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SILENT DEMYELINATION WITH FOCAL AND DIFFUSE RETICULOBLASTOSIS OF THE BRAIN

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Concomitance of demyelination with blastomatous proliferation is known from numerous clinico-pathological reports. It appears in many types and forms. In 1938 Scherer described a case of multifocal demyelination with blastomatous glial proliferation, considering this as a special form of multiple sclerosis. Progressive multifocal leucoencephalopathy is also representative of a typical demyelinating process concomitant with blastomatous astrocytic transformation. In a great proportion of cases this accompanies malignancies, involving mostly, although not exclusively, the reticulo-endothelial system. In 1974 Ulrich and Wüthrich published a case of typical multiple sclerosis, in which neuropathological examination revealed foci of reticulosarcoma connected or independent of demyelinating plaques. They quoted a similar unpublished case from Vildi's collection. One case from a series described by Schaumburg et al. (1972) and that published by Castagne et al. (1974) showed similar pathological features. It seems justified to include into the same group cases described by Dymecki et al. (1966) under the name of granulomatous encephalitis with focal demyelination and Haddenbrock et al. (1962), diagnosed as „phlebite granulomateuse sur sclérose en plaques".

The presented case, although similar in its general pattern to the above mentioned series, differs from most of them in its clinical and neuropathological picture.

CASE REPORT

The case concerned a 70-year-old female, otherwise healthy till her final disease, who had been suffering for the 2 last years from chronic spastic bronchitis. In the course of the last year some characterologi-

cal changes were noticed, accompanied with equilibrium disturbances and vertigo. Two months prior to her death she was hospitalized because of an acute febrile disease, diagnosed as bronchopneumonia. Neurological examination at the time of hospitalization revealed spasticity and weakness of upper extremities and bilateral ataxia. While at home, the general and neurological state of the patient was getting worse. Vertigo was increasing. So were gait disturbances. At the second admission to the hospital, three weeks prior to her death, the patient was emaciated, pale and semiconscious. Anisocoria (left pupil larger than the right one) and rightsided hemiparesis with pyramidal signs were present. In the course of several days complete left ophthalmoplegia appeared. This together with right spastic hemiplegia formed a typical alternate midbrain syndrome.

Laboratory tests revealed hypochromic anemia, increased white cells count, enhanced sedimentation rate (48/h), elevated globulin level in the blood and rise of proteins in cerebro-spinal fluid (73 mg%). Eeg was bilateraly abnormal, with theta and delta waves on the left side. The clinical symptomatology and course suggested either neoplastic or vasogenic process located in mesencephalic region. The general and neurological state of patient was rapidly worsening and she died three weeks after admission to hospital.

On general autopsy thrombosis of the right pulmonary artery and left illiac vein, infarction of the upper lobe of the right lung, emphysema and chronic bronchitis were found.

Naked eye brain examination revealed numerous and varying in size and shape foci of demyelination, which were widespread throughout the white brain structures, with an obvious predilection to the periventricular areas and a tumor involving midbrain and upper pons.

A microscopic brain examination was carried out on sections taken from different areas of both cerebral hemispheres, midbrain, pons, medulla and cerebellum, stained with routine and special neuropathological methods.

The microscopic picture of the brain was dominated by numerous demyelination foci, present both in cerebral hemispheres, brain stem and cerebellum. Aside large foci, often confluent, occurring mostly in the periventricular areas of brain hemispheres (Figs 1, 2), there existed small ones, located first of all in brain stem (Fig. 3) and cerebellum (Fig. 4). Most of the foci showed the features of an old demyelination. They had sharp borders, contained a scarce amount of regressive astrocytes (Fig. 5) and a dense network of glial fibers (Fig. 6). Axon cylinders, although reduced, remained preserved (Fig. 7). In some of the foci perivascular lymphocytic cuffs were present (Fig. 8).

Recent foci, with only partial destruction of myelin were also seen (Fig. 9). Their borders were less sharp, and the nerve tissue within

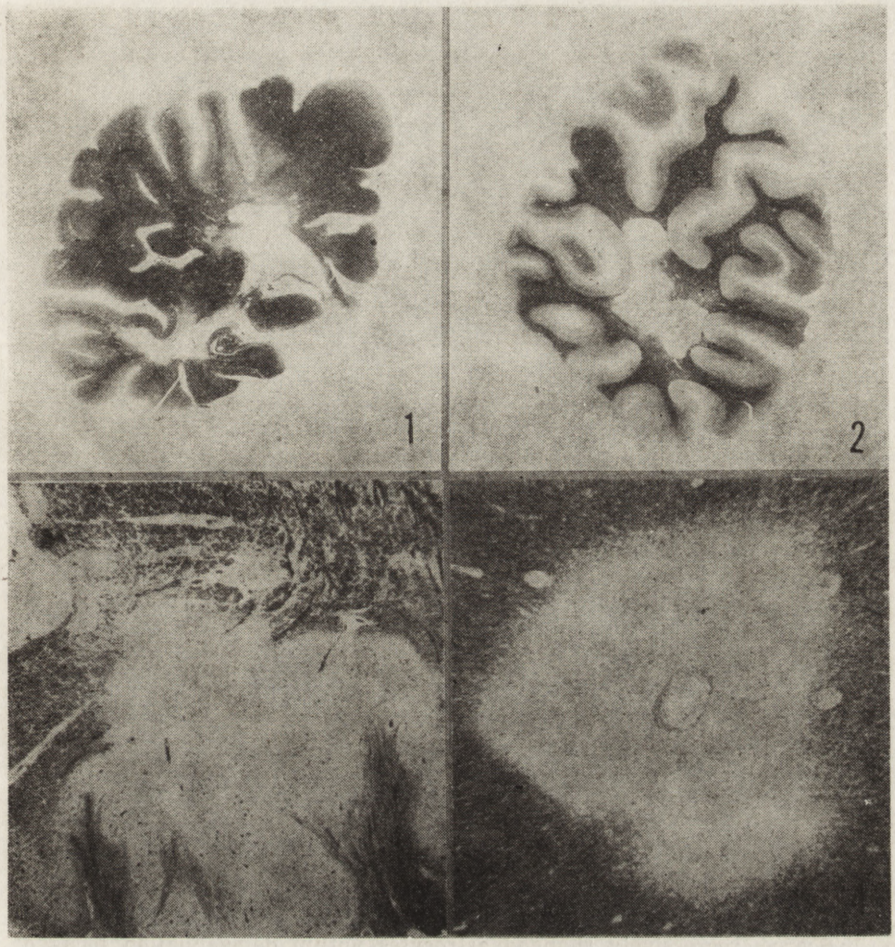

Fig. 1. Numerous demyelination foci within the white matter of the cerebral hemisphere. Klüver-Barrera. Magn. glass

Ryc. 1. Liczne ogniska demielinizacyjne w istocie białej półkuli mózgu. Klüver-Barrera. Pow. lupowe

Fig. 2. Confluent, sharply limited demyelination foci in the white matter of occipital lobe. Klüver-Barrera. Magn. glass

Ryc. 2. Zlewające się, ostro ograniczone ogniska demielinizacyjne w istocie białej płata potylicznego. Klüver-Barrera. Pow. lupowe

Fig. 3. Small demyelination focus in the vicinity of inferior olive. Klüver-Barrera. × 25

Ryc. 3. Małe ognisko demielinizacyjne w sąsiedztwie oliwki dolnej. Klüver-Barrera. Pow. 25 ×

Fig. 4. Perivascular demyelination in the cerebellar white matter. Klüver-Barrera. × 25

Ryc. 4. Okołonaczyniowe ognisko demielinizacji w istocie białej móżdżku. Klüver-Barrera. Pow. 25 ×

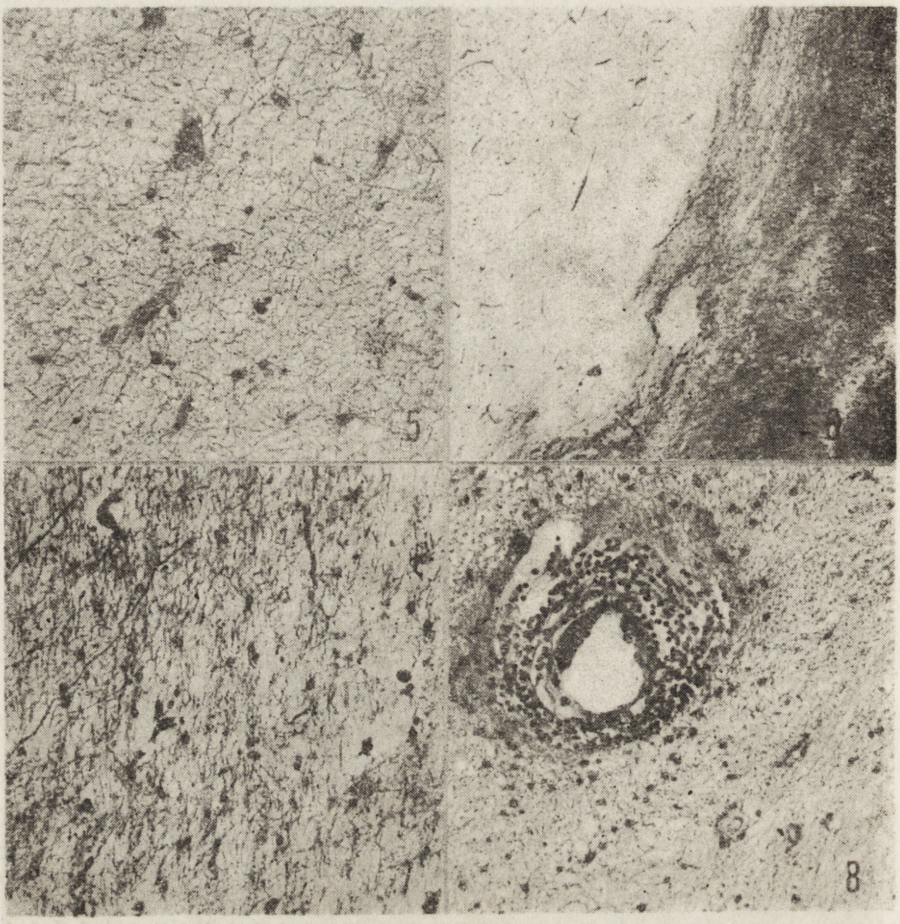

Fig. 5. Scarce, regressively changed astrocytes within demyelination focus of the occipital lobe. H—E. × 200

Ryc. 5. Ubogokomórkowe ognisko demielinizacyjne w płacie potylicznym. H—E. 200 ×

Fig. 6. Dense fibrogliosis within demyelination focus in the temporal lobe. Kanzler-Arendt. × 60

Ryc. 6. Zbita glejoza włóknista w ognisku demielinizacyjnym w płacie skroniowym. Kanzler-Arendt. Pow. 60 ×

Fig. 7. Reduced number of axon cylinders within an old demyelination focus. Bielschowsky. × 200

Ryc. 7. Przerzedzenie włókien osiowych w starym ognisku demielinizacyjnym. Bielschowsky. Pow. 200 ×

Fig. 8. Perivascular lymphocytic infiltration within an old demyelination focus. H-E. × 200

Ryc. 8. Okołonaczyniowy naciek limfocytarny w starym ognisku demielinizacyjnym. H-E. Pow. 200 ×

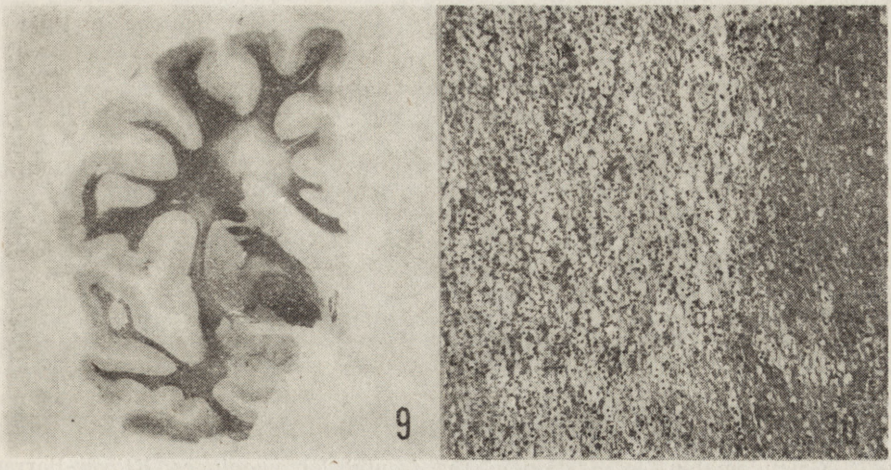

Fig. 9. Sharp demyelination focus, surrounded by diffuse palor of myelin within the frontal white matter. Numerous small ill-limited demyelination foci spread throughout the hemispheric white matter. Globus pallidus and neighbouring internal capsule involved by neoplastic growth. Klüver-Barrera. Magn. glass

Ryc. 9. Ostro zarysowane ognisko demielinizacji w istocie białej płata czołowego otoczone wieńcem spłowienia mieliny. Ponadto widoczne drobne, nieostre ogniska demielinizacyjne rozsiane w istocie białej półkuli. Gałkę bladą i przylegającą część torebki wewnętrznej zajmuje ognisko nowotworowe. Klüver-Barrera. Pow. lupowe

Fig. 10. Recent demyelination focus with rarefied tissue and poor myelin staining. H-E. × 100

Ryc. 10. Świeże ognisko demielinizacyjne w płacie czołowym z rozrzedzeniem utkania i słabym wybarwieniem osłonek. H-E. Pow. 100 ×

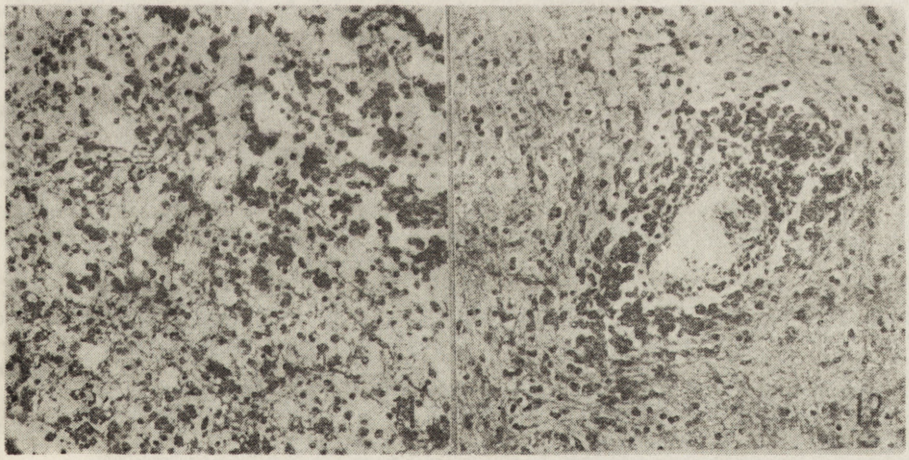

Fig. 11. Nests of granulomatous infiltration within a focus of recent demyelination. Klüver-Barrera. × 400

Ryc. 11. Gniazda komórek nacieku granulomatycznego w świeżym ognisku demielinizacyjnym. Klüver-Barrera. Pow. 400 ×

Fig. 12. Perivascular granulomatous infiltration within unchanged white matter. Klüver-Barrera. × 200

Ryc. 12. Okołonaczyniowy naciek granulomatyczny w niezmienionej istocie białej. Klüver-Barrera. Pow. 200 ×

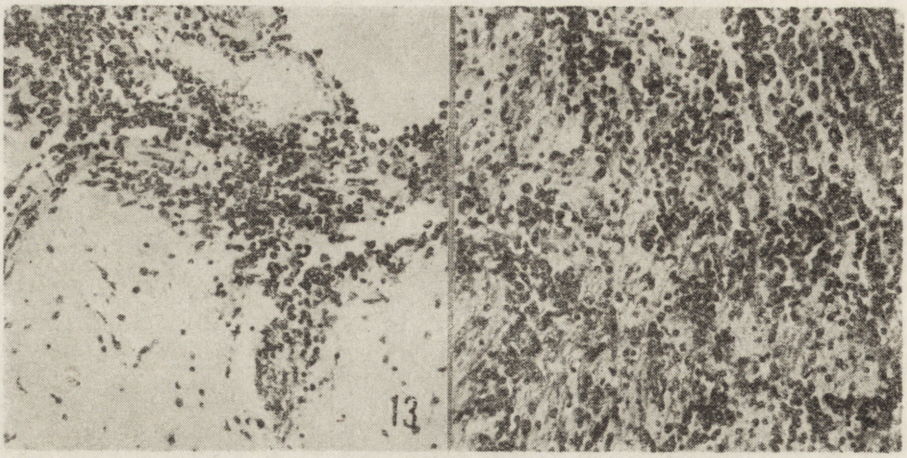

Fig. 13. Granulomatous infiltration of the frontal leptomeninges. Klüver-Barrera. × 200

Ryc. 13. Granulomatyczny naciek w oponie miękkiej płata czołowego. Klüver-Barrera. Pow. 200 ×

Fig. 14. Dense granulomatous infiltration in the internal capsule. Klüver-Barrera. × 200

Ryc. 14. Zbity naciek granulomatyczny w torebce wewnętrznej. Klüver-Barrera. Pow. 200 ×

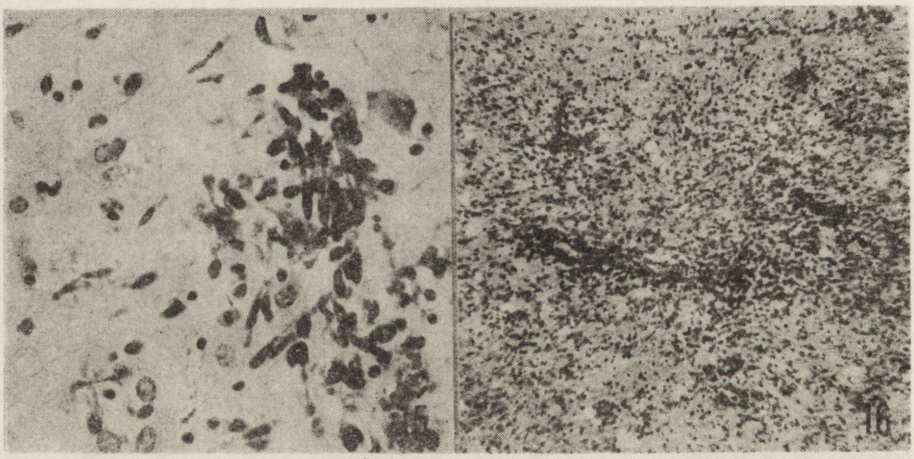

Fig. 15. Nodular microglial infiltration in the cerebral cortex. Numerous rod-shaped cells. Klüver-Barrera. × 400

Ryc. 15. Grudkowy naciek mikrogleju w korze mózgu. Liczne komórki pałeczkowate. Klüver-Barrera. Pow. 400 ×

Fig. 16. Diffuse granulomatous infiltration within the occipital white matter. H-E. × 100

Ryc. 16. Rozlany naciek granulomatyczny w istocie białej płata potylicznego. H-E. Pow. 100

their limits was rarefied to various extents (Fig. 10). Part of them contained diffuse infiltration composed of histiocytes, lymphocytes and immature plasma cells (Fig. 11). A great proportion of myelin sheaths and axons within these foci were spared, being separated by agglomera-

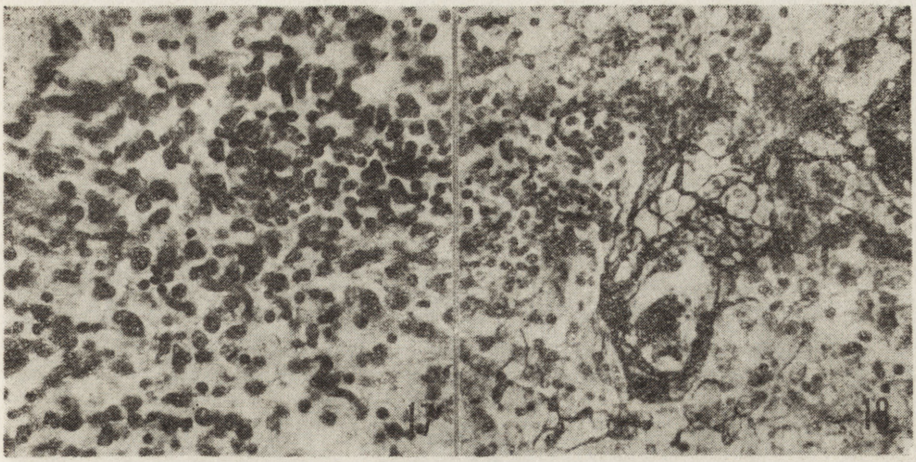

Fig 17. Cellular elements of the granulomatous infiltration within the occipital white matter. H-E. × 400

Ryc. 17. Elementy komórkowe rozlanego nacieku granulomatycznego w istocie białej płata potylicznego. H-E. Pow. 400 ×

Fig. 18. Extravascular argentophilic fibers' network between the cells of granulomatous infiltration in the occipital white matter. Griedley. × 400

Ryc. 18. Pozanaczyniowa sieć włókien srebrochłonnych pomiędzy komórkami nacieku granulomatycznego w istocie białej płata potylicznego. Griedley. Pow. 400 ×

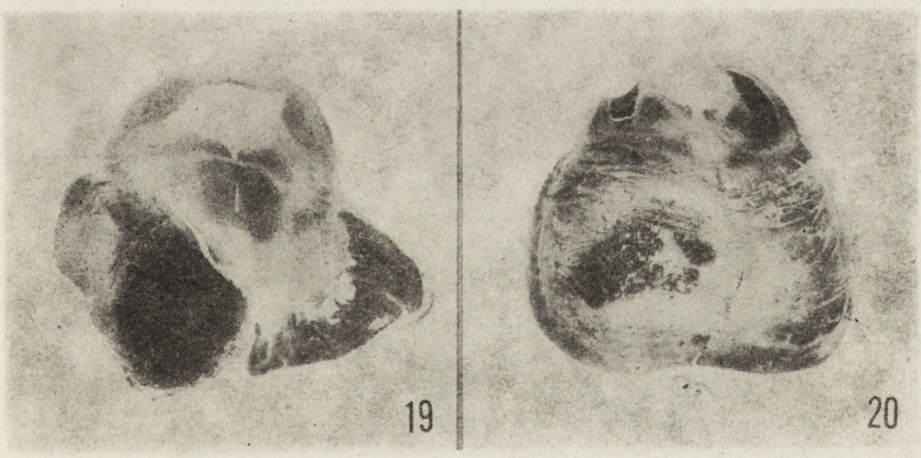

Fig. 19. Well-limited tumor involving the left basal portion of the midbrain. Klüver-Barrera. Magn. glass

Ryc. 19. Ostroograniczony guz zajmujący podstawną część śródmózgowia. Klüver-Barrera. Pow. lupowe

Fig. 20. Pontine portion of the tumor, surrounded by diffuse rime of demyelination. Klüver-Barrera. Magn. glass

Ryc. 20. Mostowa część guza otoczona rozległym pierścieniem demielinizacji. Klüver-Barrera. Pow. lupowe

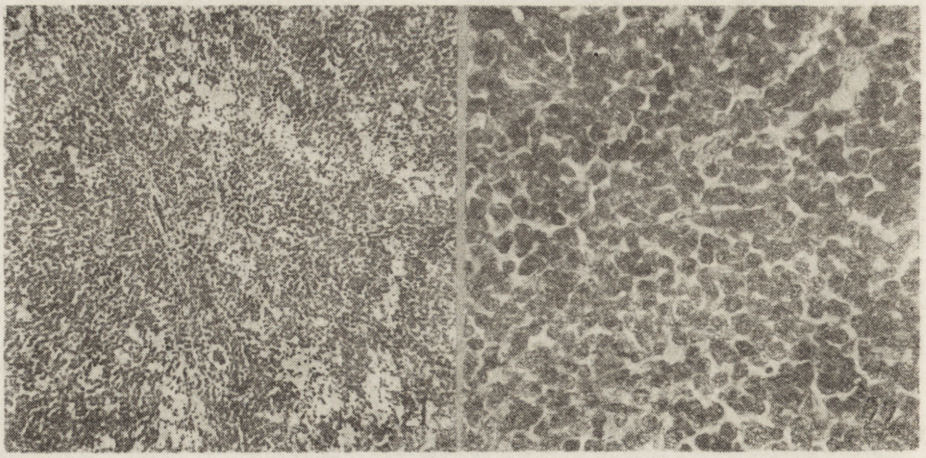

Fig. 21. General cellular characteristics of the midbrain tumor. Klüver-Barrera. × 100

Ryc. 21. Utkanie bogatokomórkowego guza śródmózgowiowego. Klüver-Barrera. Pow. 100 ×

Fig. 22. Predominating type of cells from the midbrain tumor. Numerous mitotic figures. Giemsa. × 400

Ryc. 22. Podstawowy typ komórek guza zlokalizowanego w śródmózgowiu. Liczne figury mitotyczne. Giemsa. Pow. 400 ×

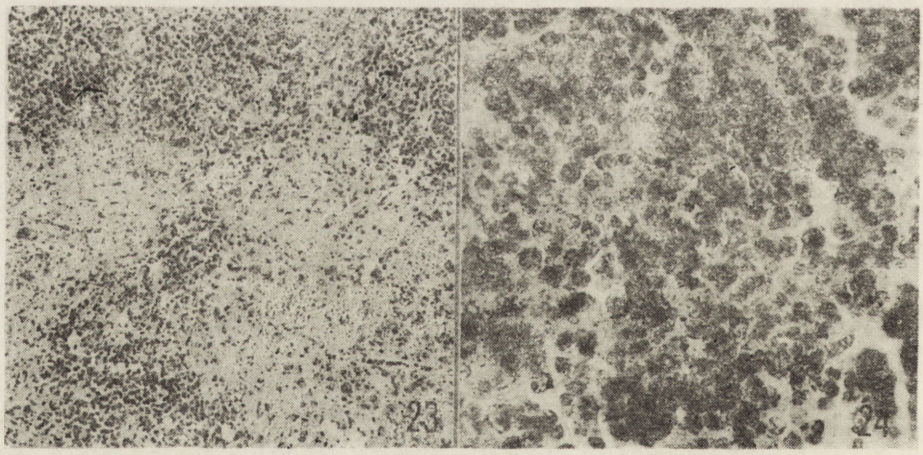

Fig. 23. Large nests of cells with light foamy cytoplasm within the pontine portion of the tumor. Klüver-Barrera. × 100

Ryc. 23. Rozległe gniazda jasnych komórek o piankowatej cytoplazmie w utkaniu mostowej części guza. Klüver-Barrera. Pow. 100 ×

Fig. 24. Cytoplasm of foamy cells filled with lipid granules and droplets. Oil red. × 400

Ryc. 24. Cytoplazma piankowatych komórek wypełniona ziarnistościami tłuszczu. Czerwień oleista. Pow. 400 ×

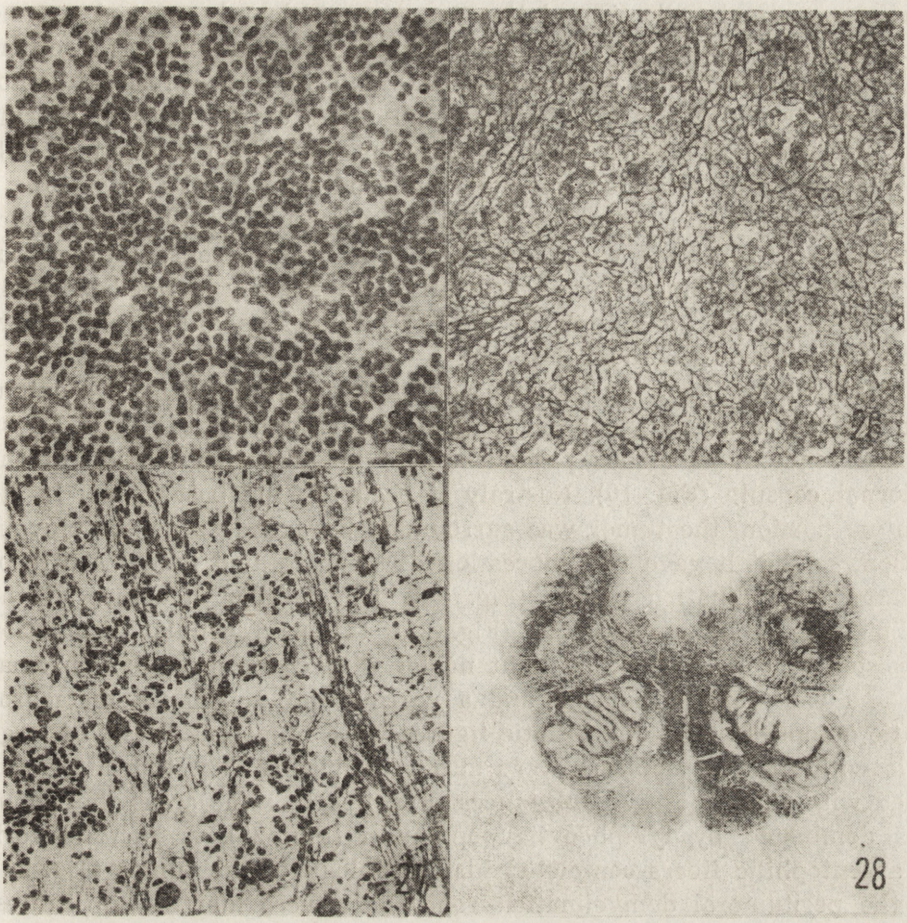

Fig. 25. Area of tumor with prevalent lymphocytic cellular population. H-E. × 400

Ryc. 25. Pole guza o przewadze utkania limfocytarnego. H-E. Pow. 400 ×

Fig. 26. Dense network of argentophilic fibers separating groups of cells and individual cells of the tumor. Griedley. × 200

Ryc. 26. Gęsta sieć włókien srebrochłonnych otaczająca grupy komórek i pojedyncze komórki guza. Griedley. Pow. 200 ×

Fig. 27. Loose infiltration within peritumoral demyelination. Klüver-Barrera. × 100

Ryc. 27. Luźne utkanie nacieku w obszarze okołoguzowej demielinizacji. Klüver-Barrera. Pow. 100 ×

Fig. 28. Medulla with secondary demyelination of the left pyramidal tract and small demyelination focus in the vicinity of right inferior olive. Klüver-Barrera. Magn. glass

Ryc. 28. Opuszka z wtórną demielinizacją lewej piramidy i drobnym ogniskiem demielinizacyjnym w sąsiedztwie prawej oliwki dolnej. Klüver-Barrera. Pow. lupowe

tes of abnormal cells. The latter were most densely packed around blood vessels. Similar perivascular infiltrates were also present in otherwise unchanged white matter (Fig. 12) and in some areas of leptomeninges (Fig. 13) and also between the fibers of the internal capsule (Fig. 14). Cortical perivascular infiltrations contained numerous cells with features of activated microglia and rod-shaped cells (Fig. 15).

In various brain areas, spreading perivascular infiltrations were present (Fig. 16). They were composed of multiform cells, with irregular nuclei, containing coarse chromatin and sharply defined nucleoli, and a scanty rim of cytoplasm (fig. 17). Numerous argentophil fibers accumulated between them (Fig. 18).

Brain tumor involving the left side of the midbrain (Fig. 19), extended rostraly towards the basal ganglia, infiltrating here corpus Luysi, medial segment of globus pallidus and a neighbouring portion of the internal capsule (Fig. 10). Rostraly it occupied the basal pons. In its pontine portion, the tumor was surrounded by a wide zone of demyelination, so that it gave the impression of being located in the centre of a large demyelination plaque (Fig. 20). The cellular structure of the tumor was rather monotonous (Fig. 21). Its main cellular population consisted of cells with large light nuclei with coarse granular chromatin and well formed nucleoli and a narrow rim of slightly metachromatic cytoplasm (Fig. 22). Mitotic figures were numerous. Nests of large cells with foamy cytoplasm were also present (Fig. 23); the latter was filled with sudan-positive substances (Fig. 24). In some areas the tumor was dominated by lymphocytic population (Fig. 25). A rich network of argentophilic fibers completed histopathology of the tumor (Fig. 26). In the peritumoral demyelination zone there were identical infiltrates. However, they were much looser and contained numerous hypertrophied astrocytes (Fig. 27). The secondary demyelination of the left pyramidal tract was seen on the level of medulla (Fig. 28).

DISCUSSION

In the present case, two pathological processes, namely demyelination and neoplasia coexist together. These two basic processes are accompanied by widespread changes characteristic of granulomatous encephalitis. Demyelination reveals structural features of multiple sclerosis. Morphology of most demyelination foci suggests a chronic process. Lack of clinical manifestations of demyelinating diseases is similar to that observed in case of Schaumburg et al. (1972). In that respect it differs from cases, described by Ulrich and Würthrich (1974) and Wildi (1974) in which clinical symptomatology of M.S. lasted for several years. A clinically silent course of multiple sclerosis is well known from numerous descriptions, first of all from that of Mackay and Hirano (1967).

Neoplasia in our case shows the features of a process known under the names of reticulosarcoma or microglioma (Schaumburg et al. 1972, Vuia, Hager 1973; Adams 1975; Reznik 1975; Zimmermann 1975), which has been included into a large group of malignant lymphomas (Lennert 1975; Lukes, Collins 1975). Similar to other descriptions, tumor in our case is multifocal in its nature (Schaumburg et al. 1972; Jellinger et al. 1975; Adams 1975) and in all probability seems to be a primary cerebral process. However, this supposition is based only on the general autopsy observations. Its univocality is greatly limited due to the lack of microscopic examination of body organs. Concomitance of lung and brain involvement by lymphoblastic processes seems to be a relatively frequent feature (Liebow 1972).

Neuropathological findings in our case raise the question of a possible relationship between neoplasia of reticulosarcomatous nature and the inflammatory process of the type of granulomatous encephalitis. Features of both processes were found to be present. In this sense the case seems to be supportive of the opinion, that delimitation between granulomatous encephalitis and reticuloblastosis of neoplastic nature is by no means sharp (Vuia 1966; Osetowska 1980). This inclines us to consider the case described by Dymecki et al. (1966) as belonging to the same group as ours, despite of its diagnosis as an inflammatory process.

The fundamental question is the possible interdependence of two pathological processes observed in the presented case. Simple incidental coexistence cannot be ruled out. However, concomitance of neoplastic growth of lymphoma type with demyelinating process is exceptionally rare compared with the total number of neuropathologically verified cases of multiple sclerosis. Lymphoma's precipitating action on the development of the demyelinating disease, although theoretically possible, seems doubtful in our case in the light of advancement of myelin damage (mostly old inactive plaques). The third possibility is the influence of demyelinating process with an immunopathological pathogenic background on the neoplastic proliferation of lymphoreticular system. The appearance of malignant lymphomas in cases of immunological defects and in the course of diseases with immunological pathomechanisms is well known (Cammorata et al. 1963; Brand, Marinkowich 1969; Gregory, Highes 1973; Wishart 1973; Jellinger et al. 1979). Also convincing is the high rate of lymphomas, involving predominantly the central nervous system in the recipients of transplanted organs, with a longlasting immunosuppression (Penn, 1978). The question which remains unanswered is whether immunopathology present in cases of multiple sclerosis can be a factor precipitating lymphoma's growth (Tourtellotte et al. 1980; Sindie et al. 1980).

Pathogenic factor or factors of unknown nature common for both

Table 1. Cases from the literature

No	Author(s) (year)	Clinical diagnosis	Disease duration	Therapy: Steroids, immunosuppression	Demyelination		Neoplastic growth or granulomatous process
					Type	Advancement	
1.	H.H. Schaumburg et al. 1972	brain tumor anamnesis: *Neuritis optica retrobulbaris*	7 months	X-rays	disseminated	old	reticulum cell sarcoma (focal)
2.	J. Ulrich, R. Würthrich 1974	M.S.	7 years	Cortizone, ACTH Azathioprine	disseminated	old	reticulum cell sarcoma (multifocal)
3.	K. Wildi 1974	M.S.	9 years	Cortizone, Azathioprine	disseminated	old	reticulum cell sarcoma (multifocal)
4.	P. Castaigne et al. 1974	brain tumor	4 months	Synacthène, ACTH	disseminated	recent	reticulum cell sarcoma (focal)
5.	J. Gruner 1961	M.S. *Mycosis Fungoides*	9 years	ACTH	disseminated	old	cerebral reticulosis (multifocal)
6.	S. Haddenbrock et al. 1962	S.M.	2 years	ACTH	disseminated	old	phlébite granulomateuse (multifocal)
7.	J. Dymecki et al. 1966	multifocal neurological syndrome with dementia	9 years	ACTH(?)	disseminated	old	granulomatous encephalitis (multifocal)

processes — demyelination and neoplasia was suggested by Castaigne et al. (1974). The suggestion was based on the simultaneousness of both pathological processes, which does not seem to be the case in our patient.

Ulrich and Würthrich (1972) in the discussion of their case stressed very strongly the possible pathogenic role of immunosuppressive therapy — steroids and mostly azathioprine, being more and more often used in the treatment of multiple sclerosis (Aimard et al. 1980). Use of azathioprine therapy was also common in cases of brain lymphomas of recipients of transplanted kidneys (Penn 1978). On the other hand, quite a proportion of brain reticulum-cell sarcomas were described in people without immunosuppressive therapy (Zimmermann 1975; Jellinger et al. 1975). Concomitance of brain lymphoma with multiple sclerosis in a patient submitted to azathioprine therapy is known from only one description in literature (Ulrich, Würthrich 1972), although this factor was also involved in a case described by Jellinger et al. (1979). However, one cannot ignore the fact that in 5 of 7 cases collected from literature, immunosuppressive therapy was in use (Table 1). This was not true in our case.

REFERENCES

1. Adams H.: The classification of microgliomatosis with particular reference to diffuse microgliomatosis. Acta neuropath., (Berl.) 1975, 6 (Suppl.), 119—123.
2. Aimard G., Confavreux C., Devic M.: Long-term immunosuppressive treatment with azathioprine in multiple sclerosis. A 10 year trial with 77 patients. In: Progress in multiple sclerosis research. Eds. H. J. Bauer, S. Poser, G. Ritter. Springer. Berlin, Heidelberg, New York 1980, 371—375.
3. Brand M., Marinkowich V. A.: Primary malignant reticulosis of the brain in Wisbott-Aldrich syndrome. Arch. Dis. Child., 1969, 44, 536—542.
4. Cammarata R. J., Rodman G. P., Jensen W. M.: Systemic rheumatic disease and malignant lymphoma. Arch. intern. Med., 1963, 111, 330—337.
5. Castaigne P., Escourolle R., Brunet P., Gray F., Rouques C., Le Bigot P.: Reticulosarcome cérébral primitif associé à de lésions de sclerose en plaque. Rev. Neurol., 1974, 130, 181—188.
6. Dymecki J., Wald I., Gadomska B.: Granulomatous encephalitis with multifocal demyelination. Neuropat. Pol., 1966, 4 (Supl.), 729—734.
7. Gregory M. C., Hughes J. T.: Intracranial reticulum cell sarcoma associated with immunoglobulin deficiency. J. Neurol. Neurosurg. Psychiat., 1973, 36, 769—776.
8. Gruner J.: Two cases of cerebral reticulosis. In: Encephalitides. Eds. L. van Bogaert, J. Radermecker. Elsevier, Amsterdam 1961, 194—203.
9. Haddenbrock S., Guazzi G. C., Torck P., Moya G.: Phlébite granulomateuse sur sclérose en plaque. Acta Neurol. Psychiat. Belg., 1962, 62, 111—134.
10. Jellinger K., Radaskiewicz Th., Slowik F.: Primary malignant lymphomas of the central nervous system in man. Acta neuropath., (Berl.), 1975, 6 (Suppl.), 95—102.

10a. Jellinger K., Kothbauer P., Weiss R., Sunder-Plassmann E.: Primary malignant lymphoma of the CNS and polyneuropathy in a patient with necrotizing vasculitis treated with immunosuppression. J. Neurol., 1979, 220, 259—268.
11. Lennert K.: Morphology and classification of malignant lymphomas and so-called reticuloses. Acta neuropath., (Berl.) 1975, 6 (Suppl.), 1—16.
12. Liekow A. A., Arrington C. R. B., Friedman P. J.: Lymphoid granulomatosis. Human Path., 1972, 3, 457—536.
13. Lukes R. J., Collins R. D.: New approaches to the classification of lymphomata. Brit. J. Cancer, 1975, Suppl. 2, 1—28.
14. Mackay R. P., Hirano A.: Forms of benign multiple sclerosis. Arch. Neurol., 1967, 17, 588—600.
15. Osetowska E.: Tissue neuropathology of viral and allergic encephalitides. Pol. Med. Publ., Warsaw 1980, 241—247.
17. Penn I.: Malignancies associated with immunosuppressive or cytotoxic therapy. Surgery 1978, 83, 492—502.
17. Reznik M.: Pathology of primary reticulum cell sarcoma of the human central nervous system. Acta neuropath., (Berl.) 1975, 6 (Suppl.), 91—94.
18. Schaumburg H. H., Plank C. R., Adams R. D.: The reticulum cell sarcoma-malignant group of brain tumours. A consideration of their clinical features and therapy. Brain 1972, 95, 199—212.
19. Scherer H. J.: La glioblastomatose en plaque. J. Belg. Neurol. Psychiat., 1938, 38, 1—17.
20. Sindic C., Cambiaso C. L., Masson P. L., Laterre E. C.: Clinical relevance of the determination in the cerebrospinal fluid with special reference to multiple sclerosis patients. In: Progress in Multiple Sclerosis Research. Eds. H. J. Bauer, S. Poser, G. Ritter. Springer. Berlin, Heidelberg, New York 1980, 176—179.
21. Tour tellotte W. W., Potvin A. R., Potvin J. H., Ma B. J., Baumhefner R. W., Syndulko K.: Multiple sclerosis de novo central nervous system IgG synthesis: measurement, antibody profile, significance, eradication and problems. In: Progress in Multiple Sclerosis Research. Eds. H. J. Bauer, S. Poser, G. Ritter. Springer. Berlin, Heidelberg, New York 1980, 106—116.
23. Ulrich J., Würthrich R.: Multiple sclerosis: reticulum cell sarcoma of the nervous system in a patient treated with immunosuppressive drugs. Europ. Neurol., 1974, 12, 65—68.
23. Vuia O.: Das solitäre und multifokale polymorphe Granulom im Nervensystem. Arch. Psychiat. Ztschr. ges. Neurol., 1966, 208, 309—325.
24. Vuia O., Hager H.: Primary cerebral blastomatous reticulosis. Clinical, pathohistological and ultrastructural study. J. neurol. Sci., 1973, 19, 407—423.
26. Wildi R.: Cit. by Ulrich J. and Würthrich R., 1974 (pos. 22).
26. Wishart J. M.: Reticulosarcoma of the vulva complicating dermatomyositis treated by immunosuppression. Proc. Roy. Soc. Med., 1973, 66, 330.
27. Zimmermann H. M.: Malignant lymphomas of the nervous system. Acta neuropath., (Berl.) 1975, 6 (Suppl.), 69—74.

NIEMA DEMIELINIZACJA Z OGNISKOWYM I ROZLANYM ROZROSTEM RETYKULOBLASTYCZNYM MÓZGU

Streszczenie

Przedstawiono przypadek 70-letniej kobiety uprzednio zdrowej, która przez okres dwóch ostatnich lat leczona była z powodu spastycznego zapalenia oskrzeli. W ostatnim roku życia wystąpiły zmiany charakterologiczne z równoczesnymi

zaburzeniami równowagi i zawrotami głowy. Po przebyciu, na dwa miesiące przed śmiercią, ostrej choroby gorączkowej, zaczął narastać w sposób postępujący zespół neurologiczny, który ostatecznie przyjął postać typowego zespołu Webera z niezbornością. Symptomatologia i przebieg kliniczny sugerowały rozpoznanie procesu nowotworowego bądź naczyniopochodnego, umiejscowionego w okolicy śródmózgowia. Na sekcji mózgu stwierdzono guz w części podstawnej konarów mózgu i mostu. Badanie mikroskopowe wykazało obecność licznych, głównie starych ognisk demielinizacyjnych we wszystkich strukturach mózgowia, z ich wyraźną przewagą w półkulach mózgowych. Procesowi temu towarzyszył uogólniony rozrost retykuloblastyczny oraz uformowany guz o cechach *lymphoma malignum*. Rozrost retykuloblastyczny występował zarówno w ogniskach demielinizacyjnych jak i poza nimi. Omówiono możliwą współzależność patogenetyczną dwu procesów, wysuwając hipotezę o wyzwalającej roli zaburzeń immunopatologicznych, związanych z klinicznie niemą chorobą demielizacyjną.

НЕМАЯ ДЕМИЕЛИНИЗАЦИЯ С ОЧАГОВОЙ И РАЗЛИТОЙ РЕТИКУЛОБЛАСТИЧЕСКОЙ ГИПЕРПЛАЗИЕЙ ГОЛОВНОГО МОЗГА

Резюме

Представлен случай 70-летней здоровой женщины, которая в течение двух последних лет была лечена по поводу спастического бронхита. В последнем году жизни выступили характерологические изменения с одновременными нарушениями равновесия и головокружениями. После перенесения, на два месяца перед смертью, острой лихорадочной болезни начал нарастать прогрессирующим способом неврологический синдром, который в полсдние времья принял форму типичного синдрома Вебера с атаксией. Клиническая симптоматология и клиническое течение подсказывали диагноз неопластического или ангиогенного процесса, локализированного в области среднего мозга. Во время обдукции головного мозга констатировано опухоль в базальной части ножек мозга и варолиева моста. Микроскопное исследование проявило наличие многих, главным образом старых демиелинизационных очагов во всех структурах головного мозга, с их отчетливым преобладанием в мозговых полушариях. Этому процессу сопутствовала обобщенная рекитулобластическая пролиферация и сформированная опухоль с признаком lymphoma malignum. Ретикулобластическая пролиферация выступала как в демиелинизационных очагах, так и вне их. Авторы обсудили возможную патогенетическую взаимозависимость обоих процессов, выдвигая гипотезу о высвобождающей роли иммунопатологических расстройств, связанных с клинически немой демиелинизационной болезней.

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