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INCREASED INTRACRANIAL PRESSURE IN A CASE OF SPINAL CERVICAL GLIOBLASTOMA MULTIFORME

ANALYSIS OF THESE TWO RARE CONDITIONS

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SUMMARY - The authors describe a rare case of increased intracranial hypertension consequent to a spinal cervical glioblastoma multiforme in a young patient. They analyse the physiopathology of intracranial hypertension in spinal tumors and the rarity of such kind of tumor in this location, and its clinico-pathological aspects.

KEY WORDS: intracranial hypertension, spinal glioblastoma multiforme, young patient.

Hipertensão intracraniana em caso de glioblastoma multiforme da medula cervical: análise dessas duas raras condições

RESUMO - Os autores descrevem um raro caso de hipertensão intracraniana conseqüente a glioblastoma multiforme situado na medula cervical de uma paciente jovem. Analisam a fisiopatologia de hipertensão intracraniana em tumores medulares e a raridade desse tipo de tumor nessa localização, assim como seus aspectos clínico-patológicos.

PALAVRAS-CHAVE: hipertensão intracraniana, glioblastoma multiforme espinhal, paciente jovem.

Increased intracranial pressure is mainly consequent to encephalic intracranial processes besides systemic conditions[3]. Only rarely intracranial hypertension may be due to spinal cord tumors[8,9,14]. Besides its rarity this condition arises controversial arguments as to its physiopathology. The glioblastomas may arise in several parts of the central nervous system. They are more frequently present in the hemisphere chiefly in the frontal and temporal lobes. They may also exist in the cerebellum and brain stem. However they arise rarely in the spinal cord where they may be primary or metastatic[20]. They usually occupy the cervical or thoracic segments and have been either spherical or elongated[21].

In this paper we aim at discussing a conjunction of two rare conditions as increased intracranial pressure and a spinal cervical glioblastoma multiforme in order to analyse the physiopathology of the first and clinico-pathological aspects of the latter.

CASE REPORT

EMA, a 26 years old female patient from Pacajus (Ceará) was hospitalized in August 1, 1991 at the University Hospital for severe headache, numbness in the left leg and right hemithorax and paresis of the right leg. Her disease has begun on May 9, 1991 with numbness in the left leg and right hemithorax. On July 21 she also began complaining

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of severe frontal headache associated with vomiting, weakness, giddiness and gait disturbance and on August 1st she was hospitalized. There was no history of contact with tuberculosis, purulent meningitis, viral infection, or contact with pets. Her familial history was unrelevant. At the admission the patient was in a regular status, eupneic, afebrile, hydrated, acyanotic, icteric (++/6+), oriented and cooperative. Her BP was 120/80 mmHg, pulse 68, temperature 36.7º C and respiratory rhythm 20/min. There was nuchal rigidity. The pupils were isocoric, but there was an impairment of the left photomotor and consensual reflexes. The thorax was symmetrical. The heart rate was regular, the bullae were sound and there was a Hamman sign without however murmur. There was also a slight decrease of the murmur in the inferior third of the right lung. The abdomen was normal. The neurological examination showed, at that time, an oriented patient with normal cortical functions. There was a paresis of the left III, VI and VII cranial nerves, a bilateral papilledema, a decrease of force in the right lower limb, an axial ataxia, hyperreflexia in the lower and upper limbs with right Babinski, right Hoffmann and bilateral patellar clonus. There was a tactile and pinprick hypoesthesia in the left lower limb and in the right submammilar area. With this picture a hypothetical diagnosis of meningitis (tuberculosis or cryptoccocosis?) or a brain stem tumor was proposed. This way the patient was treated for intracranial hypertension with corticosteroids and for a possible meningitis with cephalosporin and metronidazol. The patient evolved with slight improvement of her headache. The laboratory results showed: hematocrit 36%, hemoglobin 10.7g%, leucocytes 6300, VHS 29 mm, urea 30 mg/100ml, creatinine 0.7 mg/100ml, Na^+ 142 mg/100ml, K^+ 4.2 mEq/l, and platelets 74000/mm^3. As the patient presented a papilledema, no lumbal punction was carried out. Subsequently the patient complained again of headache and developed convulsions. At the 5thday from admission, a CT scan was done that showed a moderate supratentorial hydrocephalia with normal IV ventricle. As the patient did not improve with antibiotics and as leucogram showed normal, a treatment for tuberculosis (rifampin, hydrazide and pyrazinamide) was started, despite undefined evidences for tuberculosis.

In the 7th day, as the intracranial hypertension did not improve, a system of ventricular continous drainage was performed with subsequent improvement of the patient. The study of this ventricular CSF showed 1 cell/mm^3 (100% mononuclear), glucose of 87.5 mg/dl and protein of 38 mg/dl. The electrophoresis showed pre-albumin 4.0%, albumin 63.7%, alfa-1 4.0%, alfa-2 5.0%, beta-1 8.6% and gamma 14.7%). There were no neoplastic cells and the CSF culture was negative for pyogenic germs, BK and fungi. The immunofluorescence and Weinberg reaction were negative for cysticercosis. The patient improved of headache and convulsions. In the 12nd day the patient presented signs of drug intoxication (icterus, nausea, vomiting and increased levels of transaminases) so that the antituberculosis drugs were stopped. In the 14th day a new CT scan showed again the same hydrocephalia. At the 19th day the patient became more torporous with paresis of the left III, VI and VII cranial nerves, with right patellar areflexia, and a Brown-Séquard-like syndrome (left leg hypoesthesia and right leg paresis with bilateral Babinski and patellar clonus). A ventriculoperitoneal shunt was then performed. At the 25th day she also presented sphincter incontinence, and a bilateral paresis of the V and XII cranial nerves. ANF, LE cells, HBSAg, FTA and ELISA tests for HIV were negative. The clinical picture became more severe with irregular respiration, involuntary movements of the face and right upper limb, loss of force and hyporeflexia in the upper limbs. As the time, the results of the CSF culture BAAR were negative. At the 35th day she also presented dysphasia and positive bilateral Lasègue, and paresis of the left XI cranial nerve. A CT performed at the 47th day with thin slices of the brain stem, showed again a supratentorial hydrocephalia with normal IV ventricle and hyperdense lesion of intrasellar localization. A new ventriculoperitoneal shunt was performed in the 49thday, but afterwards the patient presented bradicardia, somnolence, irregular respiration and unconsciousness. At the 53rd day the pupils were anisocoric and slightly photo-reactive. At that moment paresis of the left III, V, VI, VII and XII cranial nerves were firmly established and there was a tetraplegia and areflexia of the upper limbs. The patient died for septicemia (Citrobacter freundii) at the 57th day (Table 1).

Anatomopathological Report - A female corpse, with an apparent age of 25 and 30 year, presenting areas of hypostasis in the dorsal and abdominal region. Bilateral mydriasis and wrinkling of the palmar face of the hands and feet. The vertebral canal was open according Franco's technique. Macroscopically, the cervical portion of the spinal cord was increased in volume (4.0 x 2.0 cm). The microscopical study of this region revealed a tumor that consisted of a glial heterotypical neoplasia with hyperchromatic cells sometimes aligned in pallisades. There were areas of extensive necrosis. These characteristics led to the conclusion of a spinal glioblastoma multiform (Fig. 1). The cranial cavity was also open and examinated. The encephalon weighed 1365.2g. It had an increased volume with an edema and focal areas of softening and hemorrhage corresponding to the shunting valve. Besides this, there was a herniation of cerebellar amygdalae and hydrocephalia. The microscopical analysis of the encephalon, brain stem and cerebellum did not show neoplastic changes.

COMMENTS

The case we present has two peculiarities that may become this report interesting: on one side we are confronted with a patient presenting a syndrome of increased intracranial pressure

Table 1. Steps in the Evolution of the Patient: May 9,1991 to July 21, 1991. Motor signs, headache, vomiting..

Admission day	
01	Signs of increased intracranial pressure. Impairment of the left III - VI - VII cranial nerves. Incomplete Brown-Séquard syndrome.
05	Convulsions. CT Scan: moderate supratentorial hydrocephalia.
07	Set up of continuous CSF drainage.
12	Symptoms of drug intoxication (anti-TB treatment). Signs of increased intracranial pressure.
14	New CT-Scan: moderate supratentorial hydrocephalia.
19	Set up of a ventriculo-peritoneal shunt. Stupor. Impairment of the left III - VI - VII cranial nerves. Brown-Séquard syndrome.
25	Sphincter disturbances and abnormal facial movements. Impairment of bilateral V and XII cranial nerves. Respiratory disturbances.
35	Dysphasia. Bilateral Lasègue. Impairment of the left XI cranial nerve.
47	CT-Scan (with thin slices of the brain stem): supratentorial hydrocephalus and normal IV ventricle.
49	Set up of a new ventriculo-peritoneal shunt. Bradicardia, chaotic respiration. Coma.
53	Impairment of the III - V - VI - VII - XII cranial nerves. Tetraplegia.
57	Exitus letalis.

consequent to a spinal tumor and on the other side this tumor is a gliobastoma multiforme of very rare occurrence in this spinal location.

The glioblastoma represent around 50% of all primary gliomas; while they are more frequent in the frontal and temporal lobes, they are rare in the brain stem and still rare in the spinal cord where they represent about 7.5% of all intramedullary gliomas and barely 1.5% of all spinal tumors[4,7,20,24]. The cervical and thoracic region are the most affected, and especially the high cervical region[4,5,11,15-17,20,24] as found in our case. The spreading of this tumor to the brain stem may possibly explain the involvement of cranial nerves. Unhappily an accurate histological analysis of slices from the brain stem was not carried out to confirm or exclude this hypothesis. The histological analysis of somes slices however did not show abnormalities. In any way, a seeding of cranial roots through the CSF did not occur as the analysis of the CSF showed absence of neoplastic cells. Moreover the patient presented sensory disturbances due possibly to impairment of the ascending reticular formation in the mesencephalon.

As in the literature, the clinical history is of less duration (less than 1 year) and it reached in our case 3 months from the initial symptoms up to admission. The subsequent follow-up reached a

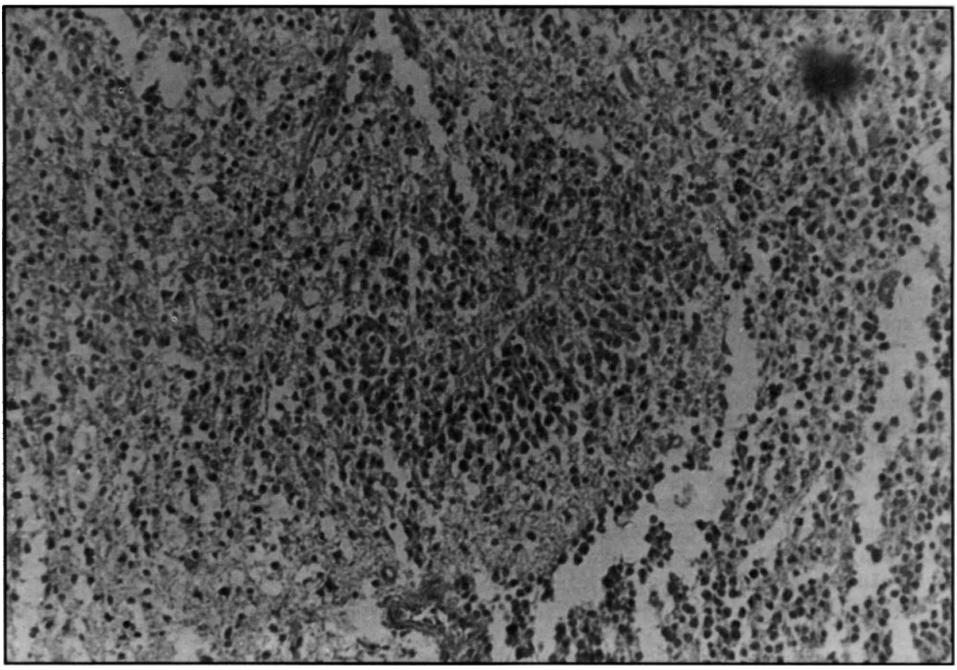

Figure 1. Glioblastoma multiforme: areas of necrosis, palissades and cellular pleomorphism. HE, 100 X.

fulminant course of around 2 months due to the in life undefined diagnosis and consequent absence of suitable palliative treatment (surgery, radiotherapy, chemotherapy)[1,4,6,10,13,22] excepting for the ventriculo-peritoneal shunt.

This kind of tumor has a predilection for the earlier decades of life as in our 26 years old patient. In the literature the mean age ranges from 18.1 to 28.5 years[7,21].

As we can see in Table 1, the evolution picture of the case evolved from initial signs of increased intracranial pressure, signs of cranial nerves impairment and of spinal cord lesions (incomplete Brown-Séquard syndrome). These three components progressed along around 2 months to a worse condition resulting in exitus letalis. However the main components of this picture concerned the cerebrum and brain stem and only in the final days a complete motor picture with tetraplegia and signs of bulbar involvement predominated.

The syndrome of increased intracranial pressure in this case was of paramount importance since it marked the whole process and was resistant to all procedures that attempted to lower it.

The occurrence of increased intracranial pressure as a consequence of spinal tumor is very rare[3,8,9,14] mainly as an initial symptom[18]. Up to 1990, only 34 cases of hydrocephalus and spinal cord tumor have been reported[19]. The hypotheses to explain this condition consider that there is a hyperviscosity of the CSF that leads to its increase, what in turn causes a slowing down of CSF circulation from the cranial to the spinal spaces and therefore an increased CSF pressure[2,12,23]. The decrease of the CSF circulation may lead to deposition of colloidal macromolecular substances producing a basal arachnoiditis[3].

These physiopathogical explanations fit to the evolutive aspects of our case: the deposition of substances that produce arachnoiditis may perhaps explain the impairment of cranial nerves, besides parenchymal brain stem lesions from tumoral origin. The hyperproteinorachia (38 mg/100ml, even from ventricular fluid) found in our case corroborates the physiopathological hypotheses above mentioned.

The association of increased intracranial pressure and malignant spinal cord tumor is bound to a worse prognosis and rapid progression of the picture[19], as in the case we describe, with an evolution of 57 days.

Most of the cases described have clinically signs and symptoms of spinal cord lesions whereas in our case brain stem signs were always present, which misled the diagnostic reasoning. Unfortunately we did not perform basic and important exams, not available in our hospital, as myelography, spinal CT and magnetic resonance. The several CT scans performed did not disclose, for their limitation in posterior fossa imaging, suggestive signs of this kind of lesion. Other interesting aspects of the clinical evolution are the initial presenting symptoms of intracranial hypertension as explained by the hydrocephalia the patient presented and less defined initial signs of myelopathy as expected from the site of the lesions and as described in the literature[2].

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