anterior ischaemic optic neuritis is the most dramatic complication of GCA. Two-thirds of the patients had premonitory visual symptoms like blurring, diplopia or amaurosis fugax the week preceding visual loss [27]. Cervical and muscular pain can occur, usually in the early phase of the disease. Transient ischaemic attack (TIA) and stroke are rare (about 7%) and often in the vertebrobasilar territory [28]. Very rare stroke is the first symptoms of GCA [29].

Steroids are the first line drugs in treatment of GCA. High-dose glucocorticosteroid should be initiated immediately when clinical suspicion of GCA is raised. In uncomplicated GCA prednisolone 40-60 mg (not less than 0.75 mg/kg) daily until resolution of symptoms and laboratory (most important decrease of ESR) abnormalities is recommended. In visual loss or history of amaurosis fugax as well as in stroke intravenous methylprednisolone 500mg to 1 g daily for three days before oral prednisolone should be given [25]. During steroids treatment it should be remembered to take care of bone and gastrointestinal protection applying weekly bisphosphonate and calcium/vitamin D supplementation, and proton pump inhibitors (eg omeprazole, lansoprazole). Prednisolone is continued in full dose during four weeks or until withdrawal of symptoms and lab abnormalities. Then dose is reduced by 10 mg every two weeks to 20 mg, then 2.5 mg every two to four weeks to 10 mg, and in the end by 1 mg every one to two months, provided there is no relapse. Sometimes the dose reduction is more rapid. In case of relapsing disease it ought to use steroids again [25].

Primary angiitis of the CNS (PACNS)

PACNS is a rare form of vasculitis of unknown cause, which accounts for just 1% of the systemic vasculitides [30]. The mean age of onset is 50 years, and men are affected twice as

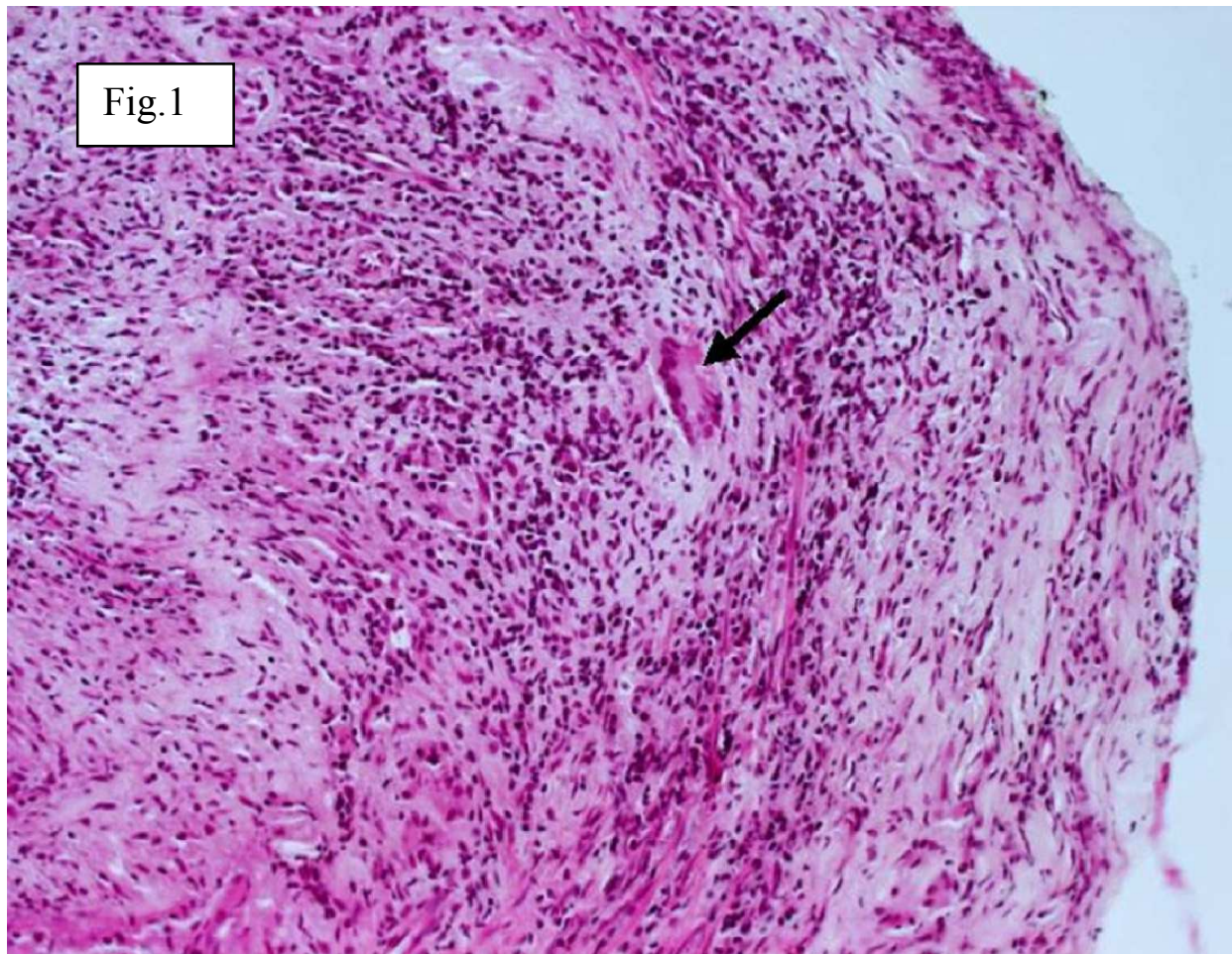


Fig. 1. GCA - biopsy of the wall of the superficial temporal artery (typical multinuclear large cell) (from collection of prof. J. Bogousslavsky)

often as women. It causes focal and diffuse neurological symptoms. Headache and encephalopathy are the most frequent initial symptoms. Stroke or focal symptoms develop in less than 20% of patients at the onset of the disease [30]. Serologic markers of inflammation are typically normal. Seizures have been reported in less than 25% of patients, fevers, weight loss, and night sweats have been reported in less than 20% of patients. Similarly, the erythrocyte sedimentation rate is elevated in less than 25% of patients. Substantial number of patients suspected of having PACNS will have signs and symptoms caused by vasospasm rather than true vasculitis of intracerebral vessels. Sometimes it is difficult to distinguish these two disorders. PACNS typically affects middle-aged men, reversible vasoconstriction syndrome is primarily seen in women aged 20 to 40 years. Puerperous period, migraines, illicit drug use, and use of over-the counter cold medicines, is frequently connected with vasoconstriction syndrome. Sometimes PACNS mimics cerebral tumors [31]

MRI should be the neuroimaging modality of choice for patients with suspected PACNS, and abnormalities are present in 90% to 100% of patients. MRI may demonstrate ischemic and hemorrhagic lesions most commonly seen in the subcortical white matter, followed by the deep gray matter, the deep white matter, and the cerebral cortex. Infarcts may be seen in approximately 50% of cases; when present, infarcts are usually seen bilaterally in multiple-

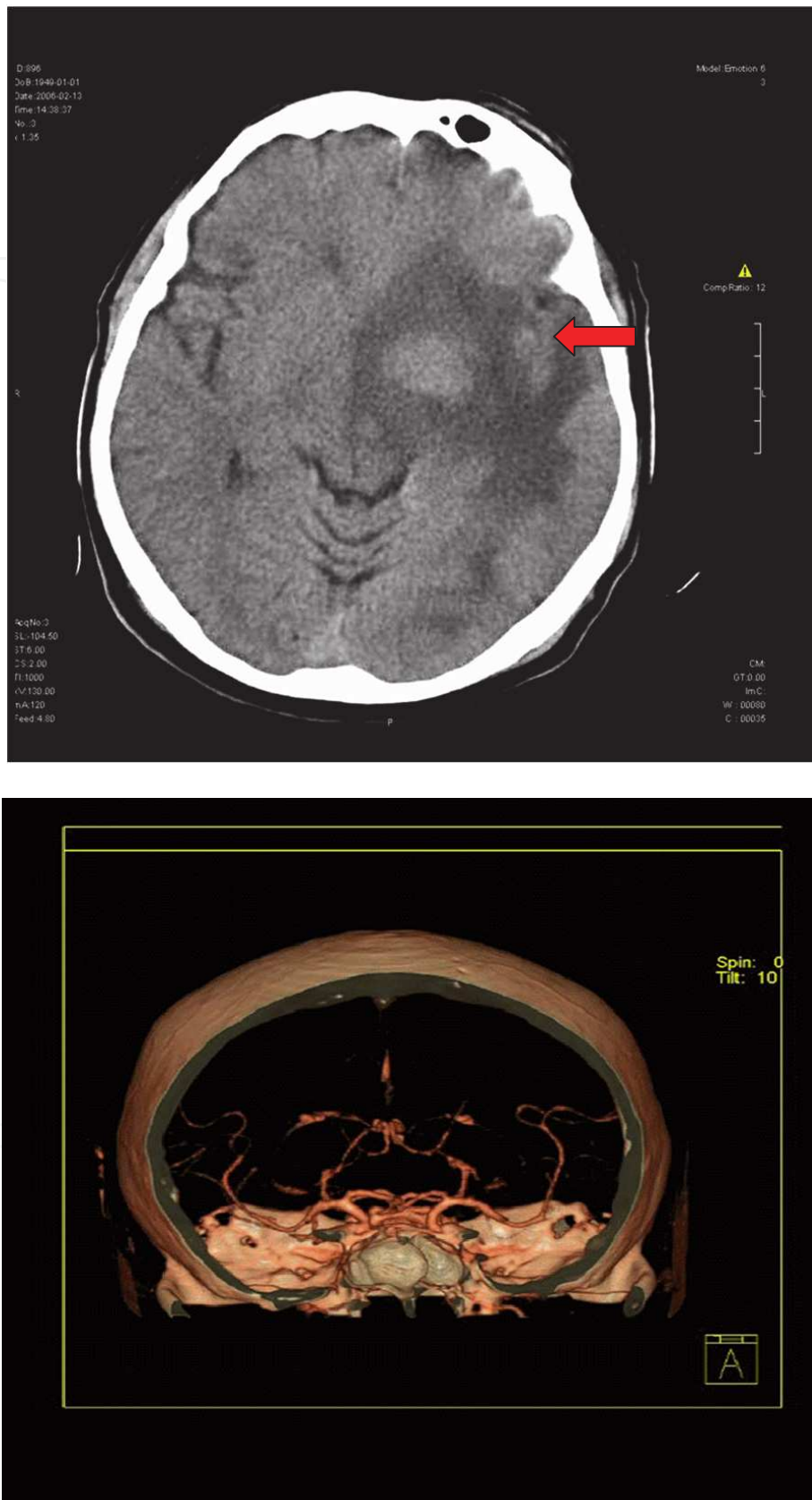


Fig. 2. CT and agio CT of the brain with lesion on left hemisphere, and typical segmental arterial stenosis in PACNS (own case)

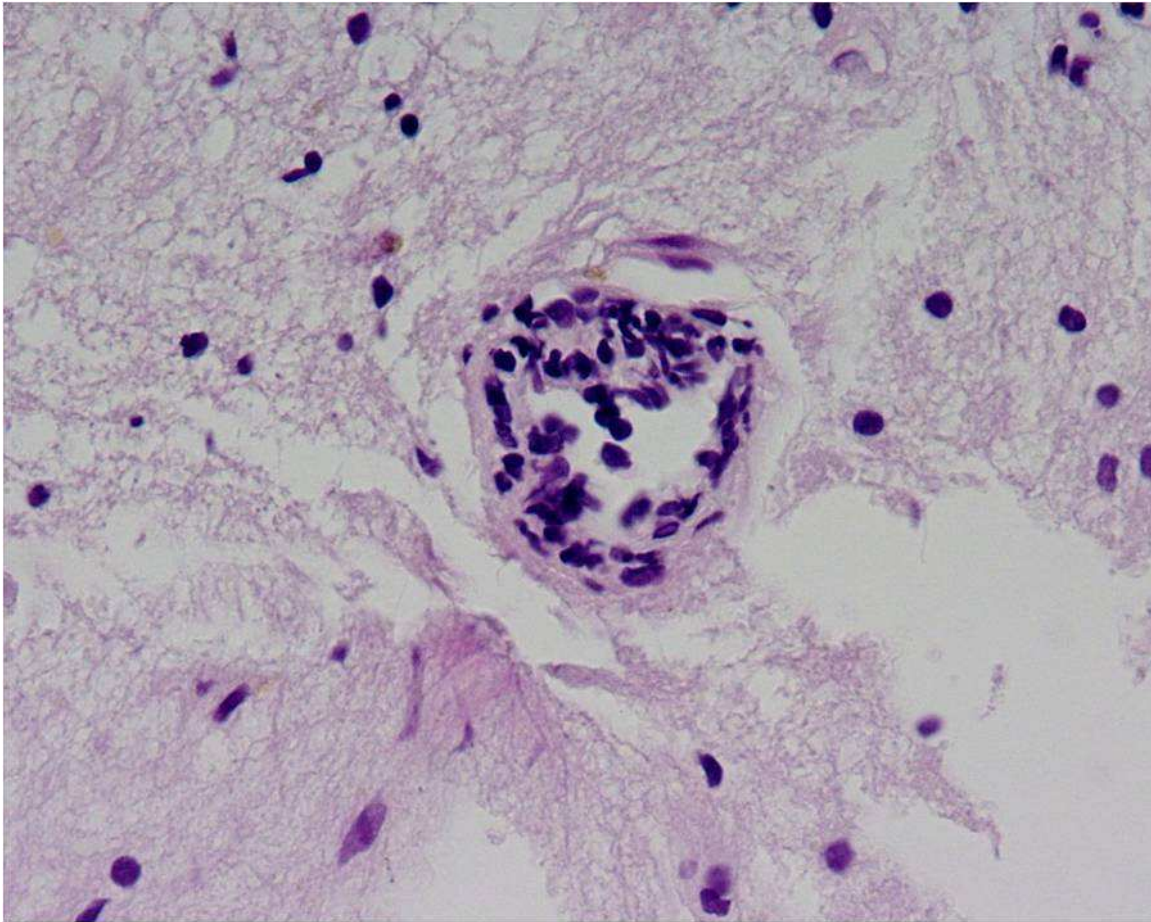


Fig. 3. Brain stereotactic biopsy from lesion in PACNS (lymphocytic cellular infiltrates of vessel wall) (own case)

vessel tributaries, affecting the cortex as well as the subcortex area. Both subarachnoid and intraparenchymal haemorrhages occur in approximately 10% of cases. Mass lesions mistaken for malignant neoplasms can be seen in as many as 15% of cases. Other common patterns include diffuse small-vessel changes of ischemic demyelination. Less common patterns include confluent white-matter lesions, which can be mistaken for multiple sclerosis [18, 30, 32]. Angiography sometimes demonstrates bilateral vessel stenoses or occlusion consistent with an angiitis (fig.2) [12, 18, 30, 32]. In cerebro-spinal fluid some findings have been reported [15, 32]. Pleocytosis and a protein elevation are frequent but not specific [15].

Criteria for the diagnosis of PANCS were suggested in 1987 by Calabrese and Mallek [33]: 1. Acquired neurological deficit unexplained after complete evaluation, 2. Diagnostic cerebral angiogram with narrowing of vessels, areas of dilation and/or beaded vessel appearance, displacement of vessels or vessel occlusions, 3. No evidence of systemic vasculitis or any other condition that could mimic the angiogram findings.

Definite diagnosis can give brain/leptomeningeal biopsy, which has a 53% sensitivity, and not rare it is negative [12, 30, 32, 34, 35]. Besides that brain biopsy remains the gold standard for the diagnosis of PACNS and stereotactic biopsy is generally only recommended for mass lesion. The brain biopsy is proposed all patients with a suspected diagnosis of PACNS before starting the plan of prolonged immunosuppressant treatment [30], but not always patients decide for this invasive procedure. The histological findings of PACNS consist of granulomatous inflammation, fibrinoid necrosis of vessel wall or exclusively lymphocytic

cellular infiltrates (fig.3.). There was no correlation between the histological pattern and clinical manifestations or prognosis [18, 36].

No controlled therapy studies for CNS angitis have been performed yet. In general, a combination of steroids and pulse cyclophosphamid (CYC) is recommended [18]. Birnbau propose treat patients with definite PACNS initially with oral CYC 2mg/kg/d and corticosteroid (usually in the form of prednisone 1mg/kg/d. if the patients has immediate life-threatening disease, it begin therapy with methyprednisolone 1g intravenously daily for 3 days. Thereafter, the patient is given oral prednisone 1mg/kg/d for 1 month, and the dose then is tapered slowly over 12 months [30]. During such high doses of prednisone, all patients should be received bisphosphonate, calcium and vitamin D, as well as pantoprazole, and supplementation of kalium periodically. Appropriate adjustments for CYC therapy, considering the patient's age, renal status, and other comorbidities, have been described elsewhere. In addition, all patients should be given Pneumocystis infection prophylaxix (eg trimethoprim and sulfamethoxazole 80mg/40mg, 1 tablet daily). Because the myelosuppressive effects of CYC are unpredictable it should be complete blood cell counts at least every 15 days imaging [30]. The therapy with CYC is management until induction of remission. After that is stoped and is continued by others immunosupresants like azathioprine or methotrexate. These agents are continued for 2 to 3 years [30]. The prognosis of PACNS is uncertain and careful; most patients dying within 1 year. Altered consciousness or mental state and diffuse cerebral dysfunction are associated with poor outcome, while focal symptoms are associated with better outcome [12, 18, 30, 37].

In this disease it should remember that a substantial number of patients suspected of having PACNS will have signs and symptoms caused by vasospasm rather than true vasculitis of intracerebral vessels. The vasoconstriction syndrome is reversible is primarily seen in women aged 20 to 40 years. A history of syndromes or provocative agents associated with vasospasm, including the puerperium period, migraines, illicit drug use, and use of over-the counter cold medicines, is frequently elicited. The most common presenting syndrome is of a thunderclap headache of sudden onset and sufficient severity to warrant diagnostic evaluation for subarachnoid hemorrhage. Focal symptoms can predominate with stroke or TIA as a presenting feature. Cerebrospinal fluid is normal. Prednisone with calcium channel blockers are used for treatment reversible vasoconstriction syndrome [30]. The prognosis in this disturbance is good.

Takayasu's arteritis

Takayasu's arteritis is a variant of giant cell arteritis affecting the aorta and its main branches to the limbs and the head. This disease is much more common in Asians than in Europeans and predominates in young female subjects. ACR diagnostic criteria include at least three of the following: 1. age at disease onset < 50 years, 2. Claudication of extremities especially upper limbs, 3. Decrease brachial artery pulse, 4. Blood pressure (systolic) difference > 10 mmHg between arms, 5. Bruit over subclavian arteries or abdominal aorta, 6. Arteriographic narrowing or occlusion of the aorta, its primary branches or large arteries (not due to arteriosclerosis, fibromuscular dysplasia or similar causes [38]. The disease starts with nonspecific systemic signs and symptoms such arthralgia, fever, fatigue, headaches, rashes and weight loss. In Takayasu's arteritis there are no specific laboratory abnormalities. An elevation of ESR or CRP and a mild anaemia are possible, but the acute phase response may be normal. Anti-endothelial antibodies (AEA) can bee present [18]. Main neurological symptoms are syncope and visual troubles. TIA or stroke are rare, and they are more often secondary to hypertension related to involvement of the renal arteries.

Digital subtraction angiography (DSA) is the gold standard for the demonstration of vessel stenoses or occlusion. MRI, and MRA, CT angiography, PET and high-resolution ultrasound are used more frequently for the investigation of Takayasu's arteritis [39].

Steroids are useful in therapy of the disease, and fifty percent of patients respond to them alone in early phase of the illness. The prognosis is relatively benign, 90% of patients being alive at 10 years [12]. In 20% of the patients the disease is monophasic and selflimited. Some patients need steroids plus CYC or low - dose methotrexate or azathioprine. Angiotensin II receptor antagonists are applied for treatment renovascular hypertension. Concomitant therapies include low-dose aspirin and statin even in normolipidemic patients [18]. Angioplasty and other vascular surgical procedures are used if it's necessary besides pharmacological treatment [40].

6. Primary, medium-sized vessel vasculitis

Periarteritis nodosa (PAN)

PAN is the systemic necrotizing vasculitis only of medium sized arteries without the involvement of smaller vessels. This disease may be associated with hepatitis virus infection. The diagnostic criteria of the ACR for the diagnosis of PAN include at least three of: 1. loss of weight > 4 kg, 2. livedo reticularis, 3. testicular pain, 4. myalgias, 5. mononeuritis or polyneuritis, 6. blood pressure elevation > 90 mmHg, 7. creatinine > 1,5 mg/dl, 8. hepatitis B or C virus antibodies, 9. pathologic arteriography (aneurysm, occlusion), 10. typical histology finding in biopsy of muscle with necrotizing granulomatous inflammation (fig. 4.) [12, 18, 41, 42].

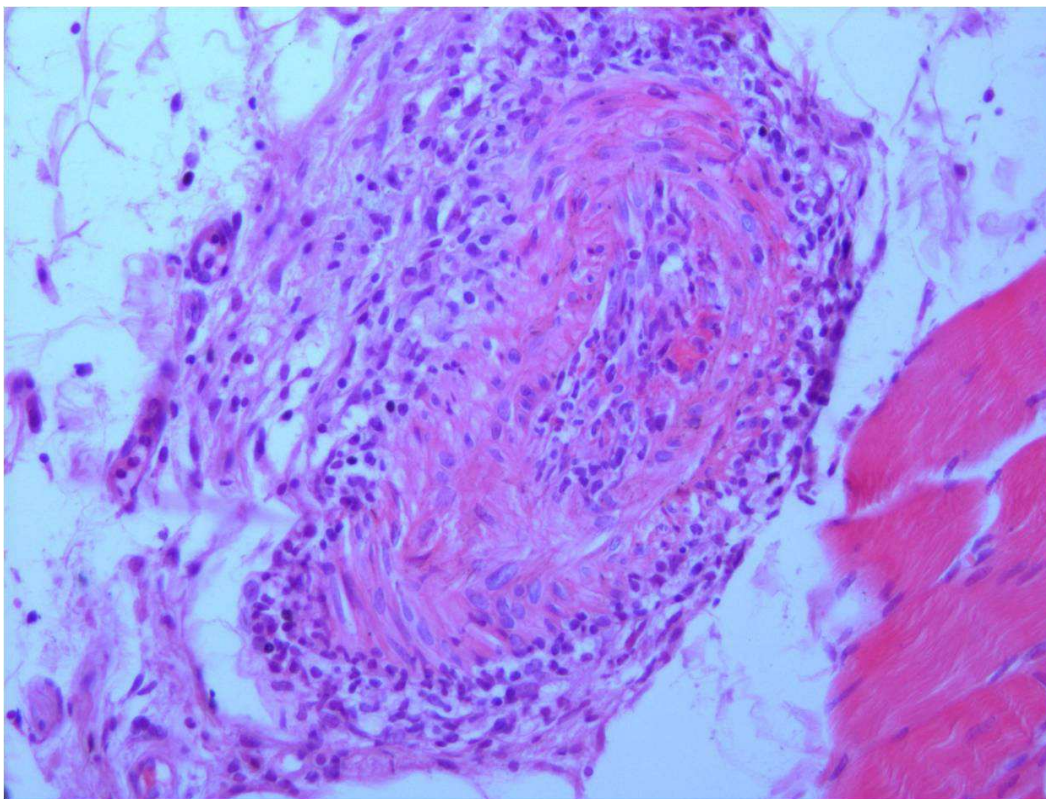


Fig. 4. Polyarteritis nodosa (PAN) - muscle biopsy with typical granulocyte infiltrates of small vessel wall with occlusion of vascular lumen (own case)

Twenty of patients with PAN had brain involvement in the monograph by Schmidley [15]. Ischemic stroke, hemorrhages and a progressive encephalopathy, seizures may occur. This disease is severe that's why it should be treated aggressively with steroids (pulses of methylprednisolone in severe forms) and cytotoxic drugs, mainly CYC. In emergency situations plasmapheresis may be tried [18]. Eighty percent of subjects who receive appropriate treatment are alive at 5 years [12].

Kawasaki disease

This is a rare medium-sized vessel vasculitis that causes stroke or encephalopathy in children. Its manifestations include fever, conjunctival infection, infected or fissured lips, infected pharynx or "strawberry tongue, erythema of palms or soles or oedema of hand or feet, polymorphous non-vesicular rash and cervical adenopathy. Treatment includes iv immunoglobulin and aspirin [12]

7. Primary small vessel vasculitis

Wegener's granulomatosis

This is rare small vessel (arteries and veins) arteritis frequently associated with autoantibodies: cANCA/PR3 and sometimes with MPO-ANCA [18]. Men are affected more frequent than women. The ACR criteria for classification of Wegener's granulomatosis include at least two of the following four: 1. necrotizing ulcerating inflammation of nose, sinuses, mouth, or pharynx, 2. irregular lung infiltrates, 3. nephritis, 4. granulomatous vascular and perivascular inflammation. In Wegener's granulomatosis a nonseptic meningitis with enhancement of the basal meninges and the development of an occlusive or communicating hydrocephalus are possible. Poyneuropathies, myelopathies and cerebrovascular symptoms frequently occur but stroke is a very rare complication [43, 44]. Nishino et al. and Fauci et al. described neurologic involvement in Wegener's granulomatosis in 22 - 33.6% of patients [45, 46]. Prednisone or CYC are used for treatment [45, 46, 47]. During CYC treatment don't ought exceed a cumulative dose > 30g, and higher doses should be strictly avoided [47]. During CYC therapy are useful: antiemetics, bladder protection with NaCl infusions and uromitexane perfusor as well as ovarien protection. Alternative treatment option is methotrexate 20-25 mg per week [48] or rituximab [49]. Azathioprine can be effective in supporting therapy. The presence of ANCA at diagnosis, cardiac or renal involvement increase the risk of relapses.

Behcet's disease

This rare disease is chronic-relapsing vasculitis affecting predominantly the venous system. It is more prevalent in the Middle East, Far East and the Mediterranean. Diagnostic criteria for Behcet's disease include at least three of the major signs (oral ulcers, genital ulcers, ocular lesions, mainly uveitis, cutaneous involvement-erythema nodosum) or the combination of two major signs and one minor (gastrointestinal or central nervous system (CNS) or peripheral nervous system (PNS) involvement, arterial lesions, thrombophlebitis, arthritis and familiar history) [50]. The course of Behcet's disease is fluctuating. The diagnosis of Behcet's disease is entirely based on clinical grounds since no pathognomonic laboratory or histologic findings exist. CNS involvement occurs in about 30% of patients after an average of 5 years. The most common clinical syndromes are encephalitis,

meningoencephalitis, and neuropathy as well as headaches, and brainstem symptoms. Cerebral oedema and cerebral stroke can also be present. Cerebral oedema is occurred in cerebral sinus thrombosis [18]. For the treatment of sinus thrombosis corticosteroids in combination with oral anticoagulation are recommended. Pulses of methylprednisolone of 1000mg (3 -5 days) followed by oral prednisolone (2 - 3 month) are used [51]. Occasionally may be used other immunosuppressive agents [52].

Churg -Strauss syndrome

Churg - Strauss syndrome is the rarest of the small-vessel vasulitides. Patients report in interview allergic diathesis and asthma. Pathologic hallmark of the disease are eosinophil-rich granulomas. p/ANCA/MPO are present in 40% of patients [53]. The criteria for the diagnosis of Churg-Strauss syndrome include at least four of: asma, history of allergy, eosinophilia (>10%), mono- or polyneuropathy, migratory or transitory pulmonary infiltrates and sinusitis [12]. Renal, lung, and central nervous system are involvement. 6-8% of patients present central nervous system symptoms like ischemic stroke, ischaemic optic neuropathy as well as intracerebral haemorrhage can occur in the disease [53, 54]. These patients may be treated with prednisone alone, but sometimes combined therapy with CYC is needed. The remission rate is 80-90%. Patient survival varies between 60% and 97% at 5 years [56].

8. Summary

Vasculitis is a rare, miscellaneous, and potentially treatable cause of stroke. The diagnosis contain labourious tests, neuroradiological image (CT, MRI with arteriography option) and often requires invasive procedures, including brain-meningeal biopsy. Treatment of vasculitis has an empirical basis and most common it is immunosuppressive. Multicentre therapeutic trials are ongoing. A lot of organs are damaged in vasculitis. These sickness required a great physician acumen and carefulness, as well as cooperation with others specialists. Fast reconnaissance of disease and accustoming of proper treatment can bring big benefits patient.

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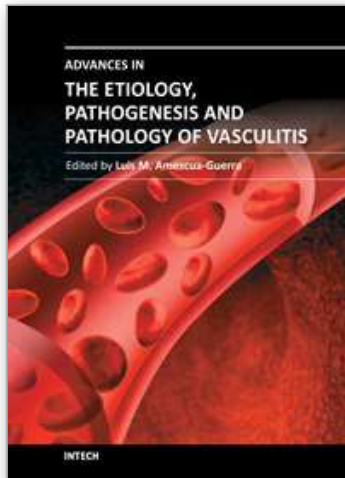
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This book represents the culmination of the efforts of a group of outstanding experts in vasculitis from all over the world, who have endeavored to devote their work to this book by keeping both the text and the accompanying figures and tables lucid and memorable. Here, you will find an amalgam between evidence-based medicine to one based on eminence, through an exciting combination of original contributions, structured reviews, overviews, state-of-the-art articles, and even the proposal of novel pathogenetic models of disease. The book contains contributions on the etiology and pathology of vasculitis, the potential role of endothelial cells and cytokines in vascular damage and repair as well as summaries of the latest information on several primary and secondary vasculitis syndromes. It also covers selected topics such as organ-specific vasculitic involvement and quality of life issues in vasculitis. The editor and each of the authors invite you to share this journey through one of the most exciting fields of the medicine, the world of Vasculitis.

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University Campus STeP Ri
Slavka Krautzeka 83/A
51000 Rijeka, Croatia
Phone: +385 (51) 770 447
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InTech China

Unit 405, Office Block, Hotel Equatorial Shanghai
No.65, Yan An Road (West), Shanghai, 200040, China
中国上海市延安西路65号上海国际贵都大饭店办公楼405单元
Phone: +86-21-62489820
Fax: +86-21-62489821

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