Pseudotumor Cystic Demyelinating Plaque: Report of Two Cases

Pseudotumor Kistik Demiyelinizan Plak: İki Olgu Sunumu

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Summary: The authors presented two patients with suggestive of intracranial cystic masses in magnetic resonance imaging whose neurologic deficits completely disappeared without surgical treatment.

The first patient was a male teenager who experienced sudden neurologic deficits five years ago and MRI showed a cystic mass in the right posterior parietal lobe. The last during the second attack, disclosed disappearance of this lesion and a new lesion with similar features in left posterior parietal region and another lesion in the middle cerebellar peduncle. Whole spinal cord disclosed by magnetic resonance imaging and also noticed a large upper thoracal lesion suggestive of a demyelinating plaque.

The second patient was a male adult who suffered progressive left hemiparesia for two months. The MRI revealed a right parietal cystic mass with ring enhancement. Stereotactic biopsies were taken from the lesion. The histopathological findings were consistent with demyelinating plaque. The patient showed progressive improvement and he was discharged with advice of further rehabilitation.

The interesting aspect of these two patients was the large cystic cerebral lesions, which suggested neoplastic

Özet: Yazarlar, nörolojik defisitleri herhangi bir cerrahi tedavi uygulanmadan tamamen düzelen ve manyetik rezonans (MR) incelemelerinde intrakranial kistik tümör izlenimi veren iki olgu sunmaktadırlar. Birinci olgu genç bir erkek hasta olup beş yıl önce ani gelişen nörolojik defisitleri nedeni ile çekilen MR' ında sağ posterior parietal lobda kistik lezyon saptandı. Beş yıl sonra geçirdiği ikinci atak sırasında çekilen MR' ında bu lezyonun kaybolduğu ve sol posterior parietal bölgede benzer radyolojik özelliklere sahip yeni bir lezyonun varlığı gözlendi. Ayrıca orta serebeller pedinkülde ve tüm spinal kordun MR incelemesinde üst torakal bölgede demiyelinizan plak izlenimi veren iki ayrı lezyon saptandı.

İkinci olgu ise iki aydır ilerleyici sol hemiparezisi olan yetişkin erkek hasta idi. Kranial MR'ında sağ parietal lobda çevresel kontrast tutan kistik lezyon görüldü. Lezyondan sterotaktik biopsi ile örnekler alındı. Histopatolojik bulgular demiyelinizan plak ile uyumluydu. Bir süre sonra yakınmaları düzelmeye başlayan hasta, rehabilitasyon önerileri ile takibe alındı.

Bu iki hastada ilginç olan, neoplastik patoloji izlenimi veren kistik serebral lezyonların varlığı, buna karşılık bu

pathology. On the contrary, the clinical outcome, radiological disappearance of the lesions and the pathologic findings were consistent with of multiple sclerosis.

Key Words: Brain tumor, cerebral cyst, demyelinating plaque, multiple sclerosis

olgularda klinik gidiş, lezyonların radyolojik olarak kayboluşu ve patolojik olarak multipl skleroz olduğunun gösterilmesidir.

Anahtar Sözcükler: Beyin tümörü, demiyelinizan plak, multipl skleroz, serebral kist

Case 1

A 13 year-old boy had been admitted an another centre in May of 1996 upon 40 days history of headache, vertigo, nausea, left hemihypoesthesia and sudden left hemiparesis. At that time magnetic resonance imaging (MRI) had revealed a subcortical white matter cystic lesion in the right parietal lobe measuring 5x4x4 cm. Medical history dislosed that the complaints had resolved spontaneously in weeks (Figure: 1a,1b).

He was hospitalized in our department in April of 2001 due to weakness and hypoesthesia of the lower extremities. The neurologic examination disclosed paraparesis, bilateral hypoesthesia to the level of T-7 and bilateral extensor plantar responses. Cranial MRI revealed the disappearance

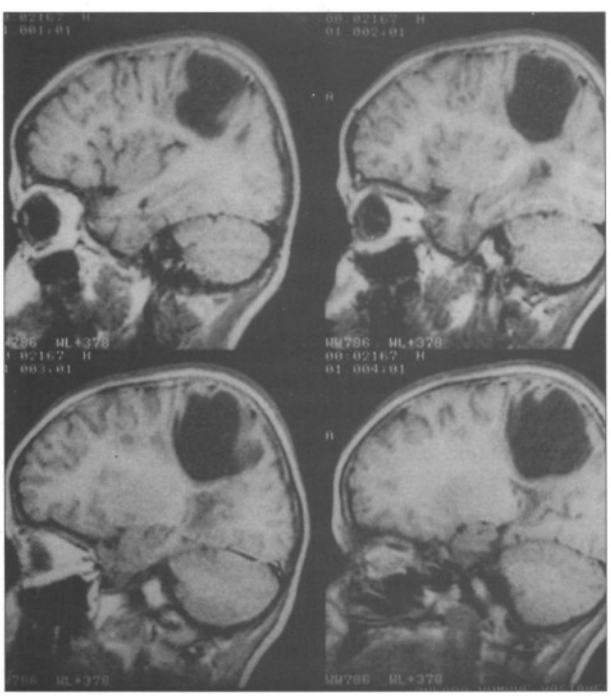

Figure 1a.Precontrast sagittal T-1 W MR imaging showing a cystic lesion in the right parietal lobe.

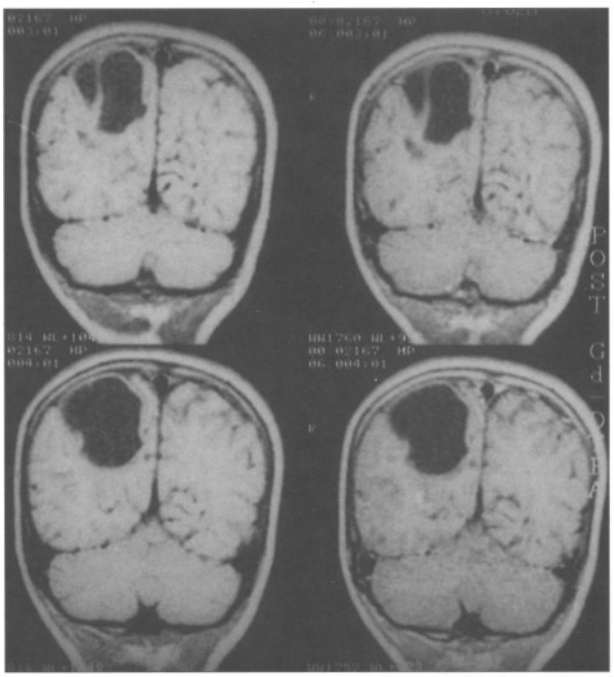

Figure 1b. Postcontrast coronal T-1 W image reveals that the lesion is non- enhancing.

of the cystic lesion in the right posterior parietal region and a new lesion in the subcortical white matter of the left posterior parietal lobe with surrounding encephalomalasic region (Figure 1c). Another lesion was detected, hypointense on T_1-W and hyperintense on T_2-W images measuring 18x15 mm, in the middle cerebellar pedincule. The lesion was an irregular shape with moderate perilesional edema and peripheral enhancement.

The whole spinal axis was examined by MRI and a partially enhancing plaque measuring 12x7 mm at T_1-T_5 was observed (Figure 1d). Serological examination for sarcoidosis, vasculitis, HIV, toxoplasmosis, lyme disease, syphilis and other neurotrophic viruses were negative and the serum level of folic acid, vitamin B_{12}, angiotensin converting enzyme were normal. Oligoclonal band was not found in cerebro spinal fluid (CSF). The

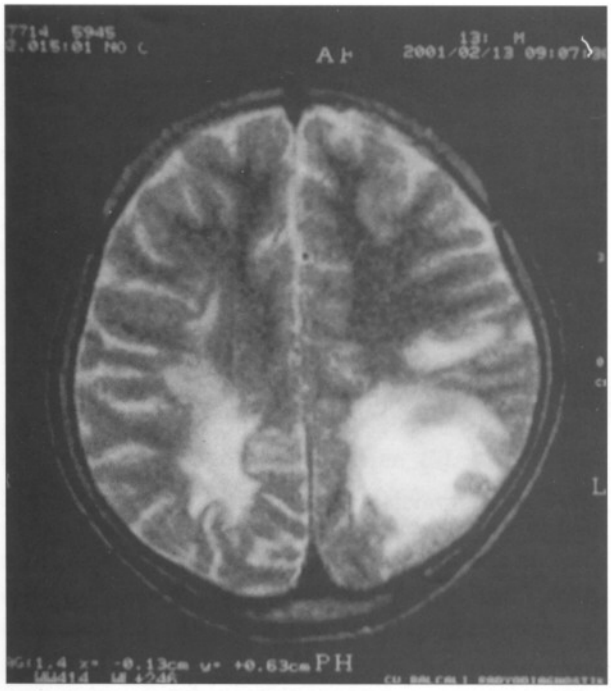

Figure 1c. T-2 W MRI five years later showing a new lesion in the left posterior parietal lobe and right posterior parietal encephalomalasia.

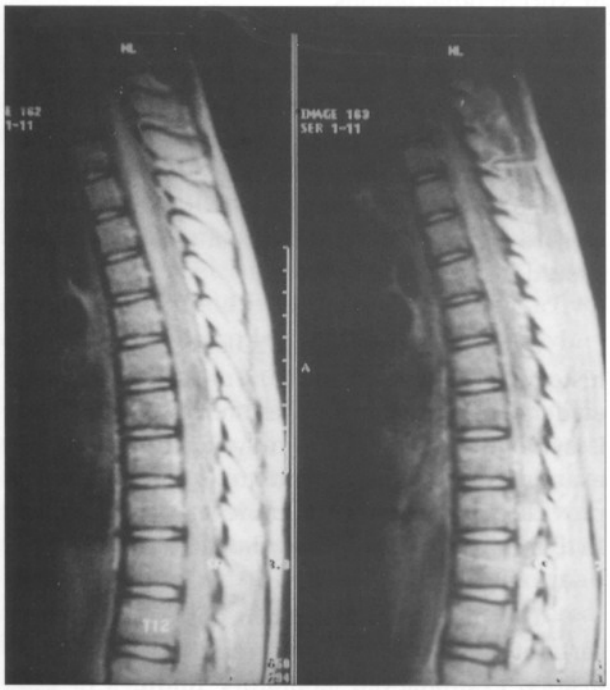

Figure 1d. Sagittal thoracal MRI of the same patient revealing a partially enhancing plaque formation extending from T1 to T5.

neurologic findings completely improved in five days without any treatment and the patient was discharged in good health.

Case 2

A 21 year-old male was admitted to our department with two months history of headache, progressive left hemiparesis. He has been using different addictives for ten years which he quitted since the last two years. The neurologic examination disclosed left hemiparesis of 3/5 with left extensor plantar responses. MRI revealed a subcortical white matter cyst with thin, peripheral enhancement in T1-W images localized in the right parietal lobe (Figure 2a, 2b, 2c). T2-W images showed a hyperintense mass measuring 2x3x4 cm. in the same localization. Serological examinations for sarcoidosis, vasculitis, HIV, toxoplasmosis, Lyme disease, syphilis, other neurotrophic viruses were negative. The serum levels of folic acid, vitamin B_{12}, angiotensin converting enzyme were normal. The toxoplasma IgG was 1/40 (+) in serum. Oligoclonal band was not detected in the CSF. We performed stereotactic biopsy and the pathology specimen showed myelin losses in

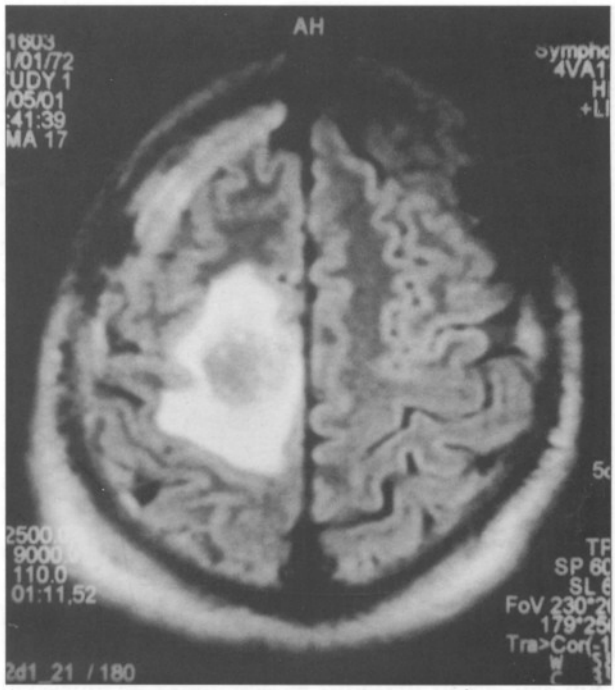

Figure 2 a. Axial T-2 W image of the second patient revealing a hyperintense mass measuring 2x3x4 cm. in the right posterior parietal lobe.

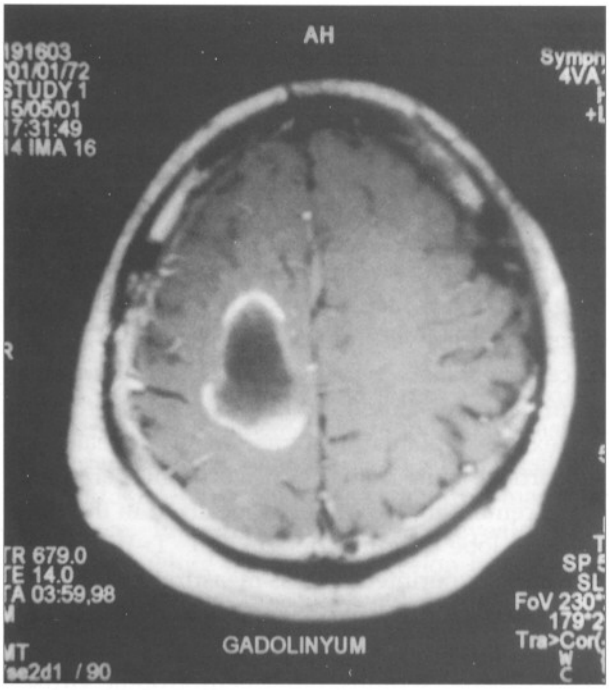

Figure 2 b. Post contrast T-1 W axial and a sagittal images shows the cystic lesion with peripheral enhancement.

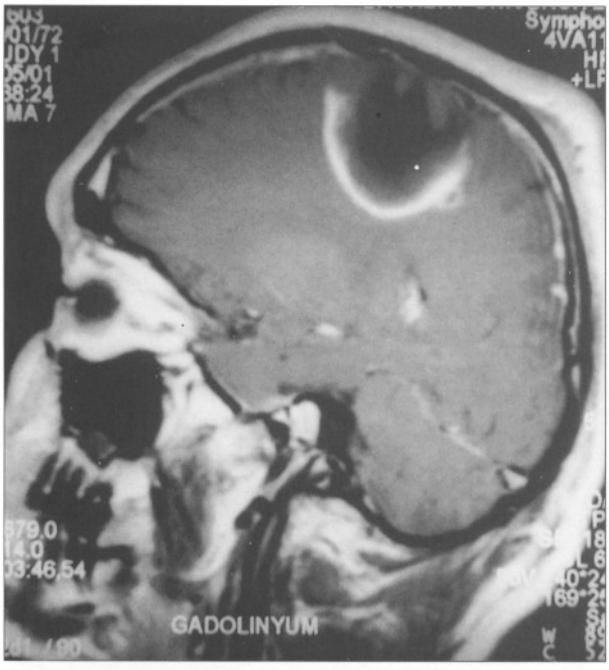

Figure 2 c. Post contrast T-1 W axial and a sagittal images shows the cystic lesion with peripheral enhancement.

myelinisation matrix and there were same myelin fibres in the cytoplasm of histiocytes named foamy cells. His complaints gradually diminished over 8 weeks and the patient was discharged with minimal left hemiparesis.

DISCUSSION

The incidence of Multiple Sclerosis (MS) is about 3-5 cases per 100 000 population yearly and childhood MS is distinctly less common, approximately composing 0.3 - 2 % of all cases (1,18).

The diagnosis of MS is based on the ability to demonstrate, on the basis of history, neurologic examination and laboratory tests, the existence of lesions involving different parts of the central nervous system (CNS). The main criteria for diagnosis of MS as described by Sadiq (19) is as follows:

The clinically definite MS requires either evidence from both history and neurologic examination of more than one lesions or evidence from history of two episodes, signs of one lesion on examination, and evidence from evoked responses or MRI of other lesions.

Laboratory-supported definite MS requires evidence of two lesions in either history or examination. If only one lesion is evident in either of those categories, at least one more lesion must be evident in evoked response or MRI. In addition, cerebro spinal fluid (CSF) IgG content and pattern should be abnormal.

Clinically probably multiple sclerosis is either history or examination, but not both, provide evidence of more than one lesion. If only one lesion is evident by history and only one by neurologic examination, evoked potentials or MRI may provide evidence of one or more lesion in addition. In this category, CSF IgG studies are normal.

The diagnosis of MS can not be made with certainty everytime. The relapsing, remitting history, examination, laboratory findings of CSF, MRI and evoked response are all helpful to diagnosis. However, some cases are diagnosed only at autopsy (19)

The typical MS plaques are often seen as round or ovoid areas near the periventricular and subcortical white matter or in the spinal cord. Lesions in the spinal cord are usually less than two vertebral body segment long, peripherally located and mostly found in the cervical region. The MRI appearance is iso/hypointense on T1-W and hyperintense on T2-W. Contrast MRI is more reliable in identification of the age of the lesions. Solid and ring like enhancing lesions on T1-W can be detected which are not appearent on T2-W (5, 6, 21). The contrast enhancement is thought to favor active demyelination of plaque and a local break-down of the blood-brain barrier. (5). Active MS plaques with inflammatory response may be enhancing with surrounding edema and tumor like mass effects. (4, 15, 17). MS plaques are demonstrated better with flair images than with conventional T2-W images or proton-density images (20).

Magnetization transfer is another tecnique which can demonstrate changes in myelin structure. These tecniques may permit useful differentiation of potential reversible from irreversible lesions to guide and evaluate the results of therapy (12)

Rare different radiologic plaque appearances have been described. CT ring sign which imitates a tumoral appearance is an example (5, 8, 11). In multiple sclerosis, the CT ring sign is very rare and may be difficult to differantiate a plaque from the primary or secondary brain tumors and abscess, ecpecially when there is a mass effect. Gadolinium DTPA ring enhancement of MS lesions is more common in MR and is thought to represent active demylination of the plaque (11).

The other radiologic form is cystic manifestation (3, 9, 10, 21). In the early phase of cyst formation, cystic necrosis around the plaque is observed. Later, remyelination of the central core of the lesion is speculated, as similarities in signal intensity between the core and the normal appearing white matter were partially recovered both on the T1- and the T2-weighted images (3).

The irregular mass lesions with homogeneus or inhomogenous irregular borders and involving the gray matter suggest neoplasm or infection. Active enhancing lesions are more easily misinterpreted as tumor and thus are operated on (21). Multiple cystic MRI lesions were seen in multicentric glioma, multiple metastasis, primary brain tumors, brain abscesses, CNS infections and postvaccination. Progressive multifocal leukoencephalopathy (PML) has clinical and radiological features similar to MS. But PML differs from MS with its rapid unset and the presence of immunodeficiency syndrome (4, 6).

The correlation between clinical status and MRI findings in MS patients is weak and therefore new MR techniques are being developed to increase MRI sensitivity for detecting disease activity and its pathological specificity. Large cystic lesions in MS do not cause symptoms related to the mass effect. Likewise the question of whether different radiologic plaque appearances have any clinical implications has not been answered yet.

MRI is the most reliable diagnostic tool for confirming, assesing the progression of MS, although it is not specific (2, 4).

The clinical relapses are not necessarily related to the apearence of new lesions. Some plaques are very large, measuring 3 cm in diameter or more. They tend to be more diffuse or irregular ring and large plaque is distincly less uncommon (1, 18). The cyst formation is not releated to the degree of neurological impairment. MS in early childhood may present atypically, with symptomatology suggesting diffuse encephalomyelitis, meningeal reaction, brain edema, seizures, impaired consciousness and in some cases take a lethal course (7).

In our first patient neurological impairment has completely resolved while in the second patient neurological impairment improved. In both instances there was no treatment. The interesting features of these cases were the cystic presentation of the MS plaque, its spontaneous disappearance and formation of a new one in the opposite hemisphere in one patient. As to our knowledge a large spinal MS plaque and bilateral cerebral hemispheric cystic and at different times, has not been reported also. Our literature review revealed only three cases of bilateral cystic MS plaques (3, 9, 10, 14).

Our second patient was laboratory definite MS with MRI findings mimicking a atypical brain neoplasm. In many instances cystic MS plaques with ring enhancement led surgical interventions for diagnosis. However, the clinical features and radiological findings do not necessitate a surgical procedure in majority of the patients.

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