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Case Report

Bilateral post-traumatic hygromas in patient with frontotemporal dementia

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ABSTRACT

Background: Frontotemporal dementia (FTD) is a highly disabling neurologic disorder characterized by behavioral alterations and movement disorders, involving patients with a mean age of 58 years. We present a unique case of a patient suffering from FTD who developed post traumatic bilateral hygromas.

Case Description: A 52-year-old male patient, with an history of head trauma 3 months before, was admitted to our department for recurrent motor seizures. Anamnesis was positive for FTD with severe frontal syndrome. Brain computed tomography and magnetic resonance imaging (MRI) showed the typical "knife-blade" appearance of the cortical atrophy associated to bilateral hemispheric hygromas exerting mild mass effect. Brain MRI showed the signs of the cortical and "anti-cortical" vein. The two subdural collections were evacuated through two bilateral burr holes and controlled drainage. Despite anti-epileptic drugs therapy, in the early postoperative period, the patient presented further tonic-clonic seizures. The patient showed progressive recovery and was transferred to the neurorehabilitation center. After 6-month follow-up, he completely recovered.

Conclusion: In FTD, severe cortical atrophy leads to space increase between arachnoid and pia mater that could affect the anatomical integrity especially after trauma, with possible development of hygromas. The coexistence of radiological findings of the cortical vein and sign of the "anti-cortical" vein can make difficult an exact differential diagnosis between a primitive hygroma and a Virchow hygroma from resorption of previous blood collection. Surgical treatment may be indicated in selected patients, but it is burdened by higher postoperative risks compared to the general population.

Keywords: Cortical veins, Dementia, Hygromas, Seizures, Subarachnoid space

INTRODUCTION

Frontotemporal dementia (FTD) is a severe dementia involving patients in the fourth–fifth decade, with a mean age of onset at 48 years old and with a prevalence, regardless of gender, of about

15/100.000 individuals. It is a highly disabling disease with a prognosis of 3–10 years from the onset. Exitus is mainly associated with causes secondary to frontal involvement causing progressive limitation of movements till forced bed stay which may lead to pneumonia, cardiovascular, and respiratory failure. Clinically, it is possible to find signs and symptoms related to frontal (behavioral alterations and movement disorders) and temporal atrophy (speech disorders with mixed aphasia and memory alterations). Magnetic resonance imaging (MRI) and PET allow to document the severity of cortical atrophy. We present an adult patient presenting the unique association of FTD and bilateral hygromas that rapidly worsened the symptoms related to FTD.

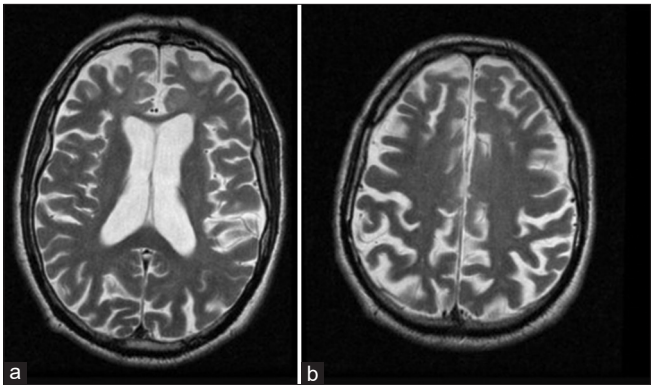

Figure 1: Brain MR T2-weighted axial images showed a typical frontotemporal cortical atrophy and the absence of hygromas.

CASE ILLUSTRATION

History and physical examination

A 52-year-old male patient suffering from FTD with severe frontal syndrome (kleptomania, restlessness, and impulsivity), alteration of attention and memory, semantic aphasia, echolalia, and bladder and bowel incontinence was admitted at our department for recurrent motor seizures. Three months earlier, he was referred to the emergency department for a head trauma related to a seizure episode. He had performed brain MRI after trauma, which showed no acute relevant signs [Figure 1]. Anti-epileptic drugs (AEDs) were administered (levetiracetam).

Brain computed tomography (CT) showed the typical "knife-blade" appearance of the cortical atrophy associated to bilateral hemispheric hygromas 20 mm thick and exerting mild mass effect [Figure 2].

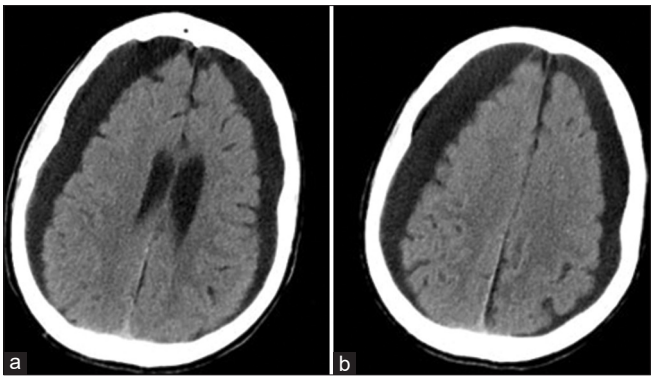

Figure 2: Brain computed tomography scan axial images 3 months after head trauma showed the typical "knife-blade" appearance of the cortical atrophy associated to bilateral hemispheric hygromas 20 mm thick, exerting mild mass effect.

Axial T1-weighted gadolinium-enhanced MRI sequences showed the signs of the cortical vein and "anti-cortical" vein: the first is typical of subdural fluid collections, the latter indicates sub-arachnoid space enlargement [Figure 3].

The patient underwent two bilateral parietal burr holes as well as slow controlled drainage of the subdural collections. Unfortunately, in the early postoperative period, the patient presented further tonic-clonic seizures, despite the treatment with AEDs. An immediate postoperative brain CT scan showed a satisfying drainage of the subdural collections and a thin sylvian subarachnoid hemorrhage [Figure 4]. The patient showed progressive recovery, and he was referred to a neurorehabilitation center. After 6-month follow-up, he completely recovered.

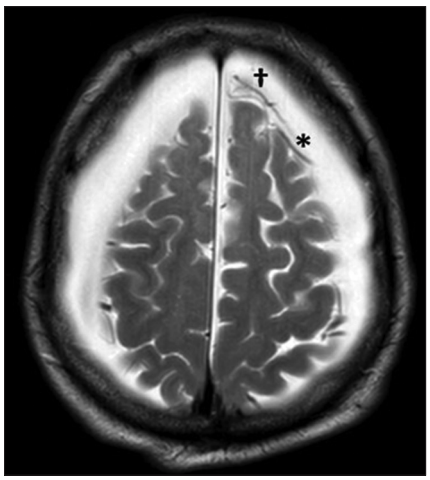

Figure 3: Brain MR T2-weighted axial images showed the coexistence of the cortical vein sign (black asterisk) and the "anti-cortical" vein sign (black obelisk).

DISCUSSION

FTD

Cortical atrophic degeneration is a progressive and irreversible disease that slowly reduces the functionality of the frontal and temporal lobes as confirmed by the CT or MRI imaging and by reduced accumulation of the 18-FDG of

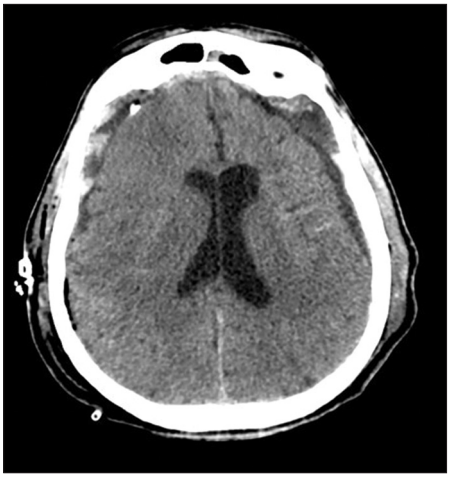

Figure 4: Brain computed tomography scan axial images showed a satisfying drainage of the subdural collections and a thin sylvian subarachnoid hemorrhage.

PET study.[14] Brain CT scan shows a conspicuous reduction of the gyri volume, associated to other common findings typical of cortical atrophy likewise ex-vacuous hydrocephalus and loss of white-grey matter substances separation. MR is a sensitive imaging which allows to highlight the degree of gyri atrophy and the greater representation of the sulci in the affected areas. MRI is useful the differential diagnosis among the several patterns of atrophy disease, associated with different disease phenotypes.[5]

Pathophysiology

Although the phenotype is predominantly sporadic, in 30% of cases, there may be a characteristic genotypic predisposition (e.g., MAPT tau-gene in FTLD-tau). Characteristic findings of this disease are silvery intracytoplasmic inclusions, called Pick bodies, present in atrophic areas and straight filamentous intraneural aggregates of tau protein <30 millimicrons in size.

Clinical presentation and management

Clinical features include signs and symptoms related to frontal and temporal atrophy. The most frequent and usually early diagnosed is the frontal syndrome characterized by behavioral alterations, apathy, movement disorders (pyramidal and extrapyramidal), and disinhibition. In the temporal syndrome, on the other hand, there are speech disorders with mixed aphasia, depending on the hemisphere involved, and memory, at least at the beginning, episodic memory (unlike Alzheimer's disease).[3] During the FTD's workout, the use of neuropsychological tests, such as Mini Mental State Examination, may provide significant data. We compared the scores obtained from the pre and postoperative tests and as expected. In both cases, the tests support the diagnosis of severe pathological dementia (being the scores below the cutoff of 24/30) since the drainage of the hygromas cannot improve the dementia-related deficits. Patients with prevalent behavioral disorders usually have bilateral symmetrical or predominant right frontal cortex atrophy, while patients with prevalent speech disorders may initially experience asymmetric atrophy.

FTD and hygromas

The severe atrophy that takes place approximately symmetrically in both cortexes inevitably leads to an increase in the subarachnoid space that is associated with a greater tension of bringing veins and arachnoid trabecula, and to a possible excessive distention of the thinnest layer of this membrane that is most vulnerable to continuous stresses. From this comes the risk of development of micro-ruptures with the formation of breaches which enlarges overtime causing CSF leakage from the subarachnoid space at higher pressure to the subdural space at lower pressure. This would justify the development of bilateral hygromas which may progressively increase with gradual enlargement of the fluid collection and can contribute to symptom's worsening, as described in our case.

Primitive subdural hygroma is defined as a fluid collection in the subdural space secondary to post-traumatic lacerations of the arachnoid membrane and passage of CSF into the subdural space which differs from subdural hematoma due to its remarkable albuminic content and the absence of the two typical membranes (parietal and visceral). The degree of cerebral atrophy along with cranial morphology would affect (pulling force) on arachnoid trabecula. Brain atrophy helps to increase the virtual space between the inner cranial table and pia mater producing intracranial negative pressure, constant at each site (Boyle law). When the distance is greater than the length of the trabecula, the risk of stratification of the inner layer of the dura mater increases, the subdural space becomes real and sufficient to be filled with fluids (blood and liquor)[10] Remote intra and extraparenchymal blood collections caused by a hyperdrainage of CSF after burr holes evacuation represents a rare complication and have already been reported by Kim *et al*.[8] It can involve both the ipsilateral and contralateral hemispheres probably due to excessive stretching of the bridge veins or secondary to sudden parenchymal expansion.[1] Brain self-regulation is compromised in degenerative disease and it is further altered after prolonged compression caused by hygromas. The rapid reduction of intracranial pressure may lead to hemodynamic alterations such as hyperperfusion and hyperemia.[15]

There are few cases of bilateral hygromas described in the literature, nevertheless none of these is related to severe frontotemporal cortex bilateral atrophy. Bilateral hygromas

are often found in spontaneous intracranial hypotension (SIH),[9,17] but in the case, we described there were no obvious neurological signs of SIH or radiological signs such as CSF leakage through dural defects, reduction in CSF volume or leptomeningeal enhancement after gadolinium administration.[2] In rare cases, subdural hygromas may be secondary to resorption and/or liquefaction phenomena of a primitive subdural hematoma (Virchow's hygromas), characterized by elevated concentration of albumin which remains an important predictive value, useful for the differential diagnosis.

The coexistence of radiological signs typical of a collection between dura and arachnoid (sign of the cortical vein and typical in hygromas) and of an increase in the space between arachnoid and cerebral cortex (the "anti-cortical" vein sign and typical of grey matter atrophy) was in the present case highlighted.[12] In recent years, it has been suggested the diagnostic role to the sign of the cortical vein in other pathologies such as possible prognostic factor[18] and/or predictive neurological deterioration in some patients with acute stroke.[11] Although the sign of the cortical vein has in the past been used to distinguish the subdural space from an enlarged subarachnoid space on MRI, according to some authors, this sign would not always be effective for the exact diagnosis between cortical atrophy and subdural hygroma, and our case could fit into this last case.[7]

Based on the hypothesis that voluminous hygromas may cause irritative phenomena on the cortex,[13,19] equally attention should be paid to those cases in which a rapid reduction in CSF volume, even of an iatrogenic nature, leads to secondary epileptic seizures and dysregulation of the CSF flow, including venous congestion and encephalopathy syndrome.[4,6,16,22]

Finally, postsurgical epilepsy with cranial shaking may lead to excessive traction and rupture of some bridging veins (already under tension and stretching due to such an important cortical atrophy), favoring blood diffusion to the subarachnoid space, through a previous arachnoid breach, irritating the cortex and self-maintaining epileptic events. The surgical procedure should be performed simultaneously and slowly with mini-craniectomies or through minimally invasive technique[20,21] and special attention should be paid to slow and controlled drainage to allow to reach the desired reduction of liquor pressure onto the gyri, avoiding risks of complications.

CONCLUSION

In FTD, severe cortical atrophy leads to an increase in the space between arachnoid and pia mater that could affect the anatomical integrity of the first membrane especially after trauma, with possible development of hygromas. The coexistence of radiological findings of the cortical vein and sign of the "anti-cortical" vein can make difficult making exact differential diagnosis between a primitive hygroma and a Virchow hygroma from resorption of the previous blood collection. The evacuation of the collection must be careful, slow, and controlled. In consideration of the high risks of complications due to the modification of precarious balances in fragile patient population, a careful surgical indication is warranted and paucisympomatic patients should be excluded from the study.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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