

Clinical and Radiological Presentation of Giant Supra and Infratentorial Dermoid Cyst with Left Trigeminal Neuralgia: A Case Report and Literature Review

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Abstract

We report the unique case of a right supra and infratentorial dermoid cyst discovered in 50 years old female with a right V2 trigeminal neuralgia symptomatology. The patient underwent of MRI/ CT scan studies observed the presence of the unclear image of epidermoid / dermoid cyst, leading to its subsequent resection. We describe and discuss the imaging characteristics of dermoid cysts the particularity in radiographic diagnosis, surgical approach, the pre/postoperative management and review the relevant literature.

Keywords: trigeminal neuralgia, supratentorial, infratentorial, radiographic, surgical

Introduction

Background

Intracranial dermoid cysts are benign, with a slow growing pattern, derived from ectopic inclusions of epithelial cells during closure of neural tube. These lesions, represents around 1% of intracranial tumors, with characteristic computed tomography (CT) and magnetic resonance imaging (MRI) appearances that usually seen in the midline structure of the brain, other localizations are uncommon and the cyst can be misdiagnosed with other tumors.

Here we report a case of a 48-year-old woman presented to us with trigeminal neuralgia of right branch symptomatology over the past 5t years. A baseline non-contrast computed tomography (CT) scan performed showed medial hypodense image suggestive of dermoid cyst. When realize magnetic resonance imaging (MRI) revealed the lesion that behaves hypointense on T1WI and hyperintense on T2WI.

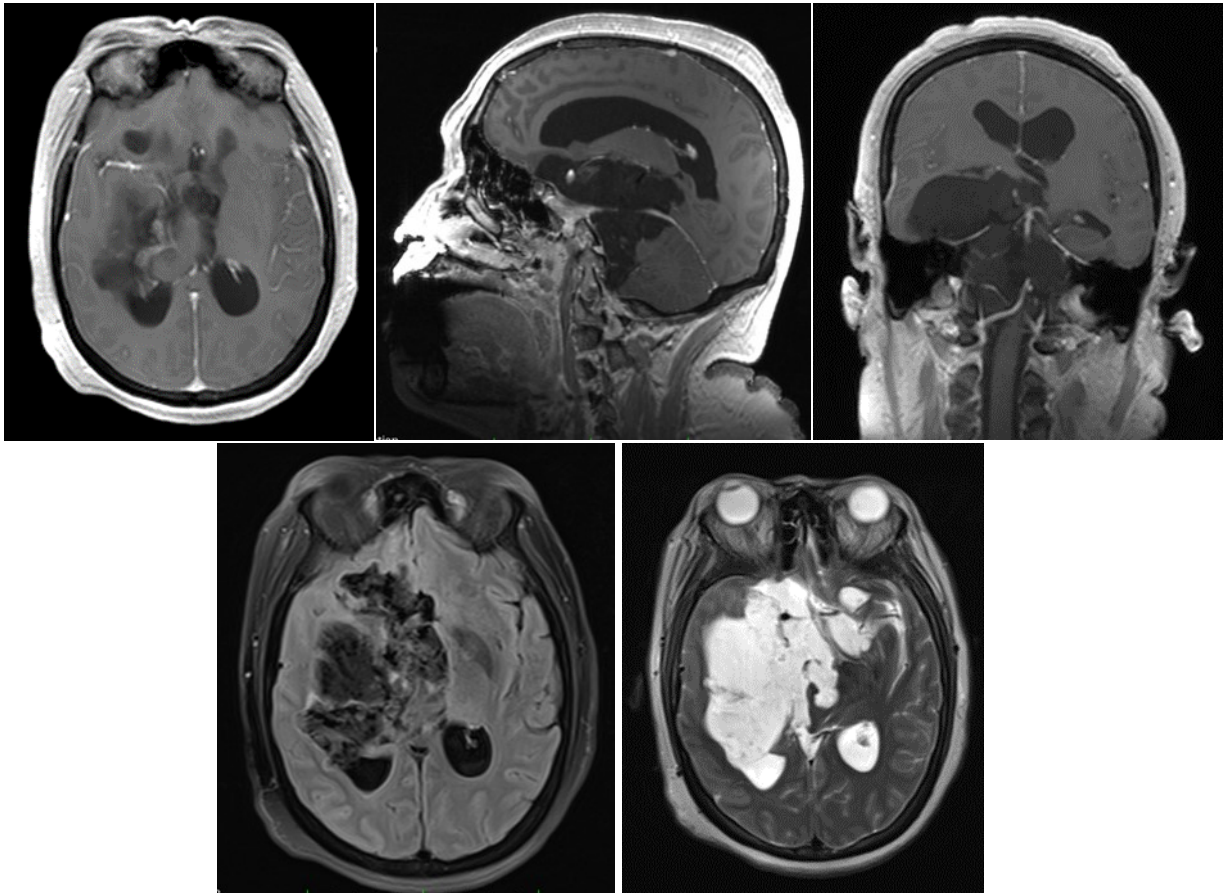
Case Report

A 50-year-old woman who presented to the emergency room due to headache, asthenia, adynamia, general discomfort and sleepiness of one week of evolution, in addition the family mentioned personality changes, disorientation episodes and tendency to drowsiness in the previous 6 months. In her medical history she had trigeminal neuralgia in management with carbamazepine, hypothyroidism and arterial hypertension in control with losartan and levothyroxine respectively, depressive symptoms without established management. In the initial neurological examination she arrived with GCS 14 points due to time disorientation, cranial nerves revealed hypoesthesia along the V1 and V2 right territory and hyperesthesia V1 left territory, without further cranial nerve deficits, left corporal hemiparesis 4/5 danieles scale without other neurological deficit.

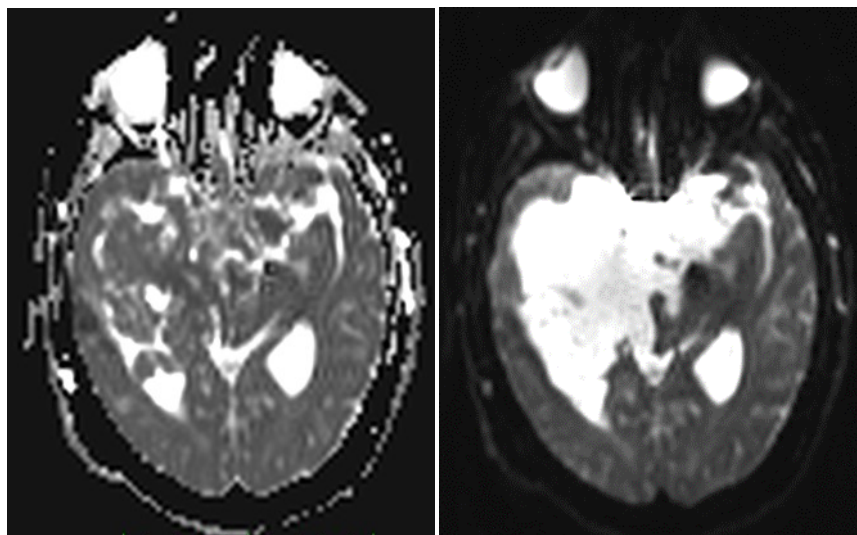
The initial approach included a simple CT where observed a hypodense supra and infratentorial extraxial heterogeneous imagen with respect to cerebral parenchyma that ranges from prepontine cisternae, right pontocerebellar angle to anterior recess of third ventricle occupying the mesial portion of ipsilateral temporal lobe, conditioning displacement of midbrain, basal ganglia with right to left vector.

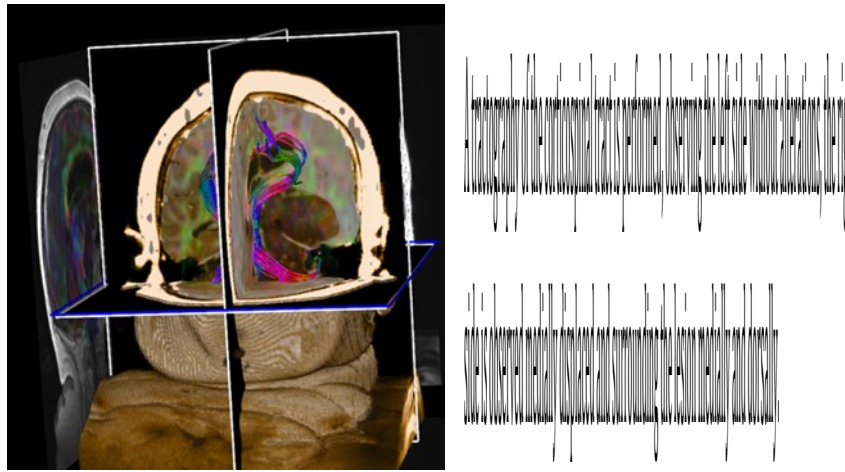
In the MRI observed a supra and infratentorial occupational lesion located from the right peribulbar cistern with extension of the ipsilateral canal auditory and basal cisterns to the sellar cistern and third ventricle, it behaves hypointense and heterogeneous in T1 sequence and hyperintense in T2, with restriction to diffusion and without reinforcement to contrast, dimensions 69x74x84 mm in dorsoventral, rostrocaudal and latero lateral direction respectively.

Patient was taken up for surgery. Right pterional craniotomy and transsylvian approach with subtotal removal of the mass was done. The patient presented an episode of seizures in the immediate postoperative period and five days later another one despite anti-seizure management, on the seventh day she presented data of CSF fistula in the region of the surgical site, for which she is managed with antifistula treatment for 7 days more after that patient recovered and was discharged in satisfactory condition. Histopathological findings were consistent with Dermoid cyst.



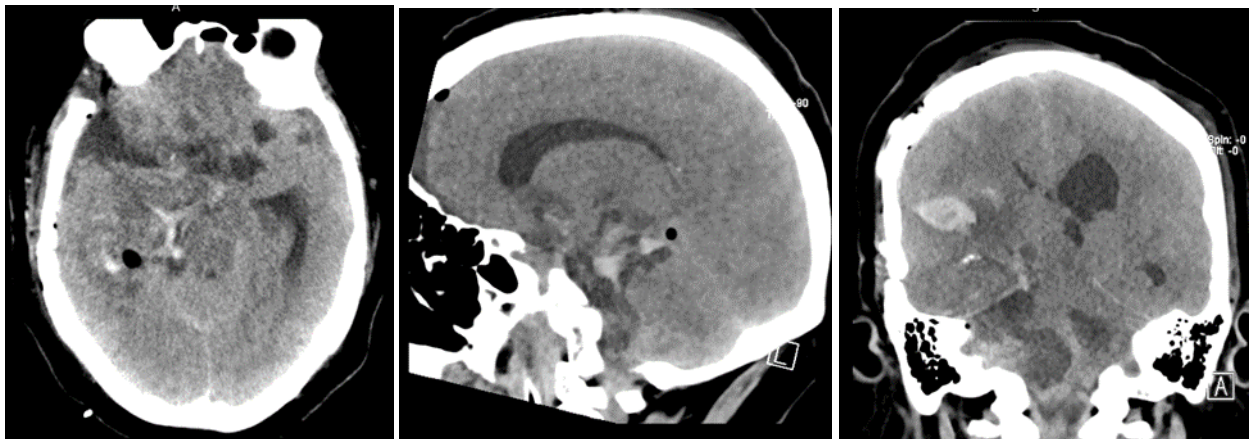
Contrast MRI is performed where observed in axial sagittal and coronal T1 weighted, T2 and FLAIR slices occupational lesion located at the level of infratentorial and basal cisterns, predominantly on the right, causing compression and involucre of structures of the brainstem, the lesion behaves heterogeneous hypointense in T1 sequence hyperintense in T2, presents restriction to diffusion and without reinforcement after contrast administration, wraps the fifth right cranial nerve in its cisternal portion.





A tractography of the corticospinal tract is performed, observing the left side without alterations, the right side is observed medially displaced and surrounding the lesion medially and dorsally.

Simple CT is observed in axial sagittal and coronal sections with data of probable CSF fistula especially in Simple CT is observed in axial sagittal and coronal sections with data of probable CSF fistula especially in axial image, hypodense and heterogeneous residual image at infratentorial level in the last two sections with and image suggestive of residual hematoma.



Discussion

Intracranial dermoid cysts are rare, benign, slow-growing congenital neoplasms and are believed to arise from ectopic ectodermal cell rests incorporated in the neural groove at the time of closure, with a dermis-like capsule containing squamous epithelium and adnexal structures (e.g., hair follicles, sebaceous, apocrine and sweat glands). Cysts develop gradually, with the accumulation of glandular secretions and epithelial desquamation.

Location Supra and infratentorial dermoid cysts are distinctly uncommon, According to Liu et. al., The location of the tumor was quite variable. In this study they founded that dermoid cyst are located most common in the peri-Sylvian region, pineal region, the suprasellar cistern, the frontal lobe, and the petroclival region, respectively the latter with significant brainstem and temporal lobe compression, however some Supratentorial intracranial dermoids generally arise with a tail in the suprasellar region with the body of the cyst expanding into the surrounding frontal and temporal lobes with mass effect.

Radiologic features

A dermoid cyst typically contains variable amounts of fat, hair and sebaceous secretions, which determine their imaging features, are usually particularly hypodense on CT scan with a range Hounsfield unit of -20 to -140, due to their lipid content. Calcification is frequently present, and the tumor does not enhance after the administration of contrast medium. Occasionally, they appear hyperdense mimicking a hemorrhage. On MRI, they are typically hypointense on T1-weighted image and vary from hypo- to hyperintense, and non-homogenous lesion on T2 weighted images according to Abrar A Wani et.,al. They typically have a high signal on fluid-attenuated inversion recovery images and are moderately restricted on diffusion-weighted images. Differential diagnosis of dermoid cyst are epidermoid cyst, arachnoid cyst, and cystic craniopharyngiomas, lesions that appear heterogeneous content.

Demonstration of fat in the dermoids and particular signal characteristics help differentiate it from others lesion like arachnoid cyst and cystic craniopharyngiomas furthermore the localization that usually are encountered off midline.

Intracranial dermoid and epidermoid cysts are usually considered to be two different entities in the radiological and surgical literature according to embryological origin. Epidermoid cysts are classically located off midline with isointense to cerebrospinal fluid on T1 and T2-weighted behaves images and have restricted diffusion, whereas dermoid cysts are located classically midline, have T1-hyperintense regions due to the presence of fat and show facilitated diffusion.

Preoperative radiological features and differential diagnosis must be concern especially with epidermoid cyst due that both are inclusion tumors with important differences between the two. Dermoid usually present and are treated in pediatric age group while epidermoid tend to present in second to fourth decades of life. The appearance is dependent on the relative lipid and protein content of the cyst. In general, dermoid have higher lipid content, but this is not always the case and radiological differentiation from epidermoid cyst is never conclusive. According to Jaskaran Gosal et al., epidermoid can be classified into two types based on their different radiological picture -classical "black" epidermoid and the atypical "white" epidermoid. These classical epidermoid are referred to as black epidermoid. White epidermoid are atypical in that they due to their high protein content and consequently high viscosity, they are hyperintense on T1WI sequence, hypo- to hyper-intense (commonly hypointense) on T2 and show no diffusion restriction. Result complex to make the difference preoperatively between the white epidermoid and dermoid as both of them have similar radiological characteristics. Diffusion-weighted sequence is a good tool due that dermoid cyst typically shows diffusion restriction while the atypical "white" epidermoid do not.

Clinical features depend on their location, seizure and headache being the most common with uncomplicated supratentorial dermoid cyst, patients can present with headache, nausea, vomiting, papilledema, cerebellar signs such as ataxia and dysmetria, cranial nerve palsies (usually of nerves 6 and 7 in our case the lesion surrounded right trigeminal nerve) bradycardia and hypertension and others mental and systemic alterations. Can occur aseptic meningitis secondary to the dissemination of the cyst content, however, rupture of intracranial dermoid cysts is a rare phenomenon the spillage of contents that may further lead to an inflammatory response, chemical and recurrent meningitis, abscess formation, extradural empyema, increased intracranial pressure resulting in local and systemic complications, actually the exact pathophysiological mechanism of the rupture is unknown, can occur spontaneously, or sometimes secondary to closed head trauma, or iatrogenic surgical complications. Stendel and associates hypothesized that glandular secretions, possibly increased by age, dependent hormonal changes, may lead to rapid enlargement and rupture of these cysts.

Surgical Management

The complete surgical removal of the primary tumor capsule and intra-cystic contents that's mean remove the capsule, cyst contents for internal debulking, and decompression, and micro surgically dissecting the capsule from adherent, or adjacent neurovascular structures. Ideally, a plane of dissection can be developed between the capsule and overlying arachnoid, but the dermoid capsules commonly have a dense adherence to the brain parenchyma and vasculature. According Abrar A. Wani et,al cyst dermoids, in comparison to epidermoids are more adherent to arachnoid, which makes the development of a surgical plane and dissection difficult. If the tumor capsule is strongly adherent to surrounding neurovascular structures, subtotal resection should be considered, leaving the adherent portion intact to avoid vascular complications. Intraventricular, or subarachnoid fat does not seem to resorb and has been demonstrated to persist for years after time of rupture.

In our patient, CT, magnetic resonance, tractography and spectroscopy images were observed and the characteristics were not entirely consistent to clearly establish an accurate preoperative diagnosis between epidermoid cyst and dermoid cyst due to the heterogeneity of the lesion, the location, the restriction pattern, the age of presentation and without rupture data at the time of the initial and subsequent evaluation during hospitalization, subtotal resection of the lesion was performed in approximately 80% due to multiple adhesions to structures of the brainstem, adjacent to it, as well as regions that due to intraoperatively distorted anatomy represents a greater challenge, taking into account not to leave an added deficit in the postoperative period.

Patient presented CSF fistula which was managed by placing an epidural drain for 4 days and anti-fistula medical treatment with acetazolamide and head position for 7 days, leaving the service with no fistula data and with clinical neurology and rehabilitation evaluations due to seizures presented in the immediate postoperative period. Which represents and early and late complication in the management of complex supra and infratentorial cystic lesions.

Conclusions

Still difficult to differentiate preoperatively between the white epidermoid and dermoid as both of them have similar radiological characteristics.

A rare case of supra and infratentorial dermoid cyst is described, with detailed presentation of preoperative imaging. These lesions are rarely described but can occur, with various clinical presentations. The described case shows that large volumes can be reached for this non-midline, slow-growing lesion, before any clinical signs can be identified.

Gross total excision remains the gold standard, allowing a rapid decompression of the brain parenchyma, however, it should be taken into account to perform subtotal resections due to the intraoperative complexity of lesions adhered to adjacent structures to avoid consequences in the patient as well as reduce early and late complications that may occur.

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