Coincidental pituitary adenoma and planum sphenoidale meningioma mimicking a single tumor

Adenoma hipofisario y meningioma de planum coincidentes simulando un tumor único

The association of pituitary adenoma and meningioma has been well described, especially in patients previously irradiated for the pituitary tumor who develop intracranial meningiomas years after treatment.[1] However, the coincidence of both tumors in contact has been seldom reported.[2–4] We present a case of contiguous pituitary adenoma and planum sphenoidale meningioma mimicking a single tumor which could be removed through an endoscopic expanded endonasal approach (EEA).

A 61-year-old patient, without previous oncological history or radiotherapic treatment, was referred to our institution after six months of progressive visual loss and bitemporal hemianopia that was confirmed by campimetry and optical coherence tomography. Neurological examination and endocrinological testing were otherwise normal, with the following serum levels: prolactin (PRL) 8.5 ng/ml; growth hormone (GH) 0.12 ng/ml; insulin-like growth factor I (IGF-I) 122 ng/ml; adrenocorticotropic hormone (ACTH) 28 pg/ml; cortisol 26.2 μg/ml; thyroid-stimulating hormone (TSH) 2.14 μUI/ml; thyroxine (T4) 1.74 ng/dl; follicle-stimulating hormone (FSH) 4.7 mUI/ml; luteinizing hormone (LH) 2.9 mUI/ml and testosterone 5.37 ng/ml. Magnetic resonance imaging (MRI) showed a 2.0 cm × 2.2 cm × 2.2 cm homogeneously enhancing sellar lesion with suprasellar extension compressing the optic chiasm (Fig. 1A and B). The pituitary stalk was displaced posterosuperiorly, though no normal gland could be identified on the MRI. The suprasellar extension was broad based, reaching above the tuberculum sellae and planum sphenoidale. A non-functioning pituitary macroadenoma was suspected and the patient was offered surgery. An endoscopic transphenoidal approach with 0° and 30° rigid nasal endoscope was performed. The rostrum sellae was completely removed up to the margins of both carotid arteries. Cranially only a limited transtuberc-ular and transplanum exposure was deemed necessary since the soft consistency of most pituitary adenomas allows a complete dissection and removal of the whole tumor after internal debulking if the suprasellar component is not exceedingly big. After successful resection of the sellar tumor, leaving glandular tissue attached to the pituitary stalk posteriorly, the diaphragm was opened and the suprasellar

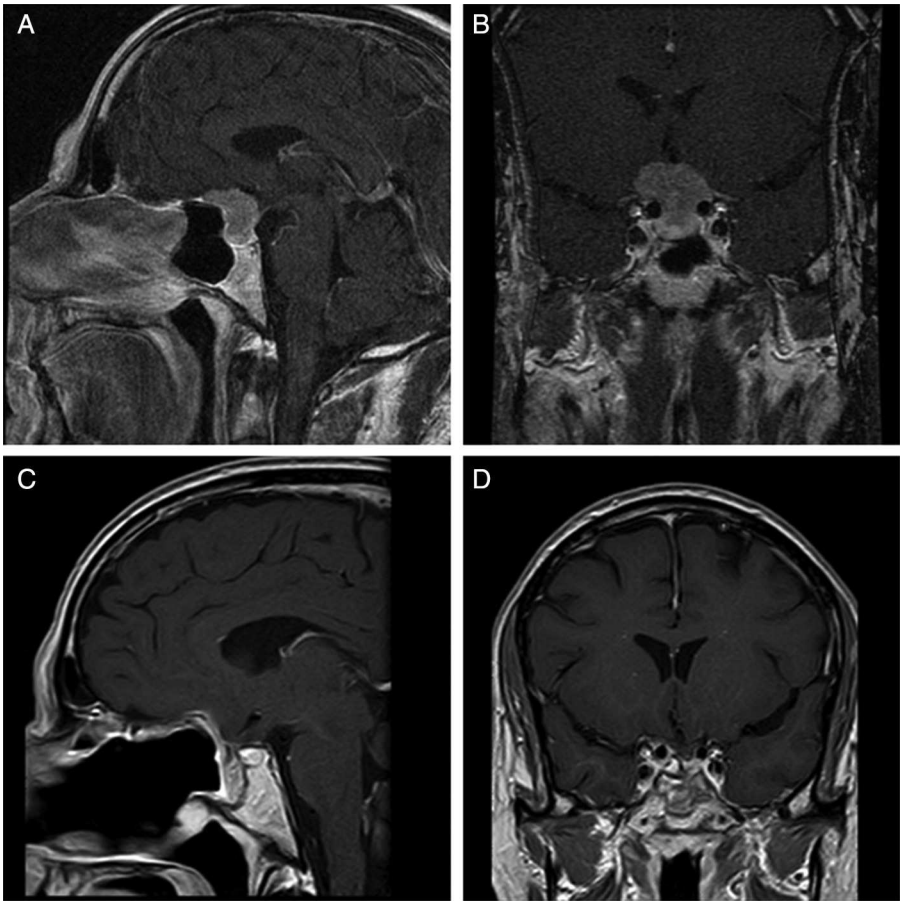

Figure 1 Preoperative magnetic resonance showing an apparently single sellar lesion with suprasellar extension to the tuberculum sellae and planum sphenoidale: (A) sagittal gadolinium enhanced T1-weighted image and (B) coronal gadolinium-enhanced T1-weighted image. Postoperative study with the same sagittal (C) and coronal (D) sequences shows a complete resection and sealing of the skull base with a nasoseptal flap.

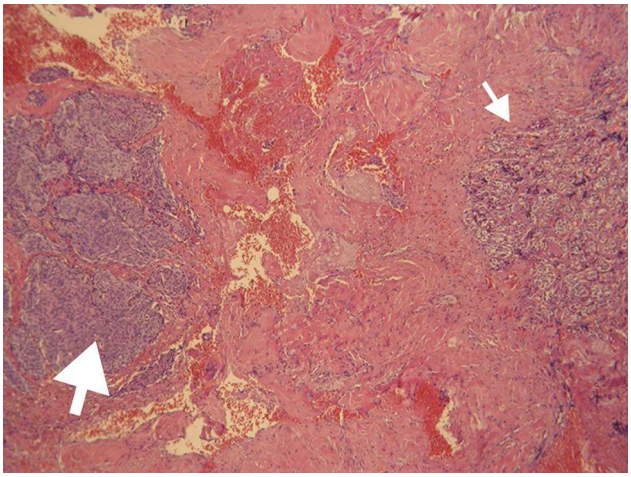

Figure 2 Photomicrograph of a section of the lesion that shows two different neoplastic tissues in contact (hematoxylin and eosin, 100×): clusters of densely packed cells of a meningothelial meningioma (big arrow) and a distorted acinar pattern of chromophobe cells corresponding to a pituitary adenoma (small arrow).

component was dissected from the optic chiasm and removed. Intraoperatively the suprasellar component appeared to have a stiffer consistency than the sellar tumor. After surgery the patient showed slight visual improvement. He developed hypopituitarism and persistent diabetes insipidus that were managed medically. The postoperative computed tomography (CT) suggested a tumor remnant at the planum sphenoidale. Meanwhile, histological study demonstrated the coincidence of two contiguous tumors: a pituitary adenoma with positive immunohistochemical staining for GH and LH, and a meningothelial meningioma (Fig. 2). Moreover the patient developed an intermittent CSF rhinorrhea when standing up. Therefore a reoperation was planned to remove the residual tumor and seal the CSF fistula. The patient was re-operated on day 8 through an expanded endoscopic transplanum approach that allowed the excision of the meningioma with decompression of both optic nerves. Effective sealing of the skull base was achieved through a multilayer closure with intradural and epidural fascia lata and a pedicled nasoseptal flap (Fig. 1C and D). The patient experienced a significant and stable visual recovery that allowed him to resume a normal life and return to work.

Radiotherapy is a well known cause of development of meningioma; several cases have been reported after irradiation of pituitary adenomas. On the other hand, coincidental meningiomas in patients diagnosed of pituitary adenoma and without previous irradiation are uncommon, with only 33 cases reported in the world literature.[3] The reported cases of coincidental pituitary adenoma and meningioma show a clear preference of the meningioma for a perisellar location at the planum sphenoidale, tuberculum sellae and the sphenoid wing.[1,3,5] Contiguity of the meningioma with the pituitary adenoma mimicking a single mass is extremely rare.[2,4] Coincidental menigiomas have been reported in patients with non-secreting adenoma, prolactinoma, acromegaly and Cushing disease. Although prolactinomas are the most frequent secreting adenomas, the higher prevalence of acromegaly among patients with coexisting meningioma has led some authors to propose an association between GH-secreting adenomas and the development of meningioma.[5,6] However, although meningiomas may express GH and IGF-I receptors and may show growth response to its stimulation in vitro, this effect has yet to be proved in vivo.[7]

The diagnosis of a meningioma in a close location to a pituitary adenoma poses a therapeutic challenge. In most reported cases the coexisting tumors were managed independently, usually addressing the pituitary adenoma first with either medical therapy or a transphenoidal approach, and treating the menigioma separately with conservative measures or a specific surgical approach.[1] Some authors used a single pterional approach for both tumors.[2,4,8] Any of these strategies implies a significant increase of the risk of morbidity as compared to the surgical treatment of a single pituitary adenoma. In the present case a solitary pituitary adenoma with suprasellar extension was suspected on the preoperative MRI; therefore an endoscopic endonasal approach was performed. Because of the presence of residual tumor on the postoperative CT and the pathological diagnosis of a coexisting meningioma, a reoperation through an endoscopic transplanum EEA was decided. This approach offers a direct view of suprasellar meningiomas without frontal lobe retraction and allows resection with minimal bleeding and good control of the optic apparatus and the anterior cerebral arteries.[9,10] Only one previous case of contiguous pituitary adenoma and suprasellar meningioma successfully resected through an endoscopic EEA has been reported.[3] The present case adds to the evidence that in the rare situation of contiguous sellar and suprasellar tumors located in the same sagittal plane, an endoscopic EEA allows an adequate exposure and safe removal of both tumors.

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http://dx.doi.org/10.1016/j.endonu.2015.03.003

Emulsión lipídica exclusiva de ácidos grasos omega-3 (Omegaven®) en el tratamiento de la hepatopatía asociada a nutrición parenteral: a propósito de un caso

Exclusively based parenteral fish-oil emulsion (Omegaven®) in the treatment of parenteral nutrition associated liver disease: Report of a case

Las emulsiones lipídicas que se incorporan en la nutrición parenteral (NPT) permiten aportar los ácidos grasos esenciales y cubrir el requerimiento energético sin un aporte excesivo de hidratos de carbono. Asimismo, tanto la composición de estas emulsiones como la dosis aportada se relacionan con el desarrollo de la hepatopatía[1,2].

Recientemente se han publicado varios artículos relacionados con el efecto de emulsiones lipídicas intravenosas basadas exclusivamente en ácidos grasos omega-3 (AGW3), en el tratamiento de esta enfermedad en el paciente adulto[3–7].

Presentamos el caso de un paciente adulto, con el período de tratamiento más largo comunicado en España, con datos de mejoría analítica y funcional con este tipo de emulsión lipídica.

Mujer de 44 años, con NPT desde 2010, tras resección intestinal masiva por tumor desmoide, quedando un intestino residual de 12 cm de duodeno, medio colon transverso y colon izquierdo. Además del soporte nutricional administrado de forma cíclica, desde un inicio recibió tratamiento con ácido ursodeoxicólico y antibióticos para el sobrecrecimiento bacteriano, para evitar el desarrollo de hepatopatía. La longitud del intestino delgado remanente imposibilitó la administración de nutrición por vía enteral.

Durante los años 2010 y 2011 se administró una fórmula lipídica compuesta por triglicéridos de cadena media (TCM), aceite de soja y AGW3 (proporción 5:4:1), con dosis 1,1-1,4 g/kg/día, relación calórica hidratos de carbono:lípidos 60:40 y una solución nitrogenada con alta concentración de aminoácidos ramificados (16 g nitrógeno/día). Ante la elevación de enzimas hepáticas, en octubre de 2011 se sustituyen los oligoelementos por otro producto con menor aporte de manganeso, con mejoría de la función hepática a finales del mismo año.

Tras valoración en centro de referencia de trasplante intestinal se realiza biopsia hepática en abril de 2012, con datos de parénquima hepático con colestasis parenquimatosa y canalicular, compatible con alteraciones post-NPT. Con bilirrubina previa de 5,1 mg/dl alcanza cifras de 15,4 mg/dl tras la biopsia hepática (complicada por hematoma y hemoperitoneo). Ante este empeoramiento brusco de la función hepática se suspende la administración de lípidos durante 3 semanas, reiniciándose en junio con aporte de otra emulsión lipídica (mezcla física de aceites de soja, TCM, oliva y AGW3 en proporción 3:3:2,5:1,5) 25 g/día, con aumento progresivo en los siguientes meses llegando a mantener pauta de administración de 50 g/48 h, con mejoría parcial de las cifras de bilirrubina al finalizar el año. Al disminuir el aporte de lípidos precisó suplementación con vitaminas K y A.

Durante el año 2013, ante mejoría de la función hepática y la pérdida de peso progresiva (peso mínimo alcanzado en abril, 44,5 kg; IMC 16,6 kg/m^2) se aumentó el aporte de lípidos llegando a administrar 65 g/día 4 días por semana y 40 g/día los días restantes (fig. 1).

Durante el seguimiento se realizaron ecografías periódicas con hallazgo de hígado de tamaño, morfología y ecogenicidad normal, esplenomegalia y barro biliar. En la realizada en octubre de 2013 se constatan datos de hipertensión portal (sin datos de varices esofágicas en gastroscopia) con porta discretamente aumentada de calibre que presenta un flujo hepatófugo. En los Fibroscan® realizados en 2012 y 2013 no se detectaron datos de fibrosis hepática (dureza 4,8 KPA, equivale a niveles de fibrosis 0-1 en biopsia).

En marzo de 2014 presenta de nuevo empeoramiento progresivo de la función hepática (bilirrubina total máxima 11,4 mg/dl) y descartadas otras causas de hepatopatía, rechazada la opción de trasplante intestinal por parte de la paciente y revisada la literatura[3–6] se le ofrece sustituir la fórmula lipídica por otra exclusiva en AGW3 (45 g/día, 0,9 g/kg, hidratos de carbono: lípidos 66:33; calorías 1.605). Este tipo de emulsión lipídica está aprobada, aunque no comercializada en España, para el uso simultáneo con otra emulsión lipídica y a una dosis máxima de 1-2 ml/kg peso. Se solicitó autorización a la Agencia Española del Medicamento y a la Dirección del Hospital para el uso en condiciones fuera de ficha técnica, tal y como se había hecho en las publicaciones revisadas (emulsión lipídica exclusiva), y la paciente firmó el consentimiento.