AUTOPSY CASES

(abs. 98-108)

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An autopsy case of primary cerebral non-Hodgkin's lymphoma associated with benign meningioma

The case of highly malignant primary cerebral lymphoma (PCL) and slow growing benign meningioma is reported: 78 years old man suffered from the diverse neurological symptoms. He was admitted to the Neurological Clinic, where CT scan and MRI identified the right parasagittal meningioma and the discrete paraventricular and brainstem lesions suspected to lymphoma. After the treatment with cortisone the majority of lymphoma lesions disappeared. The relapse appears very soon and the patient died. General autopsy examination did not recognize any malignancy outside the CNS. Neuropathologically, there were two different CNS neoplasms - malignant lymphoma and benign meningioma. The malignant lymphoma in a diffusely infiltrating manner, without evidence of mass lesion, was deeply situated adjacent to ventricular system in the cerebral hemispheres, brainstem and cerebellum. Histopathological features demonstrated the infiltration of brain parenchyma by lymphoid cells, which showed the characteristic tendency for perivascular accumulation within a concentric network of reticulin fibers. Lymphoid cells were large (20 to 30 μm), with vesicular nuclei and visible nucleoli. Mitotic figures were numerous. Imunohistochemically, they expressed LCA and pan-B markers (CD20, CD79a) and were negative for T cells marker (CD3). The diagnosis of primary cerebral diffuse large B-cell lymphoma was established. Besides the malignant lymphoma, meningioma WHO G1 (3 cm in diameter) was found parasagittally in the right parietal lobe. The association of PCL and benign meningioma is extremely rare. This finding may be coincidental or caused by some unknown predisposing pathological mechanism.

KEYWORDS: Brain Neoplasms; Lymphoma, Non-Hodgkin; Meningioma; Autopsy

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Autopsy diagnosis of Wernicke's encephalopathy in chronic alcoholics

Wernicke's encephalopathy (WE) is a rare disease due to thiamine deficiency. It occurs most often in chronic alcoholics. Because of the fact that in recent years, the classic clinical triad (disorientation, ophthalmoplegia, ataxia) is rare, or clinicians do not properly recognize the symptoms, the most diagnoses of WE are made at autopsy. We analyzed three cases of WE diagnosed at autopsy material of our Institute during the last three years (2001-2003). Brain cutting was performed after two weeks of fixation in 10% formalin. For microscopic analyses a different staining techniques (HE, reticulin, luxol fast blue, cresyl violet and immunostaining for GFAP and FVIII) were applied on representative samples of tissue. From 925 autopsy cases, we diagnosed WE in three cases (0,32%). They were chronic alcoholics. Two of them were female (37 and 56 years old) and one male (52 years old). In none of them there was clinical diagnosis of WE. Morphological changes of WE were not present in other 21 cases of chronic alcoholics in this autopsy material. Neuropathological findings: Coronal sections of brain and brainstem, revealed varied grayish or reddish discolorations, mostly symmetrically placed in the characteristic topographical distribution encompasses mammillary bodies, paraventricular regions of the thalamus, periaqueductal gray matter and floor of the fourth ventricle. Microscopic hallmark of the lesions in all cases was capillary proliferation with swelling and hyperplasia of endothelial cells. In both female, variable (slight to conspicuous) extravasations of erythrocytes about prominent blood vessels were present in the mammillary bodies and the floor of fourth ventricle. Hemorrhage was not present in male case. Number of lipid-laden microglial cells (macrophages), reactive astrocytes and demyelination varied from case to case. In the majority of lesions neurons were speared, but in some, they demonstrated acidophilic degeneration and/or central chromatolysis. Diagnosis of WE in our cases was made at autopsy. Our results prove the facts that chronic alcoholics are most often affected with this disorder, as well as, that only a subset (3 from 24 our cases) of chronic alcoholics actually comes down with the disease.

KEYWORDS: Wernicke Encephalopathy; Acoholism; Autopsy; Diagnosis

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Angiodisgenetic necrotising myelopathy: A case report

This is a rare condition, sometimes called Foix-Alajounine disease, re-named angiodisgenetic necrotising myelopathy (1951). The underlying pathology in many of these casers in now considered (Antoni, 1962) to be a spinal arterovenous aneurysm or fistula-angiomatous malformations recognized by myelography. Histological changes are more advanced in the caudal part of the spinal cord: altered capillaries at veins, often with thrombosis. The majority of the cell bodies are lost. The upper parts of the spinal cord show Wallerian degeneration of the upward directed long tracts. We present the autopsied case of 35 years old woman with 5-year long history of polyneuropathy. The predominant signs where week legs (less arms), with sensations of quizzing and massive increase of proteins in cerebrospinal liquid. Disturbance of deep sensibility was something that did not fit into the classic picture of polyneuropathy. Section of the whole brains, spinal cord and some spinal ganglia, were stained for myelin and tissue blocks for HE, Cresyl violet, PTAH, Kluver Barrera, AgNO3, Van Gieson and Holzer. Grossly: Dorsal and ventral surfaces of the spinal cord showed dilated at extremely tortuous blood vessels. Microscopically: The lower thoracic and lumbosacral segments of the spinal cord tissue shows ishemic patchy necrosis, mainly of the posterior and lateral funiculus and many enlarged thick - walled blood vessels, some in clusters, within the affected segment of the cord and both within and outside the necrotic regions. The adventitia of the colagenous thickened blood vessels contains some interspersed elastic fibers, the endothelial lining was intact and the lumen was often narrowed. Some of the blood vessels walls where hyalinized. In general, the pathological changes in the walls of the blood vessels make it impossible to identify individual vessels as arteries or veins. Most of the blood vessels appear to be veins or capillaries. Arteries seem to be relatively spared. The necrotic lesions where characterised by coagulating necrosis with evidence of phagocytosis and gliosis at the periphery of the necrotic foci in the more intact tissue only. Lower thoracic and lumbosacral segments where most severe affected. Cervical segments where most spared. Analogous changes where found in the blood vessels of the spinal cord roots. Most observers have considered that primary cause of this rare form of the myelopathy is the change in the blood vessels with congenital background. The concept of a congenital basis of the vascular abnormalities finds expression in the term - angiodisgenetic necrotising myelopathy (Foix-Alajounine Syndrome - Scholz and Manuelidis, 1951).

KEYWORDS: Spinal Cord Diseases; Arteriovenous Fistula; Autopsy; Microscopy

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Wegener's granulomatosis: A case report

Authors present a case of 47-year old female patient with clinical manifestations of uremic symptoms. This case report presents very aggressive, unusual and malignant form of Wegener's granulomatosis. Wegener's granulomatosis (WG) is usually characterized by the triad of necrotizing granulomas of the upper respiratory tract or lower respiratory tract or both, necrotizing or granulomatous vasculitis of small arteries and veins dominantly in the lungs but possibly elsewhere, and necrotizing, often crescent-shaped glomerulonephritis. The authors show a case of WG in 47-year old woman. The aim of this work was to present aggressive, "unlimited" WG. A 47-year old woman was transported from the hospital in Kosovska Mitrovica to the Institute of Nephrology Clinical Center of Niš, with uremic symptoms. At the time of hospitalization serum creatinine and urea were highly elevate; hemodialysis was applied through femoral catheter. A patient also had symptoms of lower respiratory tract, and low number of red blood cells. Laboratory examination suggested a systemic disease, but renal biopsy was not performed. After 13 days the patient died with symptoms of cardiovascular system insufficiency as well as lung infiltration. Autopsy material: 5 mm sections of formaldehyde-fixed paraffin-embedded tissues were performed and stained with HE, PAS, Van Gieson, and Gomori techniques. Morphology: macroscopically kidneys were slightly larger than normal and have normal shape and the weight 150 gr. Light microscopy showed advanced changes in the kidneys, which include diffuse extracapillary glomerulonephritis; 70% of the glomeruli were represented by proliferation of capsular epithelium or fibro-cellular crescent formation. Some of glomeruli showed capsular adhesions or advanced glomerular sclerosis. Lung was represented by several well-circumscribed firm areas, which were tan brown colored. Some of those were larger than 3 cm. Light microscopy showed hemorrhagic and necrotic areas in the center of the previous lesions. Beyond the necrosis was a zone of granulation tissue with lymphocytes, plasma cell and neutrophils. Giant cells of foreign body type, as well as epitheloid cells were also seen. In addition, the necrotizing arteritis of small vessels was also observed. There are fibrinoid necrosis, neutrophylic infiltrate of all coats, thrombosis and fibrous scarring, predominantly in the small vessels of lung, kidney, spleen and liver. Although this systemic disease is predominantly in men, the authors suggest that WG should always be considered when there are female patients with renal and lung symptoms.

KEYWORDS: Wegener's Granulomatosis; Vasculitis; Lung; Kidney; Diagnosis

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Combined myocardial infarction of left ventricle

Combined myocardial infarction of left ventricle (CMILV) is ischemic heart disease caused by either simultaneous occlusion of two coronary arteries (CA) or (more commonly) by occlusion of one and critical perfusion in other CA. Mechanism of CMILV was analyzed in 30 autopsy cases with myocardial infarction. The aim of this research was to analyze autopsy findings in cases of CMILV and compare them with clinical findings, in order to better understand clinical symptoms and improve treatment procedures in CMILV patients. We found that CMILV was caused by thrombotic occlusion of two CAs in 19 cases, which represents 63.33% of all cases.

Table 1. Causes and distribution of occlusion

Mechanism	No	%
Thrombotic occlusion of one CA	2	6.66
Thrombotic occlusion of two CAs	19	63.33
Thrombosis of one and intra-plaque hemorrhage in other CA	1	3.33
Occlusion of two CAs by massive atheromatosis	1	3.33
Thrombosis of one and subocclusion of other CA	3	10.00
Intra-plaque hemorrhage in one and subocclusion of other CA	1	3.33
Subocclusion of two CAs	3	10.00

Taking into account the thickness of left ventricle wall affected by ischemic necrosis, we found that infarction has involved nearly full thickness of the ventricular wall (transmural infarction) in 26 cases, while in 4 cases only part of ventricular wall was involved (nontransmural, subendocardial infarction). Autopsy findings were concordant with clinical findings, and clearly lead to conclusion that the most severe CMILV are the ones that affect full thickness of myocardial wall (transmural infarctions).

KEYWORDS: Myocardial Infarction; Heart Ventricle; Myocardial Ischemia; Autopsy

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Loeffler's endomyocarditis and sudden cardiac death

Sudden cardiac death (SCD) is most commonly defined as unexpected death from cardiac causes early after (usually within 1 hour) or without the onset of symptoms. In the vast majority of cases in adults, SCD is a complication of ischemic heart disease. Less frequently SCD is caused by myocarditis. Loeffler's endomyocarditis is marked by endomyocardial fibrosis typically with large mural thrombi, but unrestricted to a specific geographic area. Although the specific cause is unknown, eosinophils are considered important in the pathogenesis of this disease. A 36 years old male patient complained of the attacks of substernal chest pain that lasted several minutes. Pain occurred with progressively increasing frequency and was produced caused triggered by physical activity or emotional excitement. Respiratory infections with high temperature and marked leucocytosis (without data about eosinophils) were repeated many times during the last six months. He has dead suddenly, during the lunch. Autopsy showed pulmonary embolism with a saddle embolus with occlusion the smaller branching pulmonary artery. Many small pulmonary infarcts of basal localization and post infarction atelectasis were also observed. Heart: endocardial damage, with mural small thrombi in organisationem in the right ventricle and subsequent foci of endomyocardial necrosis in the right ventricle, accompanied by an eosinophilic infiltrate and by scarring of the necrotic areas are the features of Loeffler's endomyocarditis. Undiagnosed and untreated eosinophilic endomyocardial disease has a poor prognosis including sudden cardiac death. The release of toxic products of eosinophils, especially major basic proteins id responsible for rapidly fatal course.

KEYWORDS: Hypereosinophilic Syndrome; Death, Sudden, Cardiac; Pulmonary Embolism; Autopsy

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Sudden cardiac death of 23-year old woman with complicated libman-sacks endocarditis: A case report

Fibro-fibrinous, sterile vegetations most commonly occur on the undersurface of the mitral valve but may develop on either surface of any of four cardiac valves. Similar vegetations, which can be placed on endocardial surface or chorda tendinae, are consistent with Libman-Sacks endocarditis. This form of endocarditis occurs as a complication of systemic lupus erythematosus. It appears in 4% of patients with systemic lupus and rarely causes problems because they are not large and rarely embolize. Usually, the verrucae are clinically silent. Valvular dysfunction, particularly mitral or aortic regurgitation, is most often associated with healed form of lesion Systemic lupus erythematosus is multisystem disorder with cardiac involvement in about 50% of cases, yet clinically significant lesions are less common. Cardiovascular system involvement has been found to have a substantial effect on mortality and morbidity in patients with this disease. We describe a case of a 23 year-old-woman, who died suddenly without previous history of systemic disease. The autopsy revealed patoanatomic picture for systemic lupus erythematosus and Libman-Sacks syndrome. Namely, she had fibro-fibrinous vegetations on the mitral and aortic valves as well as on parietal endocardium of the left ventricle. The hypoplasia of aortic valve cuspis resulted in aortic insufficiency. Mitral valve alterations were complicated by ulcerotrombotic changes such as hemorrhagic infarct of left lung and anemic infarct of the left kidney. We found polyserositis, lupus nephritis, myocardial hypertrophy and dilatation of the heart, stasis and edema of the lugs. Bronchopneumonia fibrinopurulenta of both lungs was the cause of death. We presented a case of sudden death of 23-year-old woman without previous medical history of systemic disease. The autopsy revealed pathomorphologic substrate for systemic lupus erythematosus with cardiovascular involvement as Libman-Sacks endocarditis complicated by ulcerotrombotic (infective) endocarditis and its sequelae.

KEYWORDS: Death, Sudden, Cardiac; Endocarditis; Lupus Erythematosus, Systemic; Autopsy

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Amniotic band syndrome: A case report

The amniotic band syndrome (ABS) is a collection of fetal malformations associated with mesodermal bands that emanate from the chorionic side of the amnion appear to entrap or entangle various fetal parts in uterus and can affect any organ or system and cause a single or multiple anomalies. The group of defects identified as amniotic bands includes amnion rupture sequence (ARS) and body wall complex (BWC). Little is known about risk factors for either ARS or BWC, except that maternal age has been shown to affect risk inversely. ABS includes a spectrum of non-genetic anomalies, varying from simple digital band constriction to major craniofacial and visceral defects, and even fetal death. It usually is sporadic, and the incidence is approximately 1 in 15,000 live births, and affected children typically require involvement of several pediatric surgical subspecialties. It is suggest that young maternal age, low maternal education, amniocentesis, and unplanned pregnancy might increase the risk of BWC in offspring. Spontaneous lysis of an amniotic band was reported. A 17 years old woman presented for an initial prenatal visit at 22 weeks' gestation and had a first ultrasound that showed a single intrauterine pregnancy at 21 weeks' gestation. Labor induction was scheduled with a stillborn male infant delivered weighing 250 g after a spontaneous vaginal delivery. Extensive craniofacial anomalies were found. Histologically, all organs were macerated. Other congenital anomalies were not found. The possible case of intrauterine death may be an amniotic band constriction of the umbilical cord. ABS is a very rare, potentially fatal, non-genetic anomaly requiring appropriate counseling.

KEYWORDS: Amniotic Band Syndrome; Autopsy; Aborted Fetus

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DiGeorge syndrome: A case report

The DiGeorge syndrome is a primary immune deficiency disease caused by abnormal development of certain cells and tissues of the neck during growth and differentiation of the fetus. Tissues, which are dependent upon a single group of embryonic cells for their normal fetal development, are called "fields". Although the tissues and organs that ultimately develop from a "field" may appear to be unrelated in the fully formed child, they are related in that they have developed from the same embryonic or fetal tissues. Most patients with the DiGeorge syndrome have a small deletion in a specific part of chromosome number 22 at position 22q11.2. In minority of patients, the del (10p) was found. Thus, another name for the syndrome is the 22q11.2 deletion syndrome. Maternal alcoholism may result in the DiGeorge syndrome either as the single causative factor or in conjunction with other as yet unknown teratogens. Patients with the DiGeorge syndrome do not all show the same organ involvement. A given organ may be uninvolved, or so mildly involved that the organ appears to be normal. Affected children may have an upward bowing of their mouth, an underdeveloped chin, eyes that slant somewhat downward, low set ears and defective upper portions of their ear lobes. These facial characteristics vary greatly from child to child and may not be very prominent in many affected children. The primary cardiovascular anomaly always involved the aortic arch system or the arterial pole of the heart. The neural crest cells play a crucial role in development of pharyngeal (bronchial) pouch derivatives, e.g., thymus and parathyroid glands, as well as the aortic arches and the truncoconal part of the heart. In the male mature stillborn, weighting 2850 g, crown-rump length 27 cm, foot-length 7 cm, biparietal diameter 9 cm, the thymus gland was not found. The heart was enlarged, with common arterial trunk and ventricular septal defect. The DiGeorge syndrome is a very rare anomaly, requiring cytogenetic analysis. All suspected cases of microdeletion syndromes should be studied using fluorescence in situ hybridization, irrespective of high-resolution chromosome results.

KEYWORDS: DiGeorge Syndrome; Infant, Newborn

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Thoracopagus twins: A case report

Conjoined twins appear rare, 1/58000 birth in Europe. The most common type being the thoracopagus, in which conjoined site is the anterior thoracic and upper abdominal midline. In female thoracopagus twins, 2 months old the conjoined organs were heart, liver (with two billiary bladders), and part of the gastrointestinal tract (duodenum and part of small intestines). Conjoined heart had four chambers (two atrial and two ventricular) with connection at the atrial and the ventricular level. Both twins had common atrium. Membranous septum between them had one large and smaller defect as a communication. Systemic and pulmonary venous connections to each atrium were normal, except the persistent left superior vena cava, which is connected to common atrium of one twin. Two distinct atrioventricular valves were present. One had the structure of a tricuspid valve (twin A) and the other one resembled a mitral valve (twin B). The two ventricular chambers shared a common wall in which there was one defect. Great arteries in twin B were in transposition. Twin A had a right aortic arch and right ductus arteriosus. Twin B had a left aortic arch and left ductus arteriosus in process of obliteration. Each aorta had two coronary ostia. Surgical success in the separation of thoracopagus twins depends upon the level of fusion of the heart and associated cardiovascular abnormalities. In our case surgical separation was impossible.

KEYWORDS: Twins, Conjoined; Heart Defects, Congenital; Cardiovascular Abnormalities; Thorax; Abdomen

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Potter's syndrome: A case report

Syndroma Potter I means congenital defects in form of renofacial dyspalstic lesions which includes hypertelorismus, epicanthus, beaklike nose, low-set ears, receding chin and old-man look of newborn; it is accompanied with renal agenesis or hypoplastic kidneys. Congenital limb and vertebral anomalies, pulmonary hypoplasia, hydrocephalus, genital, and gastrointestinal malformations are also common. Most infants born with bilateral agenesis are stillborns, or those with dysplastic kidneys die immediately after delivery. Usually there is evidence of amnion nodosum and oligohydramnios and etipathogenesis is still unknown. We described a prematurely delivered fetus with clinical diagnosis: Graviditas hbd XXIII, Syndroma Potter I, oligohydramnios. Prenatal sonographic diagnosis showed enlarged hyperechogenic polycystic kidneys; differential diagnosis was bilateral agenesis along with presentation of enlarged discoid suprarenal glands (Power Dopller showed the absence of renal arteries); termination of pregnancy was indicated. Postabortal examination detected female fetus at XXIII gestational weeks confirmed multiple malformations: hydrocephalus internus, hypertelorismus, beaklike nose, low-set ears, receding chin, polycystic dysplastic kidneys, and solitary pancreatic cyst. Renal dysplasia is a failure of differentiation of metanephrogenic tissue that results in the presence of structures inappropriate to the gestational age of the patient with image of cystic dilated ducts lined by tall cuboidal or columnar epithelium surrounded by undifferentiated mesenchyme. Disturbance of branches of the ureteric bud to induce the differentiation of normal nephrons in metanephrogenic tissue, which leads into renal insufficiency. In about one third of prenataly US verified urinary tract anomalies, confirmed postnataly as Potter's syndrome. The incidence of oligohydramnions is about 3.5% and Potter's syndrome itself participates with 11 % in all cases of oligohydramnios.

KEYWORDS: Abnormalities, Multiple; Oligohydramnios; Kidney; Lung; Face; Urinary Tract; Fetus