Giant cavernous Hemangioma of the cavernous Sinus. 
Case Report and Review of Literature.

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Abstract

The incidence of cavernous sinuses cavernous hemangiomas is 2% of all cavernous malformations. These lesions are well circumscribed benign neoplasm, that are confined to the dural layer of the venous sinus. These lesions are difficult to accurately diagnose them presurgically and the surgical treatment is also difficult. Nevertheless with the new imagining diagnosis technologies and with advancement in microneurosurgical techniques, neurosurgeons can treat these lesions with more confidence. In this article we want to report a case of a 24 year woman who presented to our institution with galactorrea as the only clinical presentation. This patient was investigated and was found to have a giant lesion in the right middle fossa. She was taken to surgery with the suspicion of a giant meningioma but this surgery was aborted when we found that the lesion did not correspond to a meningioma. The patient was followed for 4 years and the galactorrea was treated and she was stable, but then she presented with severe headache and deterioration in her consciousness. She was treated in another institution where she was taken to surgery and a partial resection was performed, the surgery was aborted because the patient presented massive bleeding.

we want to present this case that is dealing with an infrequent lesion of a very delicate localization of the skull base. We also want to emphasize in having this differential diagnosis when dealing with lesions in this area. We also want to warn and recommend that if a neurosurgeon stumble into one of these lesions in surgery we recommend to back off and plan better the surgical management. Planning adequately the surgical procedure improve the prognosis in these patients We will present this case and review the literature concerning these rare lesions.

Key Words: Cavernous sinus Hemangioma, Cavernous Malformation, Cavernoma, Cavernous Sinus, Skull Base Surgery.

Resumen

La incidencia de los hemangiomas cavernosos del seno cavernoso es del 2% de todas las Malformaciones cavernosas. Estas lesiones son neoplasias benignas bien diferenciadas, que se encuentran confinadas en el capa dural de los senos venosos. Estas lesiones son difíciles de diagnosticar de manera segura en las etapas prequirúrgicas y su tratamiento es difícil. Sin embargo con las nuevas tecnologías de imágenes diagnosticas y los avances en la microneurocirugía, los neurocirujanos podemos tratar estas malformaciones de manera mas segura. En este artículo queremos reportar un caso de una mujer de 24 años quien consulto a nuestra institución por galactorrea. Esta paciente fue investigada y se le diagnosticó una lesión gigante de la fosa media derecha. Se llevo a cirugía bajo la sospecha que presentaba un meningioma gigante pero esta cirugía se abortó cuando se encontró que la lesión se trataba de una patología distinta. La paciente siguió en controles por 4 años donde se mantuvo estable con mejoría de la galactorrea con manejo medico, pero luego comenzó a presentar cefalea severa asociada a deterioro de la conciencia. La paciente fue tratada en otra institución donde fue llevada a cirugía en la cual una resección parcial se le realizó a la paciente. Esta cirugía se abortó por un sangrado excesivo. Queremos presentar este caso que trata de una lesión infrecuente en un sitio de alto riesgo quirúrgico de la base del cráneo. Enfatizamos en tener esta patología entre lo diagnósticos diferenciales de las lesiones de esta localización. Además, recordar que en caso que uno se encuentre con una de estas lesiones en cirugía, recomendamos que finalice la cirugía en ese momento y que planeee un manejo quirúrgico enfocado para esta patología en un nuevo tiempo quirúrgico. Un adecuado planeamiento del procedimiento quirúrgico mejora el pronóstico en estos enfermos. En este artículo queremos reportar un caso y revisar la literatura que concierne estas lesiones raras del sistema nervioso central.

Palabras Claves: Hemangioma del seno cavernoso, Malformación Cavernosa, Cavernoma, Seno Cavernoso, Base de Craneo

Introduction

Intracranial Cavernous Hemangiomas are rare lesions that conforms 5 to 16% of all intracranial vascular malformations. Extra-axial cavernous Malformations are even more rare and estimated to be 0.4 to 2% of all the intra-cranial cavernous hemangiomas. These lesions are well circumscribed benign neoplasms, which are confined to the dural layer of the venous sinus. The incidence of cavernous sinuses cavernous hemangiomas is 2% of all extra-axial cavernous malformations. Microscopically they are composed of fine vascular canals whose wall ate composed of endothelium and connective tissue and with blood circulating in these channels. There may be a relationship between the presence of a cavernous sinus cavernoma and the presence of a skin hemangioma nearby. These lesions are difficult to accurately diagnose them presurgically and the surgical treatment is also difficult. Nevertheless with the new imaging diagnosis technologies and with advancement in microneurosurgical techniques, Neurosurgeons can treat these lesions with more confidence.

Case Report

A 24 year woman consulted because of menstrual irregularities and galactorrhea. She was a prolactine level of 40 ng/ml, the rest of the endocrinological screen was normal. A CT scan was done where irregular heterogeneous lesion was found in the cranial base. A Cerebral MRI was performed and it showed a large lesion in the right parasellar region which extended to the suprasellar region and the medial cranial fossa. The lesion was hipo-intense in T1 and hyper-intense in T2. A cerebral angiogram was performed which did not show the presence of a vascular malformation or a giant aneurism. The carotid artery was displaced to the left and the middle cerebral artery was elevated. We suspected that the patient had a cavernous sinus meningioma and an orbitozigomatic approach was planed for its resection. A blue violet lesion was approached extraduraly and intraduraly. We did not find a cleavage plane. We decided to puncture the lesion which resulted in a considerable bleeding. We performed homeostasis, and did not get any lesion sample for pathological study. We decided to abort the surgery because the lesion we were treating was different of what we planned for. Studying the case we suspected that the patient had a Cavernous Malformation of the cavernous sinus. We proposed a surgical treatment for the patient and explained the risks. The patient and her family denied having a surgical treatment so we decided to treat her with ergotamine and observe the patient regularly. The patient was treated with So 3 years post surgical visit the patient’s menstrual cycle and galactorrhea had gotten better and imaging controls show that the lesion has not increased in size. On the fourth year of observation the patient presented a severe headache with deterioration in her consciousness being somnolent and asthenic. She also presented a fine and continuous tremor of her left hand. She was managed in another institution. The neurosurgeons approached the lesion thinking it was a meningioma. The patient bleeds almost 5 liters. A partial resection was accomplished. The patient presented with complete third nerve palsy and a mild hemiparesis in the post operative period. The histopathological study showed that the lesion was indeed and hemangioma. In a 6 month control the patient is still have a third nerve palsy and her motor deficit is improving and the control MRI shows a remnant of the lesion in the right cavernous sinus. The patient had denied a third intervention or any additional surgical treatment at this time.

Discussion

In the world literature there are many names for this same pathology: extra-cranial cavernous hemangioma, extra-cranial cavernous angioma, cavernous hemangioma of the cavernous sinus, hemangioma of the middle fossa, adenoma of the cavernous sinus. Intra-cranial cavernous angiomias are rare lesions and only represent 5 to 6% of the intracranial vascular malformations. Intracranial extra-axial cavernous hemangiomas are even rarer, 0.4 to 2% (30). These lesions are well defined benign neoplasm and are limited to the sinus dura. The incidence of hemangiomas of the cavernous sinus is 2% of the lesions in this anatomical structure. It is more frequent in females. Microscopically this lesion contains vascular spaces with endothelial alignment and variable sizes, from capillary to cavernous. They have an abundant vascular formation; however, these vessels do not contain a muscle layer, although they do have a small amount of elastic tissue. There is no neural tissue present, and the dura comprises the capsule of these lesions.

The symptomatology may be only a retro-orbitarian headache on the side of the lesion. It may exacerbate during exercising and this is attributed to an increased intracranial pressure; with endocrine disorder and delayed menstruation in the case of this female patient, due to displacement of the pituitary stalk, which caused galactorrhea. The most frequent symptoms are a retro-orbitarian headache. This may be accompanied by alteration of cranial nerves, facial hypothesia, trigeminal neuralgia, exophthalmus, and galactorrhea. They rarely present hemorrhage as a clinical manifestation, in contrast to intra-axial hemangiomas that show a bleeding incidence of 25%.
The most frequently found signs for hemangiomas in the literature are involvement of the oculomotor nerves. Visual involvement or of the fifth cranial nerves by neuralgia, or facial hyposthesia is infrequent (4,16,26,30). In addition, neurological deficits might be unspecific and even diplopia might be not constant; it might be present, and with less physical activity of the patient it may subside, but then it comes back again. In contrast, meningiomas of the cavernous sinus the presence of neurological deficits is frequent. They are usually silent, but may produce mass effect towards the seler region, against the cerebral parenchyma, and displacement of the middle line when they are large, which is frequent in most cases reported in the literature and may cause hydrocephalus (10,13). Furthermore, they may produce displacement of the midbrain, resulting in motor involvement and red nucleus lesion, causing a rubral tremor - which is a continuous up-and-down short displacement movement as case in the above-mentioned patient was due to movement of her left hand.

The hemangiomas of the cavernous sinus differ from the intra-axial angiomas, although their pathological structures are similar (Table 1). The difference lies in the behavior of these two lesions and, as such, they must be treated as different pathologies. The cavernoma or cavernous sinus hemangiomas being a vascular tumor mass in contrast to the cavernous angiomas, these are also vascular lesions, although small (8,12,17).

The growth mechanism for these lesions is similar to intracerebral hemangiomas. There is a progressive ectasia, endothelialization, angiogenesis within an intraleision hematoma. There is also glyotic reaction of the surrounding tissues by the blood, which is a stimulating factor for the proliferation of connective tissue (17). This reaction facilitates an autonomous vascular development on the borders of the lesion, promoting its growth (4,18).

The radiological examination always shows a meningioma-like lesion. The patient’s cranial scan displayed an unspecific lesion in the parasellar region, extending into the middle fossa. According to reports in the literature the lesion may be found on CT Scans hyperdense, hypodense or isodense, and it captures contrast homogeneously (4,10,15,18,19,26). The Magnetic Resonance Imaging (MRI) is the study of election and it shows a hipointense or isointense lesion in T1, and markedly hyperintense in T2 as the LCR, a characteristic that distinguishes this lesion. With the contrast the border of the lesion and the lateral wall of the cavernous sinus are enhanced (4,26). Although it is a vascular lesion, the angiographic study does not show it clearly, only a tenuous filling with scarce capillaries is visualized, very small for the size of the lesion. It is worthwhile pointing out that there is a nutrient artery; this is the meningohypophyseal artery, but this is also not visualized. It is describe that these lesions may be visible in the late venous phase as a tumoralike shadow (15,16). For hemangioma in the region, it is very important to differentiate between real meningiomas and this kind of tumor, since hemangiomas do not have the characteristic tail of contrast enhancement along the tentorial edge, which is missing in this case. A further important fact is that, despite the dislocation of the ICA in the parasellar compartment, its diameter is not decreased, which might happen in some highly vascularized meningiomas too, but in the majority of meningiomas it does not.

Due to its anatomic and imaging characteristics it is almost impossible to pre-surgically differentiate this lesion from a clinoid meningioma (4). Aneurisms of the cavernous sinus, giant adenomas of the pituitary gland, and tumors of the neural sheath are the pathologies for a differential diagnosis (15,17,30).

Surgery is indicated in all symptomatic patients or in those patients having large lesions with compression of the cranial nerves and parenchyma (13,30). The objective is the complete removal of the lesion, without causing neurological deficit (13,30). It is recommended to make a wide orbitozigomatic approach. There is a golden rule for surgery in the region, i.e. that the piece-meal...
Figure 5: Cerebral IRM with T2 information where we observe a giant lesion in the right parasellar region that engulfed and displaces the internal carotid artery.

La técnica es una necesidad. Sin embargo, si se realiza la técnica de retención fragmentaria de un hemangioma, es incorrecto. No es correcto porque el sangrado es tan profuso que la disección de las CNs es extremadamente difícil, si es posible en absoluto. Es claro, pero en tal caso de resección de las CNs, los déficits resultan como regla y la pérdida de sangre es muy alta. El hemangioma cavernoso es la única enfermedad en el espacio parasellar que debe ser cuidadosamente diseccionado de los nervios y coagulado, y solo cuando los nervios se deslicen del tejido y se excluyan los principales, entonces la lesión puede ser tomada. El tumor es identificado y se desliza por el plano de separación dado por la cápsula. En caso de sangrado puede controlarse con esponja de celulosa oxidada (Surgicell), hipotensión y electrocauterio o bipolar. Hay informes de autores que usan inyecciones de fibrina para controlar el sangrado durante la cirugía. La resección en bloque es recomendable para evitar un sangrado masivo (1, 2, 7). Durante la disección es necesario mantener en mente las CNs; estas suelen estar siempre en el centro. En la mayoría de los casos, la carótida está envuelta en el tejido. Durante la disección, el botón arterial debe ser identificado - en la mayoría de los casos, es la arteria meningohypofisaria - este debe ser desligado y coagulado. El seno no debe ser empaquetado con Surgicell, ya que este movimiento dificulta la reseción subsecuente del tejido y además aumenta el riesgo de lesionar las CNs (30). Se realiza una resección extradural del menor ángulo del hueso esfenoidal y del anterior clianoide. Es incorrecto si el dura es abierto antes de que se hayan completado los actos mencionados de cirugía (27). El dura debe ser abierto al final de la cirugía, cuando el tumor se ha removido mediante un enfoque extradural exclusivo, y de course when the tumor is located intraduralmente, pero esto es una ocasión rara. El dura se abre en el semicírculo (25, 26, 28). La valleja Sylviana se abre y el lóbulo temporal se mueve hacia atrás. La resección microquirúrgica es el tratamiento de elección; sin embargo, esto muestra un alto índice de mortalidad (15, 22, 26, 30). Se debe esforzarse por realizar una resección en bloque.
Figure 6: Pathological study of the lesion that confirms that the lesion is a cavernous malformation. A: Histologically all of the samples showed a vascular lesion that is constituted by anastomosed channels covered by endothelium. No atypical cyto-nuclear or mitotic activity was found. B: Between the vascular channels we found fibroblastic cells without atypical cyto-nuclear or mitotic activity. C: Deposit of mixioid material. D: Presence of few l mfoplasmocitic inflammatory cells. E: Thick venous vessels. F: Dilated Capillaries. G: Occasional vessels with micro-thrombus. H: In the periphery of the lesion some neural roots are observed. I: Dense fibrotic tissue compatible with the dural layer is found. J: Immunoperoxidase study for Epithelial membrane antigen(EMA) is negative. K: CD34 is positive in the endothelium of the vessels of this lesion, and negative in the previously described cells between these structures. L: CD31 shows the endothelial recovering of the vessels and don’t show ant atypical or proliferative nature of these vessels. This finding favors the fibroblastic character of the lesion and rule out the presence of an angiomatose meningioma, hemangiopericytoma and solitary fibrous tumor.

Figure 7: Control MRI T2 information. We observe a residua lesion in the right cavernous sinus with extension to the middle fossa.
Reporte de Casos

Revista Chilena de Neurocirugía 34 : 2010

Table 1: Differences between a cavernous sinus Cavernoma and an Intraaxial Cavernous Malformation

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Recibido: 07.04.10
Aceptado: 30.04.10

These treatments had showed to be effective, safe and consistent with tumor shrinkage. Radiosurgery is an excellent alternative to operative intervention and may even replace operative procedures if tumors are small in diameter or when the recur (11,14).

The bad prognosis associated with these lesions is improving, from a surgery mortality of 38% to 14.8% in the last series (6,15,26,30). When some remnant of the tumor is left, recurrence may occur.

Conclusions

Radiotherapy and radiosurgery are treatment options that have shown some results in a reduced series of patients. There is not sufficient evidence to use these routinely (2,29). These therapeutic methods are recommended when there is tumor remnants adhered to important structures that could not be resected, and in those cases of small and incidental lesions (3,11,15,28,20).

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