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Pseudotumorous clinical and neuroimaging manifestation of multiple sclerosis

Multiple sclerosis (MS) is autoimmunological, inflammatory disease of the central nervous system (1). The pathological process, as a definition, involves multifocal lesions but great variability of clinical syndromes facilitates atypical manifestations, being a probable reason for diagnostic pitfalls. Additionally, accurate diagnosis may be postponed in cases of unifocal manifestation, e.g. with retrobulbar optic neuritis, as the diagnosis of MS requires pathological lesions to be dispersed in time and space. We present an infrequent constellation of clinical symptoms which appeared in a female patient whose symptoms of unifocal central nervous system lesion, with seizure and intracranial hypertension, led to inappropriate diagnosis of neoplasmatic brain tumor.

CASE DESCRIPTION

Twenty-three years old female patient, at the age of 19 had undergone corrective operation of juvenile idiopathic thoracic spinal scoliosis of 3rd degree, with implantation of steel stabilization device CD Horizon (Fig. 1). Since the age of 17, she occasionally experienced paresthesias in upper and lower limbs, which were regarded as radiculopathy. Four years after the operation, she developed transitory syndrome of left leg paresis with headache, nausea with vomiting and vertigo (suggesting of focal lesion with intracranial hypertension). After a month, she experienced left hemiparesis which progressed to left hemiplegia, accompanied by visual acuity disturbances. Additionally, she observed episodic left limbs spasticity which once progressed to generalized tonic-clonic seizure. EEG showed focal, episodic delta-theta discharges in fronto-temporal regions, with secondary generalization. CT scanning showed heterogenically enhancing lesion, 22 mm in diameter, in right patietal lobe with surrounding oedema suggestive of neoplasmatic brain tumor (2) (Fig. 2). Differential diagnosis comprised demyelinization but there was no time and space dispersion of symptoms, no oligoclonal bands in cerebrospinal fluid nor disturbances of visual evoked potentials. During hospitalization we introduced symptomatic treatment (Mannitol, Dexamethasone, Furosemid) with prominent regression of motor deficits. Consequently, stereotactic biopsy of brain lesion was performed. Tissue specimen showed glial cell proliferation which was interpreted as possibly suggesting of astrocytoma grade 2. Unexpectedly, clinical syndrome was not progressing and consecutive CT monitoring showed that the lesion was not evolving as it is seen in brain tumors, i.e. regression of hypodensive area (oedema), increase in density of the lesion (gliosis) and lack of contrast enhancement.

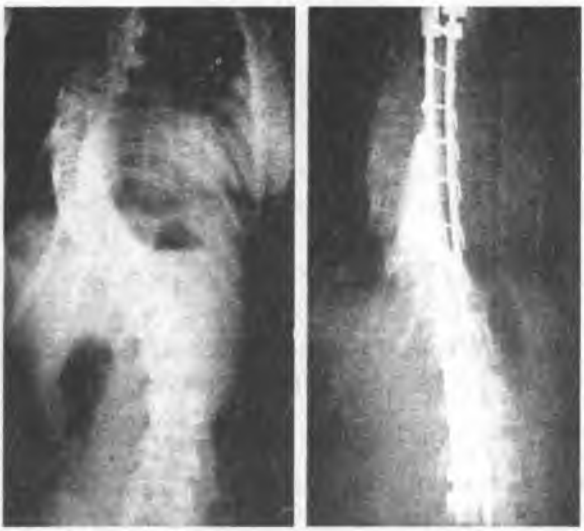

Fig. 1. X-ray of the spine before and after operation

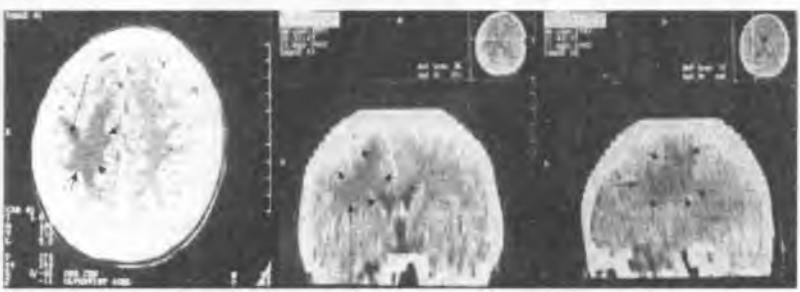

Fig. 2. Head CT revealed small, 22 mm in diameter, lesion with contrast enhancement with surrounding area of local brain swelling, in right parietal lobe, suggestive of brain tumor

Because of serious and still unconfirmed suspicion of brain tumor and good general condition of the patient, finally the decision to remove spinal stabilization device was made, to enable magnetic resonance imaging (MRI) (Fig. 3–5). First examination revealed three T2 and FLAIR hiperintensive lesions localized in periventricular area of parietal white matter, with no contrast enhancement, and one similar lesion in frontal lobe. During three consecutive years the patient experienced transient neurological symptoms comprising of limbs parestesias, disturbances of micturition, compromising of visual acuity and vertigo. Two more MRI examination were performed, showing multiple, diffuse, confluent T2, FLAIR, PD hiperintensive lesions with no contrast enhancement localized in periventricular white matter of both hemispheares and atrophic corpus callosum. Visual evoked potentials revealed prolongation of P100 latency (right eye – 118 ms, left eye – 111 ms). Somatosensory evoked potentials showed dysfunction of proprioceptive sensation in left lower limb. Serological screening for boreliosis was negative. Thorough clinical workup was performed to exclude other conditions that could resemble MS in neuroimaging studies (3). The latest MR imaging fulfils the McDonald's criteria for multiple sclerosis (4). Since then time and space symptom dispersion was comfirmed.

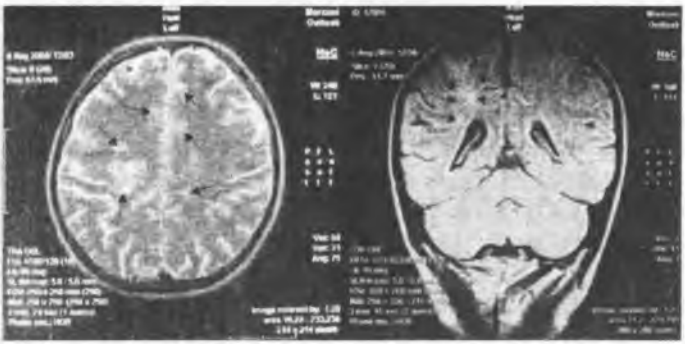

Fig. 3. Head MRI showed T2 and FLAIR hiperintensive lesions, with no contrast enhancement, localized in periventricular white matter of both parietal and frontal lobes

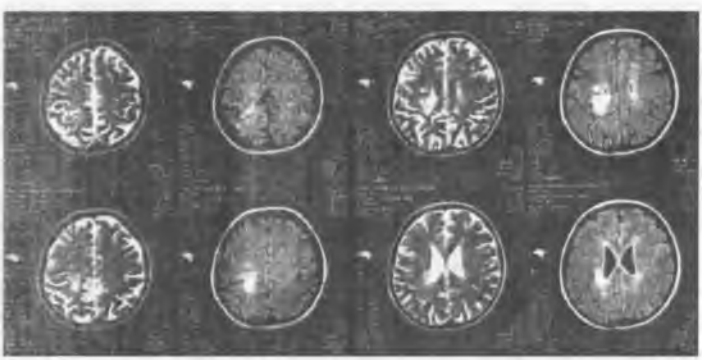

Fig. 4. Consecutive MR imaging showed multiple, diffuse and confluent T2, PD and FLAIR hiperintensive lesions, without contrast enhancement, in periventricular white matter of both hemispheres and atropic corpus callosum

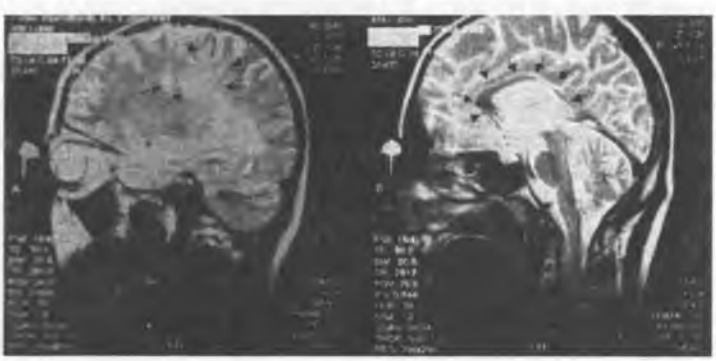

Fig. 5. T2-weighted MR images in saggital planes, showing lesion in periventricular area of right parietal lobe and atrophy of corpus callosum

DISCUSSION

Multiple sclerosis often presents history of central nervous system lesions dispersed in time and space. The diagnosis is made mostly on clinical ground with MR being the most important additional

tool. In rare cases MS presents with symptoms that strongly suggest focal lesion, forcing thorough differential diagnosis with neoplasmatic tumors, vascular diseases and infections being among others the most prominent conditions (5). Tumefactive lesions often lead to brain biopsy which often can unexpectedly aid unconsistent results as demyelinating lesion may resemble astrocytoma (6). In our patient, clinical course, with rapidly progressing neurological deficits, focal seizures and concordant CT neuroimaging studies, lack of oligoclonal bands in cerebrospinal fluid nor changes in visual evoked potentials, made the suspicion of brain tumor highly probable. In early stage of the disease special clinical background (ferromagnetic spine stabilization device) made MR imaging contraindicated. CT imaging is known to be less sensitive to MS lesions and most often no lesion is visualized at all. In our case CT showed significant oedema around relatively small lesion which is typical rather of a neoplasmatic origin than of demyelination (6). For above reasons CT imaging had to be followed by brain biopsy in order to state diagnosis and begin appropriate treatment. Decision to perfom biopsy is inevitable in such condition as non-invasive diagnostic tools can aid misleading results and lead to unnecessary operation (7). Interestingly enough tissue specimen obtained was suggestive of astrocytoma grade 2. In this case the result was certainly wrong which underlines necessity of appropriate specimen preparation and analysis as demyelination and low grade gliomas share the same histological features (8). Thus what was the reason to delay operation as the diagnosis seemed to be achieved? After the focal presentation suggestive of intracranial hypertension patient received symptomatic treatment, inluding glicocorticosteroids. Her symptoms rapidly recovered and after 2–3 weeks she felt functionally healthy and denied surgical treatment. Such treatment response can be encountered in demyelination but not in neoplasmatic lesions (9). Continuous CT imaging showed no progression and no mass effect in the follow-up period. Such an observation reflects evolution of the lesion from the stage of acute plaque to chronic tissue scar with prominent gliosis, which is not encountered in brain tumors. When the spine stabilization device was removed, MRI revealed typical periventricular lesions with no contrast enhancement and additionally one larger lesion which was responsible for focal presentation and had been sampled. Despite oligosymptomatic course, continuous CT and MR imaging helped exlude neoplasmatic tumor. In this case final diagnosis of MS was time consuming and reached on the basis of: 1) clinical features of MS, with time and space dispersion of neurological symptoms, 2) neuroimaging diagnostic criteria by McDonald (4), 3) pathological disturbances of visual evoked potentials.

The presented case leads to the conclusion that in rare cases differential diagnosis of MS and brain tumor can be challenging. Traditional clinical diagnostic criteria should still form the basis but may not be enough in focal presentation, especially when additional diagnostic tools show unconsistent results.

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SUMMARY

Small percentage of MS patients paradoxically present with focal neurological symptoms that may suggest vascular or neoplasmatic diseases. In most cases such tumefactive lesions demand biopsy to confirm diagnosis. We have presented the 23-year-old female patient with idiopatic thoraco-lumbar scoliosis, whose initial syndrome suggested etiopathology of brain neoplasmatic tumor. This preliminary clinical diagnosis was supported by CT examination that revealed single hypodense lesion with large oedema in the right parietal lobe. The multiple sclerosis was finally diagnosed traditionally on the basis of prolonged clinical observation, and confirmed on MR imaging that demanded removing of spine stabilization device.

Kliniczna i radiologiczna manifestacja stwardnienia rozsianego naśladująca guz nowotworowy mózgu

U niewielkiego odsetka chorych stwardnienie rozsiane może paradoksalnie zamanifestować się klinicznymi objawami jednoogniskowego uszkodzenia OUN, co w pierwszej kolejności nasuwa podejrzenia udaru niedokrwiennego lub procesu nowotworowego. W większości przypadków potwierdzenie rozpoznania wymaga wykonania biopsji mózgu. Przedstawiony został przypadek 23-letniej kobiety z idiopatyczną skoliozą piersiowo-lędźwiową III°, u której stopniowo narastające zaburzenia neurologiczne sugerowały rozwój guza mózgu. Wstępna kliniczna diagnoza znalazła potwierdzenie w obrazie CT mózgu, gdzie stwierdzono obecność zlokalizowanego hipodensyjnego uszkodzenia w prawym płacie ciemieniowym z rozległym lokalnym obrzękiem. Prawidłowa diagnoza SM została ustalona sposobem konwencjonalnym, po dłuższej obserwacji przebiegu klinicznego oraz potwierdzona w badaniu MR głowy, co wiązało się z koniecznością usunięcia stabilizatora kręgosłupa.