patología
Revista latinoamericana

Patología 2008;46(2):112-6

Case report

Frontal solitary plasmacytoma and meningioma (collision tumor). Case report and literature review

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RESUMEN

Se comunica un caso raro de mengioma benigno y plasmocitoma solitario en colisión. Se trata de un hombre de 46 años de edad que manifestó antecedentes de dolor facial y exostosis, alteraciones progresivas en la marcha y, un año antes, debilidad corporal. Inicialmente se le diagnóstico síndrome paraneoplásico y recibió tratamiento como tal. Al examen físico mostró debilidad, hiperreflexia simétrica, signo de Babinski positivo bilateral y exostosis frontal que afectaba también los tejidos blandos. La imagen de resonancia magnética cerebral mostró una masa hiperintensa frontal izquierda con crecimiento periférico, que afectaba los tejidos blandos, los huesos y las meninges. Desde el punto de vista histológico el tumor en la duramadre y en las meninges consistía en un meningioma sincicial típico y un plasmacitoma, ambos localizados en la misma área. Un plasmocitoma frontal es una manifestación rara de plasmocitoma extramedular y no se ha reportado previamente concomintante con meningioma. El presente artículo discute el diagnóstico diferencial entre meningioma inflamatorio, granuloma de células plasmáticas y una posible colisión de meningioma con plasmocitoma. Ambos componentes tienen la capacidad de recurrencia y transformación anaplásica.
Palabras clave: plasmocitoma solitario craneal, meningioma, tumor en colisión.

ABSTRACT

A unusual case of benign convexity meningioma and solitary plasmacytoma in collision is presented. A fourty-six year-old male presented with a history of face pain and exostosis, progressive ambulation disturbances and, one year before, body weakness. He was initially treated as paraneoplastic syndrome. On physical examination, he showed weakness, symmetrical hyperreflexia, bilateral positive Babinski sign and frontal exostosis affecting also soft tissues. The brain MRI showed a left frontal hyperintense mass with peripheral enhancement, affecting soft tissues, bone and meninges. Histologically the dura mater and meningeal tumor consisted of a typical syncytial meningioma, and a plasmacytoma, both localized in the same area. A frontal plasmacytoma is a rare presentation of extramedullary plasmacytoma and has not been previously reported in association with meningioma. The differential diagnosis between inflammatory meningioma, plasma cell granuloma and a possible collision of a meningioma with a plasmacytoma is discussed. Potentially, both components have the capacity for recurrence and anaplastic transformation.
Ked words: cranial solitary plasmacytoma, meningioma, collision tumor.

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Received: October, 2007. Accepted: January, 2008.

Also available at: **www.revistasmedicasmexicanas.com.mx**

Meningioma is a common and generally benign slow-growing tumor. Radiation exposure, trauma, type 2 neurofibromatosis, hormonal, genetic and molecular factors have been implicated in its development and growth.[1]

A plasmacytoma is defined as any discrete, usually solitary mass of neoplastic plasma cells, either in the bone marrow or in various soft tissue sites.[2]

The coexistence of two or more primary tumors is a relatively rare occurrence; it is termed collision tumor, and

is considered the coincidence of these two different tumors at the same time and location.[3] In most cases, these two different histological tumor types arise in separate locations, or in proximity, and they are focally intermingled.[1] The coexistence of two or more primary brain tumors is a relatively rare occurrence. There have been similar case reports documenting the coexistence of meningioma and glial tumors; mostly astrocytomas, glioblastomas, shwannomas, ganglioneurocytomas, pituitary adenomas, lymphomas.[1,3-10] Multiple primary brain tumors of different histology can also be found in the setting of phacomatosis or cranial radiotherapy.[1]

The simultaneous occurrence of brain tumors of different cell origins is unusual; primary brain tumors may be multifocal and metastatic tumors are frequently multiple in the central nervous system and they may coexist.[1] It has been hypothesized by some authors that one tumor may produce an oncogenic factor or other factors that may induce the growth of another lesion or may somehow favors the development of a second tumor of different origin.[1,11]

We report a case of brain collision tumor; syncytial meningioma localized in the same area of a plasmatocytoma, both tumors within the left frontal bone. The immunohistochemistry of plasma cell population with monoclonal immunophenotype is emphasized.

CASE REPORT

A fourty-six year-old male with a medical history of head trauma at the age of 8 years. He had no data of radiation therapy. Antecedents included face pain and two months of dizziness, focal motor seizures, progressive immobility and weight loss of 22 kg. He was admitted due to increased leg weakness and gait disturbances and the initial diagnosis was of a paraneoplastic syndrome.

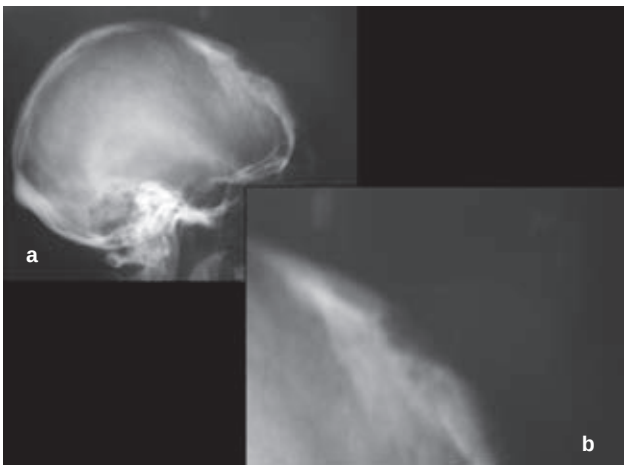

Figure 1. a. Simple cranial X-ray showing a left frontal lytic lesion. **b**. Close-up to the lytic lesion.

On physical examination he showed weakness, symmetrical hyperreflexia and bilateral positive Babinski's sign; cranial nerves II, IV and VI were normal and there were no meningeal signs. There was fontal exostosis, non-painful, bland, subcutaneous mass in the frontal region, without skin changes. Neck exam was normal.

Nerve conduction studies reported severe motor and sensitive axonal, demyelinating neuropathy. Cerebrospinal fluid glucose was 73 mg/dL, proteins 154 mg/dL, total cells 4.

Skull radiographs showed a lytic lesion (figure 1). Computed tomography scan showed a large hyperdense lesion; it showed gray matter involment and it was nearly isodense on T1 and T2. The brain MRI imaging scan showed a peripherically enhancing extra-axial, left frontal hyperintense mass, affecting soft tissues, bone and meninges (figures 2a and 2b).

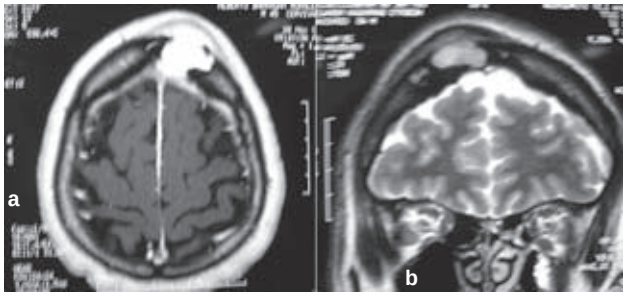

Figure 2. a. The brain MRI-image showed a left frontal hyperintense mass with peripheral enhancement, affecting soft tissues, bone and meninges. **b**. IRM coronal plane Gadoliniun-enhanced imaging showing heterogeneously enhancing in meningeal and subcutaneous lesion that spares the encephalon.

The patient underwent left craniostomy, the dura mater was thickened and infiltrated by reddish hypervascularized and granular tissue mimicking a meningioma. A gross total resection of the solid component of the dura mater and soft tissue tumor and a bone section (figures 3a and 3b) was performed. After the surgery a Tc99 bone scan and a bone marrow biopsy showed no tumor activity. Possibilities of lymphoreticular malignancy, myeloproliferative disorders and other inflammatory diseases were excluded by appropriate investigation.

Immediate follow-up was uneventful, post-operative treatment included radiotherapy (30 Gy in 10 sessions, lateral opposed field) and physical therapy. The patient was asymptomatic and his brain MRI did not show any features of intracranial process on following. Neuropsychological tests concluded that language, memory and emotional functions were normal, and he was able to carry on normal professional activity. Currently, after two years of his first admission, the patient's gait has improved; he is able to walk although there are still distal lower extremities weakness and paresthesias.

Gross aspect of surgical specimen corresponded a section of temporal bone; was irregular, 7 x 6.5 x 1.8 cm of diameter, the external surface showed red, granular, soft aspect (figure 3a), and internal surface showed a gray, and soft tumor, 2 cm of diameter (figure 3b).

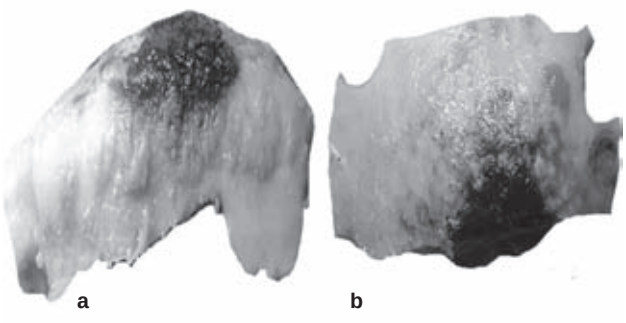

Figure 3. a. Gross aspect of the bone showing a red hemorrhagic tumor in one side and an ill-defined. **b**. Tumors showed fine, granular lesion in the inner surface of the bone.

The microscopic examination of the specimens confirmed that the dura mater and meningeal tumor consisted of a typical syncytial meningioma, formed by meningothelial cells arranged in a lobular architectural pattern; mild nuclear pleomorphism and rare mitoses were observed (figures 4a and 4b). No necrosis, hemorrhage, or psammoma bodies were found. The tumor was immunoreactive for EMA and vimentin, desmin and CD34 were negative. In other areas, the tumor showed a different histological pattern non consistent with meningioma; it was formed by mononuclear cells, mainly mature plasma cells and reactive lymphocytes. Occasional Russel's bodies were present. There was not evidence of vascular proliferation nor necrosis (figure 5a).

Immunohistochemistry revealed the presence of only kappa light chains in the plasma cells, demonstrating the

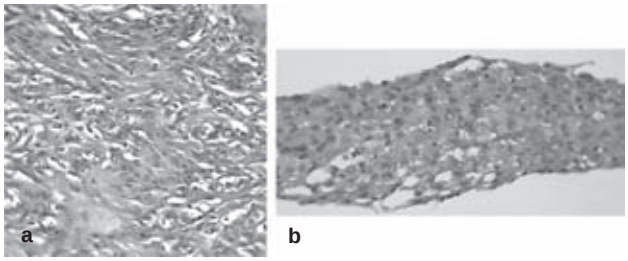

Figure 4. a. The meningioma histology showed long cells without atypia nor cell pleomporphism (H&E original magnification x 200). **b**. Close-up of the maelstrom appearance of the meningothelial cells (H&E original magnification x 400).

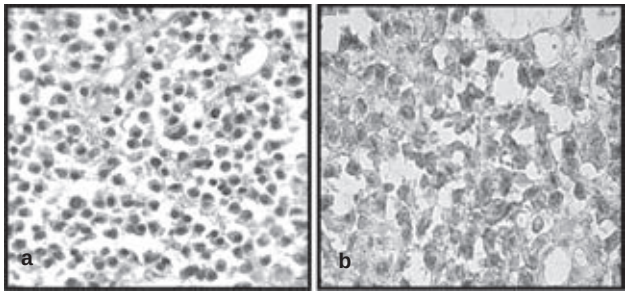

Figure 5. a. Tumor formed by plasma cells (H&E, original magnification x 200). **b**. plasma cells were immunoreactive for kappa light chains (IHC x 400).

monoclonal nature of these cells. Lambda light chains were negative (figure 5b). Diagnoses of meningioma and plasmacytoma were made.

DISCUSSION

Hematopoietic proliferations rich in plasma cells rarely occur within the central nervous system without the involvement of other organ systems, depending on their histological pattern, cellular composition, and immunophenotype. Several differential diagnoses include: solitary plasmacytoma, plasma cell granuloma, hyalinizing plasmacytic granulomatosis, hyperplasia of plasma cells, and inflammatory meningioma or lymphoplasmacyte rich (LPR) meningioma.[12-18]

Solitary intracranial plasmacytoma is exceedingly rare, benign, monoclonal plasma cell tumors that can arise from the skull,[19] occipital bone,[7] the dura or, rarely, from the brain.[14] Imaging studies bear some similarities with meningioma. A large extra-axial mass with an important lytic lesion should have led to the diagnosis of plasmacytoma.[14,20] The lesion is usually extra axial and nearly isointense with

gray matter on T2-weighted MR images, and diffusely enhanced after administration of contrast, sharing some characteristics with meningioma.[14,21] A diagnosis of solitary craniocerebral plasmacytoma should be considered when a mass with these imaging features is seen.[22,23]

Multiple myeloma may have extra osseous manifestations in the cranial region. It may be a solitary intracranial tumor without any other signs of multiple myeloma, or intracranial disease may be part of a generalized disease.[1]

Plasma cell granuloma (PCG) is a rare disease characterized by a non-neoplastic polyclonal proliferation of plasma cells. It can include lymphocytes, histiocytes and mainly, mature plasmocytes. It is of outmost importance to confirm the polyclonal nature of this disease. The distinction of PCG from the other disorders is sometimes very difficult to establish, due to lack of specific clinical and radiological features.[13,16]

Rosai-Dorfman's disease, Erdhein-Chester's disease, hematophagocytic lymphohistiocytosis or juvenile xantogranulomas are included in the differential diagnoses, where the polyclonal nature of immunoglobulins excludes the diagnosis of plasmacytoma and confirms the benign nature of the lesion.[13]

Many articles have discussed the lack of certainty on the pathogenesis of plasma cell disease, the strong inflammatory reaction argues for a post infectious origin, in association with viral diseases,[16] autoimmune disorders o may be due to post-trauma.[11,18,24]

A plausible interpretation of this sequence of events is that the inflammatory cell reaction to the meningioma caused the immunological response followed by an unusual hypergammaglobulinemia.[18,25] A peculiar type of meningioma with conspicuous plasma-cell components has been described.[13] According to the WHO classification of the central nervous system,[26] this rare clinical entity is recently designed as lymphoplasmacyte rich (LPR) meningioma.[17,27,28] This type of meningioma is usually accompanied by prominent peripheral blood abnormalities, anemia or polyclonal gammopathy, which disappear after surgical removal of the tumor. To date, the origin (neoplastic or inflammatory) of this tumor is unclear; its biological behavior and clinical course are anomalous, so it is considered closer to intracranial inflammatory masses rather than typical meningiomas.[13,25,27,29]

The collision tumor is considered as the coincidence of these two different tumors at the same time and the same location and other cases have been reported.[3,8] Synchronous multiple intracranial tumors specially occur in metastases. The association of meningioma and astrocytoma is the most common,[3,4,8] some rare cases of meningioma and lymphoma,[9] and others of meningioma and pituitary adenoma have been reported.[1] Banerjee et al.[2] published a subfrontal tumor with the features of plasmacytoma and meningioma, to our knowledge it is the first case published which presented this rare association between two different histological pattern.

The cause and pathogenesis of the association of meningioma and plasmacytoma is unclear, however, as meningiomas are common and slow-growing intracranial neoplasms,[1] the possibility of their concurrence with a second brain tumor might increase with time. In contrast, an irritative effect of meningioma facilitating the development of plasmacytoma cannot be excluded in cases in which the two lesions occurred in succession in the same area. On the other hand, a spectrum of solitary plasmacytic lesions may occur within the central nervous system and atypical plasma cell hyperplasia have the potential to evolve into plasmacytomas.[16,18] The collision tumor might have been caused by malignant transformation of a reactive meningoendothelial cell surrounding the plasmacytoma.[16] Tandem or coincidental tumors are synchronous tumors of different histogenesis in contiguous or far areas.[2] A locally acting oncogenic factor or irritative effects of a tumor inducing the growth of another neoplasm have been suggested as possible explanations.[6,11]

Although the molecular mechanisms responsible for osteolysis remain to be fully elucidated, it is clear from numerous studies that it is due, in part, to an increase in osteoclastic bone resorption. Several known osteoclast-activating factors are produced by myeloma cells (plasma cells) or by stromal cells in response to myeloma cells (plasma cell) and include interleukin-1beta (IL-1beta); tumor necrosis factor-alpha (TNF-alpha); IL-6; parathyroid hormone-related protein; macrophage inflammatory protein-1alpha and, the most recently described TNF-ligand family member, receptor activator of nuclear factor-kappa B ligand.[30]

Several etiologic factors have been proposed in the development of meningiomas, including: trauma, radiation, oncogenic viruses, chromosomal abnormalities, and hormonal factors.[1,11] Therefore, it remains unclear which one of the two tumors appeared first, if there is casual or causal coexistence or if it is only coincidental event.

CONCLUSION

We report the case of a 46 year-old male patient who developed exostosis, progressively worsening difficulty in ambulation and body weakness, after one year. He was initially considered as a paraneoplastic syndrome. On physical examination, he showed weakness, frontal exostosis and a subcutaneous mass. The cranial X-ray and IRM imagines showed meningeal infiltration and a lytic mass in the frontal bone. Histologically there was a syncytial meningioma localized in the same area of a plasmacytoma, with monoclonal immunophenotype, and both tumors in the left frontal bone; we named it a collision tumor. This is the first known and reported case of this unusual association of tumors located in the same anatomic area, preceded by a cranial trauma, and with a clinical presentation of paraneoplastic syndrome.

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