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Primary intraosseous meningioma. Case report and literature review

Meningeoma intraoseo primario. Reporte de caso y revisión de literatura

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Abstract

Meningiomas that develop in ectopic locations represent only 1 to 2% of the total number of Meningiomas. The term Primary Extradural Meningioma (PEM) signifies that the origin of these tumors is isolated from the dural covering of any part of the brain and spinal cord. Primary Intraosseous Meningioma (PIM) describes a PEM subtype that originates in the bone and accounts for approximately two-thirds of all the Extradural Meningiomas. The clinical appearance of lesions and differential diagnoses depend primarily on the lesion's location, size and neighboring structures. The radiological appearance of Intraosseous Meningiomas depends to a large extent on their location and on the effects they cause upon invading the bone. Tumors are typically osteoblastic or osteolytic. Intraosseous Meningiomas are Histologically and Immunophenotypically indistinguishable from their Intracranial Meningiomas counterparts. When surgical excision is the treatment of choice, the patient's potential cure is possible.

Key words: Primary extradural meningioma, intraosseous meningioma, meningothelial meningioma, surgical resection.

Resumen

Los Meningiomas que se desarrollan en localizaciones ectópicas representan solo del 1 al 2% del total de los Meningiomas. El término Meningioma Extradural Primario (MEP) refleja que el origen de estos tumores es aislado de la cobertura dural de cualquier parte del cerebro y la medula espinal. Meningioma Intraóseo Primario (MIoP) describe un subtipo de MEP que surge en el hueso y representa aproximadamente dos terceras partes de la totalidad de todos los Meningiomas Extradurales. La presentación clínica de las lesiones y los diagnósticos diferenciales dependen primariamente de la localización de la lesión, tamaño y estructuras vecinas. La apariencia radiologica de los Meningiomas intraóseos depende en gran medida de su localización y los efectos que producen al invadir el hueso. Los tumores son típicamente osteoblasticos u osteoliticos. Los Meningiomas intraóseos son indistinguibles Histologicamente e Inmunofenotipicamente de su contraparte los Meningiomas Intracraneales. Cuando la escisión quirúrgica es el tratamiento de elección la potencial curación del paciente es posible.

Palabras clave: Meningioma extradural primario, meningioma intraóseo, osteoblástico, osteolítico, meningioma meningotelial, resección quirúrgica.

Introduction

Meningiomas are the most common tumors of the Central Nervous System (CNS)[1]. They account for about 35% of all primary intracranial and spinal neoplasms in adults[2]. Meningiomas originate from the Meningothelial Cells or the arachnoid cell layer, therefore tumors appear where such cells are abundant; arachnoid granulations and arachnoid villi concentrated along

the venous sinuses and their tributaries. Frequent locations of intracranial or juxtacranial meningiomas include: Convexity (20-34%), parasagitals and cerebral falx (18-22%), Sphenoid bone and middle fossa (17-25%), frontobasal (10%), posterior fossa (9-15%). The incidence of spinal meningiomas is 1/9 of intracranial meningiomas[3].

Meningiomas that develop in ectopic locations represent only 1 to 2% of the total number of Meningiomas[1-12]. The term Primary Extradural Meningioma (PEM) signifies that the origin of these tumors is isolated from the dural covering of any part of the brain and spinal cord, and in turn we are able to distinguish them from Primary Intracranial Meningiomas (MIPs), which may have extradural extension and/or metastasis[4]. Primary Intraosseous Meningioma (PIM) describes a PEM subtype that appears in the bone and accounts for approximately two-thirds of all of the Extradural Meningiomas[5].

This article presents the clinical case of a 50-year-old female patient, who underwent a surgical operation of a right frontotemporal sphenoidal bone lesion, where the Histopathologic result reported a Meningothelial Meningioma. A review of the current literature on the genesis, diagnosis and treatment of this type of lesions is carried out.

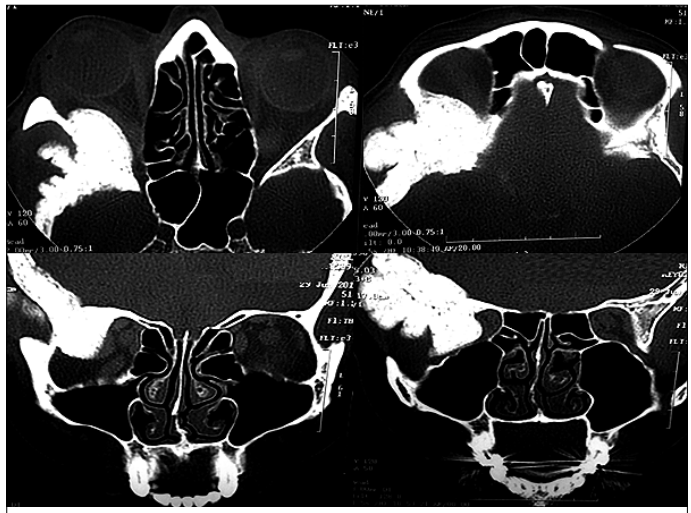

Figure 1. Bone excrescence and asymmetry due to the larger size of the orbital roof and the sphenoid's greater wing on the right-side. Extraconal mass effect.

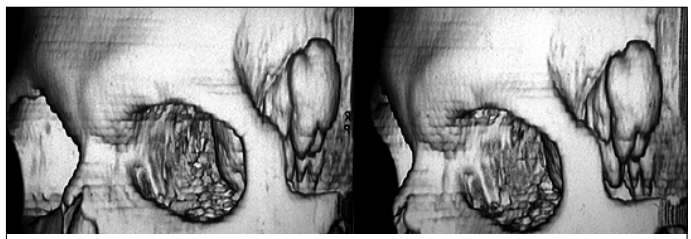

Figure 2. 3D reconstruction of multiplanar CT showing lesion projecting towards the ipsilateral temporal fossa.

Clinical case

Female, 50 years old, with no relevant history related to her current condition. Approximately 2 years ago she started with a clinical picture characterized by right temporal Cephalalgia, with a pain intensity of 8/10 according to the Visual Analogue Scale (VAS), with pulsatile characteristics, irradiation to the occipital region, with exacerbations to changes in position and a decrease of pain while resting and ingesting NSAIDs.

Presence of right temporal exostosis and ocular ipsilateral proptosis accompanied by epiphora and ocular pain.

With the guided physical exploration we found; Bilateral visual acuity of 20/30 in Snellen type arithmetic optotype, with no campimetric alterations due to confrontation, 3 mm isocoric pupils reactive to light stimulus. Limited right ocular abduction. Normal bilateral corneal reflex. Frontotemporal sphenoidal exostosis of approximately 2.5 cm in diameter with irregular borders, of stony consistency, non-mobile, non-painful and ocular proptosis.

Tomography study of skull in simple phase and with contrast, multiplanar, cuts with intervals of 3 mm which evidenced; bone excretion and asymmetrical due to the larger size of the orbital roof and the sphenoid greater wing on the right-side. Increased sclerosis was identified as well as bone trabeculation at the described locations. Such mass was projected towards the ipsilateral temporal fossa. It produced extraconal mass and directly on the lateral rectus muscle on the same side. It partially obliterated the right orbital apex (Figures 1 and 2).

The patient was subjected to a surgical intervention after being placed in a supine position, with placement of a 3-point cranial fixation system (Spetzler), where the ipsilateral pin was placed in the mastoids and the contralateral pins in the superior temporal line. Cephalic positioning was performed with Traction in a longitudinal direction, Elevation above the right atrium, lateralization, deflection and a 30º rotation. A hemicoronal incision was made and the cutaneous flap was retracted anteriorly, which is fixed to the surgical field. Interphascial dissection, subperiosteal section and dissection of the temporalis muscle was performed. The latter was pulled and fixed in the direction of the skin flap. Bone excretion (see above) was identified with smooth edges and irregular shape of approximately 2.5 cm in diameter, with emergence in pterion and temporal fossa (Figure 3). Frontotemporoparietal craniotomy was performed with a superior frontal keyhole and a lower limit of one centimeter of the lesion. Temporosphenoidal craniectomy was begun with resection of the lesion and milling of the sphenoid's greater wing, roof and lateral wall of the orbit, with a resection of approximately 85% (Figure 5). On the surgical bed, it was observed that there was no dural communication with the bone lesion (Figure 4). Bone flap plasty was per-

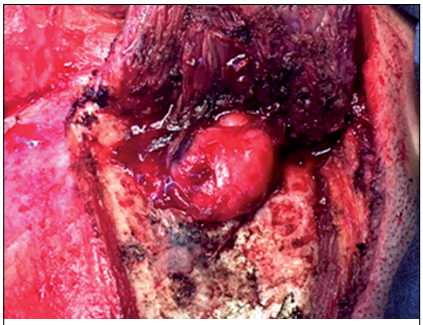

Figure 3. Intraoperative image of bone excrecence with smooth edges and irregular shape of approximately 2.5 cm in diameter, with emergence in pterion and temporal fossa. Anterior traction of the temporal muscle

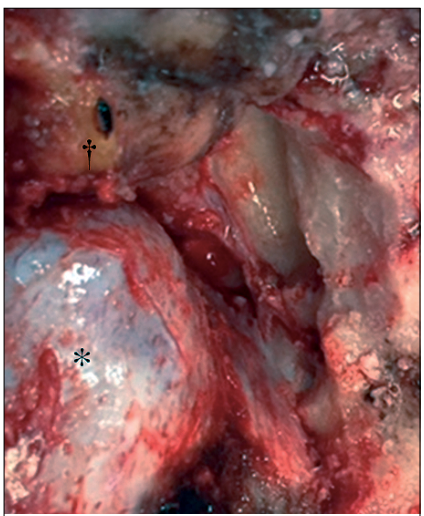

Figura 4. Intraoperative image of the dura mater in the frontopolar* and periorbital† region, after bone removal, where no dural involvement is observed.

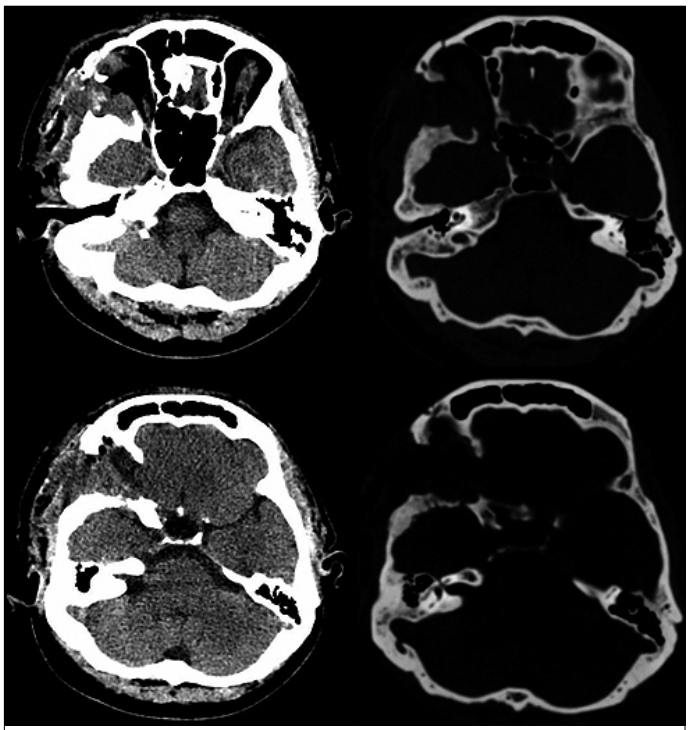

Figura 5. Simple multiplanar CT and post-surgical bone window, where craniectomy locations are observed in the frontotemporal sphenoidal region after an almost full resection of the lesion.

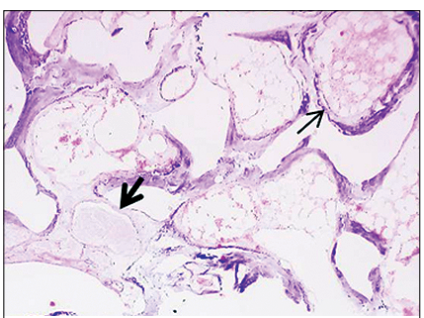

Figure 6. The meningioma lobes are in contact with bone cells of variable cellularity. (neoplastic ← cells, medullary → cells).

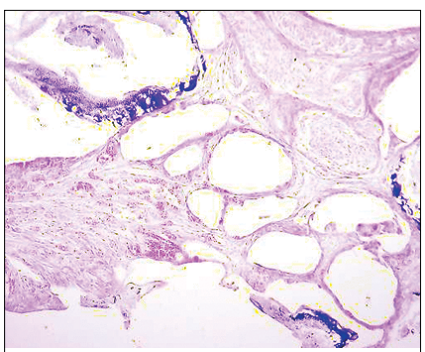

Figura 7. Neoplastic meningothelial lesion with nodular pattern is observed, delimited by fibrous septa with some cystic areas, eroding bone and areas with recent hemorrhage.

formed and the tissues were closed by planes in a conventional manner. This concluded the surgical procedure.
A surgical specimen was sent to the Pathology service, which reported a neoplastic meningothelial lesion with nodular pattern, delimited by fibrous septa with some cystic areas, which erode the bone and areas of recent hemorrhage (Figures 6, 7, 8 and 9).

Discussion

Intraosseous meningiomas are histologically and immunophenotypically indistinguishable from their intracranial meningiomas counterparts[6].
Pathogenesis of Primary Intraosseous Meningiomas is controversial[7]. There are different theories that try to explain the origin of this subtype of tumors:[5,8] it has been suggested that their formation is due to the entrapment of arachnoid cells in the sutures during the development of the skull in the pre- and post-natal stages[7], Lang et al. found that only 8% of intraosseous meningiomas were associated with a cranial suture. Clusters of arachnoid cells that may remain attached to the surface of blood vessels or nerves in their course or exit through foramen in the skull[6]. The presence of undifferentiated ectopic Mesenchymal Cells and etiology of fractures[9]. However, only about 0.2% - 4% of patients with Primary Extraditional Meningiomas have a history of Trauma to the skull[4,10].
Lang et al. found in their systematic review that of 142 patients and 168 tumors studied in the latter, 15 patients had intraosseous meningiomas that involved the orbit and the sphenoid bone (10.5%), as in the case of our patient.
The clinical appearance of the lesions

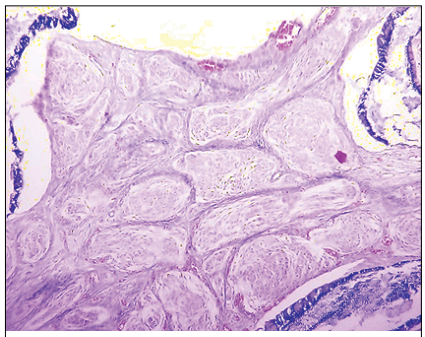

Figure 8. Neoplastic meningothelial lesion with nodular pattern is observed, delimited by fibrous septa with some cystic areas, eroding bone and areas with recent hemorrhage.

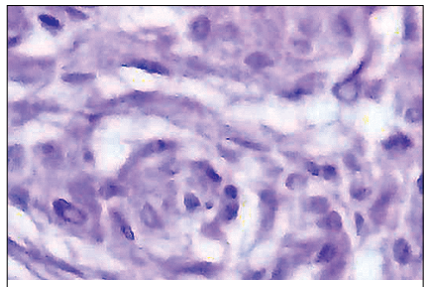

Figure 9. Meningothelial cells have nuclei with membrane reinforcement, basophils, some hyperchromatic and intranuclear pseudoinclusions.

and the differential diagnoses depend primarily on the lesion's location, size and neighboring structures[4,5,8]. Intraosseous meningiomas of the skullcap bones usually show slow growth as easily palpable subcutaneous masses. They are usually firm and non-painful, with no associated skin alterations at the growth location, and they may be incidentally detected. Neurological signs and symptoms are usually absent. In contrast to Intraosseous Meningiomas of the skull base that may occur with cranial nerve deficits, such as ophthalmoplegia, campimetric alterations, or alterations associated with the mass effect that provoke as ocular proptosis[5]. In comparison with Primary Intradural Meningiomas where the male-female ratio is 1:2, for Primary Extraditional Meningiomas it is 1:0.73-1.4. The latter had two distribution peaks in the patients' age. The first incidence peak occurs during the second decade of life and the second incidence peak appears between 50 and 70 years of age[10].

Nearly half of the patients present imaging studies interpreted as "normal"[6]. Radiological appearance of Intraosseous Meningiomas depends to a great extent on their location and the effects they produce when invading the bone. Tumors are typically osteoblastic or osteolytic, and sometimes a combination of both has been reported[11]. Meningioma cells are known to invade the Haversian canals to stimulate osteoblastic activity and they are usually manifested with hyperostosis[9]. The latter represent the most common radiological subtype of Intraosseous Meningiomas[11], whereby conventional radiographs show hyperdensities associated with the lesion. However, overlapping bone structures limit the use of this radiological modality. Skull x-rays can detect abnormalities in 30-60% of Intraosseous Meningiomas cases, and x-ray damages include hyperostosis, bone erosion, calcifications, and marks of atypical vascular elements[5,11]. On the other hand, Tomographic studies with bone windows show hyperdense, intradiploic lesions with expansion or destruction of the skullcap bone layers. The tumor is usually in the range of 65-85 Hounsfield Units, and it is evenly enhanced after the administration of a Contrast Agent[5]. In addition, Tomographic studies allow us to perform three-dimensional reconstructions of the bone and vascular structures, which becomes a useful tool for surgical planning. Magnetic resonance imaging shows meningiomas as hiso or hypointesal images compared to cerebral parenchyma, in T1-weighted images, whereas T2-weighted images show variable signals of Intensity. Angioresonance is reserved for those intracranial lesions, however it is typically useful in cases of Intraosseous Meningiomas that require early embolization[8], or to visualize changes in the vascular pattern by mass effect. When administering Gadolinium, the tumors do not exhibit the typical image of the dural tail of Intradural Meningiomas, however it is possible to visualize a reinforcement of the underlying dura mater, where such reinforcement may be secondary to dural irritation or tumor invasion[5]. It is of paramount importance to detect any possible intracranial extension and invasion to adjacent structures before surgical resection, in order to prevent potential intraoperative complications.

These tumors may be erroneously classified as primary bone lesions, where the main differential radiological diagnoses include: Osteosarcoma, Bone Metastasis, Fibrous Dysplasia, Paget's Disease, Bone Fibroma and Osteomas.[8] Other lesions that may appear as hyperdense focal bone lesions are eosinophilic granulomas, aneurysmal bone cysts, and plaque meningiomas[5].

Diagnosis of Intraosseous Meningioma represents a challenge and even histologically errors may be made by differentiating primary and secondary osteosclerotic tumors[7]. Microscopic study of lesions may reveal findings that are pathognomonic for intradural meningiomas including: psammoma bodies and eosinophilic neoplastic cells with irregular borders and nuclear pseudoinclusions grouped in spirals[5]. Due to a high morphological variability, the immuhistochemical study it is necessary to confirm the diagnosis. Most meningiomas are immunoreactive with membrane epithelial antigens (which stains the cytoplasmic membrane) and vimentin (which stains the cytoplasm), and variably other stains such as S-100, and cytokeratins[6]. Lester et al. in their series of 36 cases of Primary Meningiomas of the temporal bone mostly, 33 cases were classified as Meningothelial Meningiomas with an infiltrative growth pattern. This corresponds to our reported case.

Classifications have recently been proposed to unify the terms and avoid confusions of the meningiomas that grow in locations other than in the subdural space.

Lang et al. propose a classification for Primary Extradural Meningiomas based on the relationship that these lesions contain with the skull.

Type I; includes lesions that are purely extracalvarial without contact with the bone.

Type II; purely calvarial tumors, located completely within the bones of the skull

Type III; corresponds to calvarial tumors with extracalvarial extension, a tumor located in the skull, but with extracranial extension to soft tissues.

Types II and III are further subdivided into B when they are located at the base of the Skull and C for the tumor's convexity.

The treatment is certainly Surgical for these types of lesions. When surgical excision is the treatment of choice, the patient's potential cure is possible. Ideally, bone reconstruction should be performed during the same surgical time.

Sometimes total resection of the tumor is not possible, especially with lesions in the base of the skull, in which case decompression of nerve structures should be performed[5,11].
In the case of patients with unresectable tumors that condition neurological deficits or who have histologically exhibited malignant or atypical data, adjuvant therapy should be considered. This therapy includes external radiation or Gamma Knife, chemotherapy and hormone therapy[5]. The recurrence of Primary Extradural Meningiomas has been mainly associated with the location and extent of resection. The estimated recurrence rate for atypical Meningiomas is close to 21.1%[4].

Conclusions

Intraosseous Meningiomas are rare tumors, whose clinical and radiological data are not specific and represent a real challenge for the treating physician, therefore they require a Histological evaluation to reach a definitive diagnosis.
Surgical resection represents the main therapeutic option, however, vascular and nerve structures adjacent to the lesion, degree of extension and benign or atypical radiological characteristics have to be evaluated.
The prognosis of the patients after being surgically operated is generally good.

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