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Ruptured intraventricular dermoid cyst in a 33-year-old patient, diagnosis and surgical management. case report and literature review

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Introduction

Brain tumors constitute 20% of all tumors in the body. They can be primary or secondary. The total incidence of primary CNS tumors corresponds to 21.42 per 100,000 inhabitants, distributed with 5.43 per 100,000 inhabitants in patients between 0 and 19 years of age and 27.85 per 100,000 inhabitants in patients older than 20 years of age, with the highest incidence in the later ages. Of these tumors, 66% are benign tumors and 44% are malignant.[1,2] Among the most frequent locations of primary CNS tumors, the meninges are in first place with 36.1%, contrasting with the ventricular system which has one of the lowest prevalences with 1.1%.[1–3] The clinical manifestations of intracranial tumors depend directly on their location, size, degree of infiltration, extension and edema. According to these factors, they can have different forms of presentation due to symptoms caused by endocranial hypertension, irritative or deficit symptoms.[1–3] They can be classified in different ways, one of them regarding their relationship with the brain parenchyma (intraaxial/extraaxial) and also in relation to the tentorium (supratentorial/infratentorial).[2,3]

Dermoid cysts are non-neoplastic primary tumor lesions, frequently congenital and slow-growing. They derive from embryonic ectodermal tissue during neural tube closure, between the third and fifth weeks of embryonic development, therefore, they are of ectodermal inclusion. They are rare lesions that represent between 0.04 and 0.6% of intracranial tumors.[4,5] The cyst wall is composed of a capsule of stratified squamous epithelium containing dermal elements such as sebaceous glands, sweat glands and hair follicles. The presence of such elements in the wall distinguishes dermoid cysts from epidermoid cysts.[6] They usually occur in childhood and are more common in males. They are usually located in the midline or parasagittal region of the skull, although their most common location is the spinal canal.[7] Patients become symptomatic in old age. The most frequent symptoms are seizures and cephalalgias.

Clinical case

Female patient, 33 years old, with a history of obesity, cholecystectomy, appendectomy, under follow-up in the neurosurgery polyclinic since 2010 for intraventricular tumor with left frontal predominance, possibly dermoid, in imaging study Figure 1. Consultation at the Emergency Department of the Hospital Herminda Martin on September 16, 2020, due to a 24-hour evolution of holocranial headache, pulsatile, abrupt onset, refractory to analgesia, associated with nausea and dizziness. Glasgow scale 15 points, without neurological deficit. A non-contrast CT scan of the brain showed significant hydrocephalus and progression of the volume of a fatty mass towards the horn of the left lateral ventricle measuring 4.1 x 3.5 x 2.2 cm. and with evidence of a similar image at the level of the temporal horns of both lateral ventricles measuring up to 5 mm. icrosurgical management was decided and performed on 10/6/20 with the support of ultrasound in order to delimit the tumor and check tumor resection, without incident, and the patient was subsequently admitted to the surgical PCU with GSC 15 without focality Figure 2, 3.

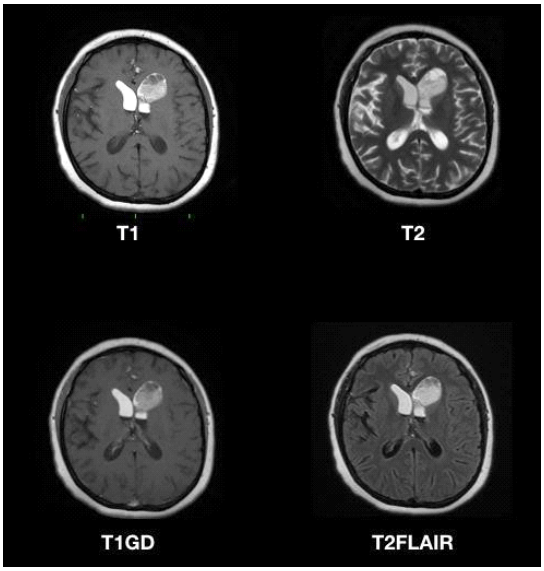

Figure 1 Magnetic resonance imaging with gadolinium taken in January 2020 of the present year, showing dermoid cyst in the different sequences.

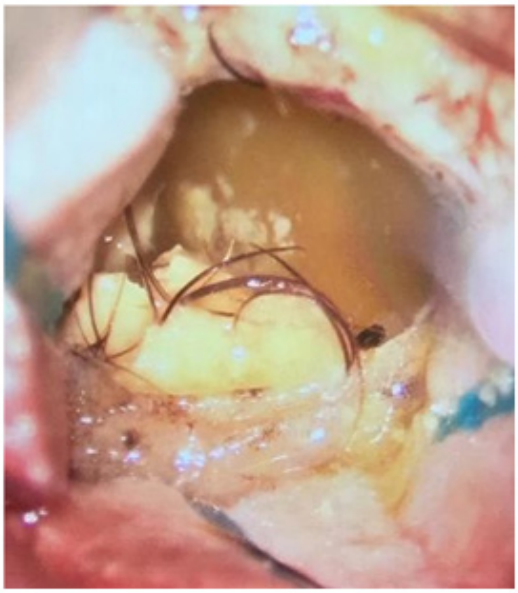

Figure 2 Cystic lesion with fanereos.

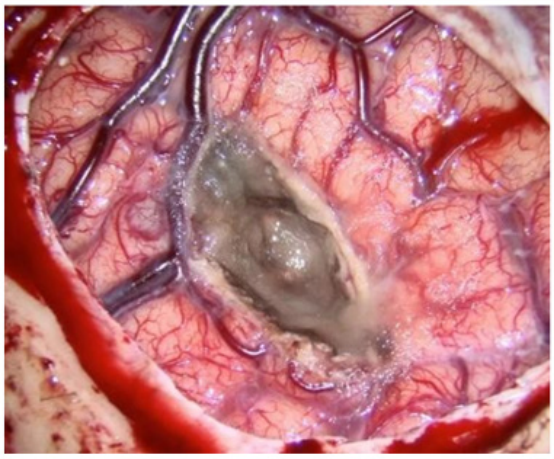

Figure 3 Surgical bed post tumor resection. Edges with absorbable hemostatic fabric.

Discussion

From a ventricular point of view, primary lesions of intraventricular location are relatively infrequent as mentioned above. The most frequently described tumor lesions located in lateral ventricles are meningiomas, papillomas, astrocytomas and subependymomas, being dermoid cysts infrequent. Clinically they will present a syndrome of endocranial hypertension due to obstructive hydrocephalus.[1,3] In relation to embryonal cell tumors, dermoid and epidermoid cysts correspond to this histological type. These are generally benign lesions that originate from the inclusion of ectodermal elements at the time of neural tube closure, which occurs between the third and fifth week of embryological development.[5] They represent between 0.3% and 1.5% of all intracranial tumors, being dermoid tumors 4 to 9 times less frequent than epidermoid tumors. They differ from each other by the presence of dermal structures such as sebaceous glands, sweat glands and hair follicles in dermoids, findings explained by the fact that the dermal and cerebral lines come from the same embryological layer. On the other hand, the absence of these adnexal findings in the dermis leads to the suspicion of epidermoid cyst, helping in the differential diagnosis.[3,9] Both types of cysts form well-demarcated tumors of the cerebral parenchyma.[3]

Dermoids have a medial location and are more frequent in the perisellar region and posterior fossa. As for those of intraventricular location, it is debated whether they are primarily intraventricular tumors or if they penetrate the cavities from the cisterns of the base or periventricular sulci.[8] The clinical presentation of dermoid cysts depends on the factors mentioned above and, therefore, is directly related to their location and size. They increase in size due to epithelial desquamation and glandular secretion, generating edema and displacing adjacent structures (cranial pairs, for example).[5] The most frequent symptoms are convulsions and cephalalgias. Tumor rupture may be asymptomatic or give rise to recurrent chemical meningitis due to spillage of its contents into the subarachnoid space and ventricular system, manifesting with seizures, vasospasm with cerebral infarction and death.[4,8]

For imaging and diagnostic follow-up, MRI has become a fundamental element and has positioned itself as the test of choice due to its image quality to differentiate tissue densities. However, the low availability of this test in the centers of the country, the time it takes to perform the test and the economic accessibility represent a limitation at the time of its use.[10,11] In MRI dermoid cysts show hyperintensity in sequences obtained in T1 due to their fat content consisting of triglycerides and unsaturated fatty acids and variable signal in T2, occasionally with a liquid-fat level. Therefore, CT is more frequently used for diagnosis and study, thanks to its greater availability, lower cost and shorter examination time. For these same reasons, it is considered the test of choice for emergency contexts.[11,12] In CT scans, due to the ectodermal content of the cysts, they will be observed as hypodense and well delimited masses, which may have calcifications. Rarely they may appear hyperdense.[4]

Regarding the case presented, the low incidence of dermoid cysts alone is already remarkable and the infrequent location of intraventricular cystic tumor lesions is noteworthy. This would even allow considering it as an incidentaloma in early stages and asymptomatic.[11] In this case, 2 main differential diagnoses were studied: an epidermoid cyst, which was ruled out by the evidence of dermal attachments such as hairs associated with the cyst, and secondly a teratoma, which is less likely because although these are tumors that are also formed from germ cells (in this case totipotential), their evolution would not coincide with a slowly progressive picture of 10 years as in the patient.[1] Being a benign tumor with low proliferative grade, it could be classified as grade 1 according to the classification of CNS tumors established by the WHO (although the results of the histopathological study are necessary to confirm this with certainty).[1] Under this background of benignity of the tumor lesion at the time of diagnosis in 2010 it was decided to perform expectant medical management associated with periodic controls.

The definitive treatment of dermoid cysts is purely surgical, being of choice the excision of the tumor by open microsurgery guided preferably by intraoperative ultrasonography or excision by neuroendoscopy. The open or endoscopic approach will be decided according to the criteria of the medical team, considering mainly the size of the tumor lesion to make the decision. It is important for the neurosurgeon to remove the tumor with a precise safety diameter to avoid possible recurrence, but also trying to generate as little damage as possible to the healthy brain parenchyma. Nodules associated with tumor lesions should always be removed under the same principle of avoiding recurrences. In this case, since the patient presented symptoms associated with hydrocephalus, it was decided to perform

a microsurgical resolution. Although the size of the lesion would have allowed endoscopic resolution, microsurgical technique was decided to increase the percentage of tumor resection. The aim of this management is to avoid the development of hydrocephalus associated with the natural evolution of endocranial hypertension syndrome, which would culminate in herniation and the consequent death of the patient.[1,3]

The patient's post-procedure evolution was favorable, thanks to the minimally invasive techniques that prevail in current neurosurgeries, where ultrasound guidance is a great contribution for the neurosurgeon at the time of performing the intervention. Currently the patient is discharged with the corresponding care and indications at home.

Conclusion

Dermoid cysts are primary CNS tumors with a low incidence and in this case with an unusual presentation with respect to intraventricular location. A national registry of brain tumors is needed to know the real regional prevalence and incidence of these tumor lesions along with others. The diagnosis of these cysts is the one used for CNS tumor lesions, favoring MRI as the gold standard, but CT is also useful in cases where imaging resources are not available or in the context of emergency. The treatment of choice for these lesions is neurosurgery. The treatment route will be decided according to the criteria of the medical team in charge in order to achieve a good outcome. This will prevent possible complications associated with the tumor lesion that may compromise the patient's life.

Acknowledgments

None.

Conflicts of interest

The author declares there is no conflict of interest.

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