Epidermoidcyst of the pinealregion: report of a case

Abstract :

Intracranialepidermoidcysts are one of the rare intracranialtumors. Theyrepresent 0.2–1% of intracranialtumors and 7% of cerebellopontine angle tumors. The pineal location isexceptional for this type of tumor. In 1928, Cushing was the first to report the pineal location of the epidermoidcyst. We report a clinical case concerning a 30-year-old young man whopresentedwithintracranial hypertension for 6 months. A clinicalexaminationfound stage III papilledema at the fundus, and the rest of the examinationwasunremarkable. MRI showed a pinealregion process. The patient underwentsurgery and underwent total excision of the lesion.

Introduction :

Epidermoidcysts (EC), alsocalled “primarycholesteatomas” or “pearled Cruveilhier tumors," are rare, benigncongenitaltumorsdevelopedfromectodermal inclusions [1]. The classic locations are: the cerebellar pontine angle (half of the cases), the temporal fossa, the suprasellarregion, and the quadrigeminalregion. The location in the pinealregionis rare.

Materials and methods:

We report the case of a 31-year-old patient whohadintracranial hypertension for 6 months. The clinicalexaminationfound a conscious patient GCS 15 withoutsensory-motordeficitwithpapilledema stage III in the fundus. A brain CT showed a lesiondeveloping in the pinealregion, hypodense, not enhancingcontrast, withhydrocephalus. MRI showed a process in the pinealregionwithregular contours, well-limited, hypo-intense in T1, hyper-intense in T2 (Figure 1), heterogeneous FLAIR, filling the supra-vermiancisternwith mass effect on the vermis, and Sylvius aqueductwith tri-ventricularhydrocephalus. The patient benefitedfrom a first ventriculo-cisternostomy (VCS), and then, through a supra-cerebellarinfratentorialapproach, hebenefitedfrom a macroscopically total excision of a tumorwith a pearlyappearance. The evolutionwasmarked by a good improvementwith the completedisappearance of clinicalsigns; radio-clinicalfollow-up wasrecommended (Figure 2).

Figure 1: Brain MRI T2 sequence axial and sagittal section showing a hyperintenselesion in the pinealregionwithregular contours.

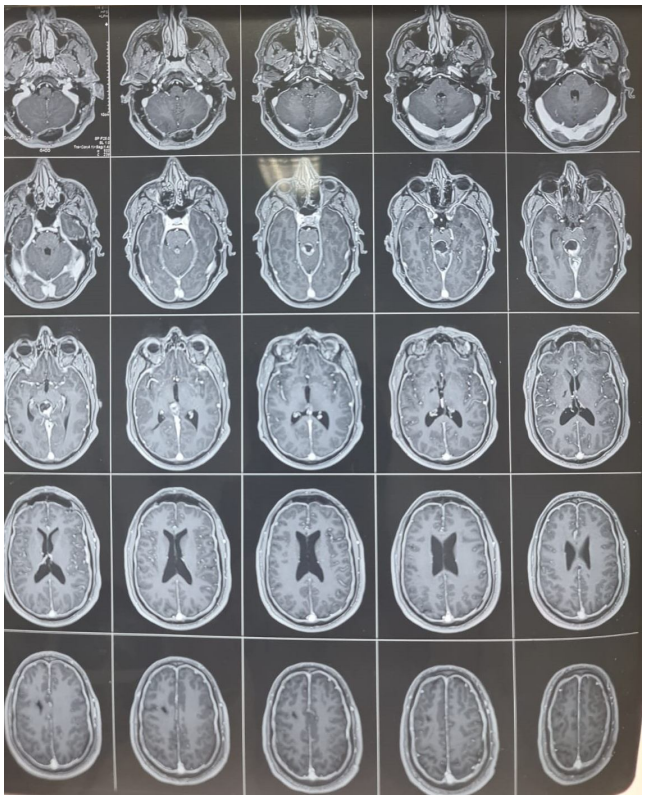

Figure 2: Axial T1-enhanced brain MRI, performedpostoperatively, showing a smallresidue of the epidermoidcyst in the pinealregion.

Discussion :

Epidermoidcysts, alsocalledprimarycholesteatomas or “Cruveilhier pearltumors," are rare, benign, dysembryoplastictumors; theyconstituteapproximately 1% of all primaryintracranialtumors [1]. Developedfromectodermal inclusions [1]. Theymostoftenresultfrom an incompletecleavage of the neural ectoderm and the cutaneousectoderm at the time of closure of the neural tube between the third and fifthweeks of gestation, withretention of ectodermalcells in the nervous system [1.2]. Theycan, more rarely, besecondary to post-traumatic or iatrogenicpenetration [3] of the epidermis at the level of the subarachnoidspaces.

This lesionisoftenlocated at the cerebellopontine angle, while the dermoidcystprefers the medial location [4]. Pineallocalizationis a very rare form of thisintracraniallesion. It represents 0.2–1% of all intracranialtumors [5]. Cushing was the first to report the pineal location of the epidermoidcystin 1928 [5]. Then, manyotherauthors have reportedsome cases of pinealepidermoidcysts [12]. Until 1974, nine cases werereported in the literature [13]. In 1999, 11 cases wereanalyzed by Mackay et al. [14]. Sincethen, manyother articles have been publishedfocusingprimarily on surgicaltreatment.

The clinicalpresentationisoftencharacterized by Parinaud's syndrome and hydrocephalus. Hemiparesis and cerebellarsignsmayalsobenoted [14].

Previousstudies have describeddepressivesymptoms in pineal injuries [6], as well as a case of schizophreniacuredafter the excision of the lesion [6].

An unusualpresentation for this rare tumor in an adolescent isdiplopia and lateralrectus muscle paralysis [7].

Theselesions are made of a pearly white, soft mass surrounded by a capsule, oftenadherent to the adjacent walls. The cystic content isavascular and presents, whencut, a yellowish content of more or lessviscousconsistency, reminiscent of candle wax and arranged in concentricstrips [8]. The lesiongrowsslowly and has a flexible and deformablecharacter, adapting to the spaces in whichitevolves [8]. Cystsgrowthrough the progressive desquamation of epithelialcells, whichtransformintokeratin and cholesterolcrystals.

A CT scan shows a cysticlesion. The densityissimilar to that of cerebrospinalfluid. Wecanfind a lesion of the quadrigeminalcisternsometimescausinghydrocephaluswithoutenhancementafter injection of contrastproduct. Variable imagingis due to the difference in cholesterol and protein content and the presence of hemorrhage. On MRI, the epidermoidcystishypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR images withoutcontrastenhancement [4]. Diffusion sequences (DWI) candifferentiatebetween an epidermoidcyst and an arachnoidcyst [14]. In diffusion, epidermoidcysts are brightcompared to othercysticlesions [5].

The main point of surgicaltreatmentis the radical excision of the epidermoidcystwithits capsule. However, itis a real challenge due to this location. Someauthorsprefer to intentionallyleave fragments of the adherent capsule in situ in the deepveins of thisregion to

avoidanyrisk. Konovalov et al. [4] specifiedthat radical excision wasonly possible in 50% of the cases presented in thisseries. Twoapproacheweredescribed by Yasargil [4] in the surgical management of pinealepidermoidcysts: the supra-cerebellar infra-tensorialapproach and the occipital trans-tentorialapproach. The latter ispreferred for a lesionwith a significantsupratentorial component [4,5].

The supracerebellarinfratentorialapproachis the mostcommonlyused for surgicaltreatment [4]. Otherapproaches are used, including the trans-callosalinterhemisphericapproach [9], the trans-ventricularapproach [5], and the combined supra-infratentorialapproach [14]. The ventriculoperitoneal shunt couldbeused in certain cases of hydrocephaluswithintracranial hypertension [5]. Therapeuticstereotaxic aspiration isalsoproposed for the treatment of epidermoidcysts. Kitchen et al. [5] reported a case of VP shunt and stereotaxic aspiration. This technique still has manydisadvantages. First of all, aspiration does not detach the capsule, whichrepresents a high risk of recurrence, spontaneous rupture of the cyst, asepticmeningitis, and malignant transformation of the epidermoidcyst.

The direct surgicalapproachappears to be more useful for these patients. Mackay [14] analyzed 12 reported cases of pinealepidermoidcystssince 1968. The outcomewas good in 10 of the 12 cases. There are two cases of asepticmeningitis. One deathwasrecorded.

As part of postoperative monitoring, diffusion sequence MRI allowspreciseverification of the completeness or incompleteness of the excision [8]. Two complications canmodify the evolution of KE: rupture and malignantdegeneration [8]. Cystic rupture is the mostfrequent complication; itismostoftensecondary to surgical manipulations, more rarelyspontaneous, and manifests as asepticchemicalmeningitis. As for malignantforms, they are rare and exist in the form of squamouscellcarcinomas [11].

In the event of incomplete excision, the growth of the residueis as slow as that of the native tumor; itneverthelessrequiresannual monitoring, allowingitsevolvingpotential to beassessed [1].

Conclusion :

The pinealepidermoidcystis a rare tumor. The treatment of choiceis a total resection of the tumor. But sometimes, thisis not always possible due to the characteristics of the tumor and the pinealregion.

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