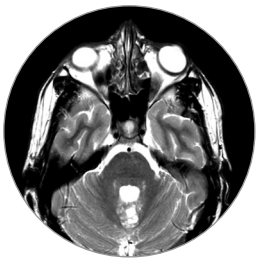

Multinodular and Vacuolating Posterior Fossa Lesions of Unknown Significance. Case Report in Colombia

Lesión multinodular y vacuolar de fosa posterior. Presentación de caso en Colombia

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Summary

There are multiple types of cystic lesions that can be found in the brain and in the posterior fossa. Among these, a new entity called MV PLUS (Multinodular and Vacuolating Posterior Fossa Lesions of Unknown Significance) has similar imaging characteristics to the vacuolating multinodular tumor, but as indicated by its acronym in English, it is located in the posterior fossa. These tumors are defined as a group of small, high intensity nodular subcortical images in the T2-FLAIR, with or without post-contrast enhancement. They can be differentiated from other cystic entities, because they are clustered lesions, without reactive inflammatory changes, in a cortex of normal appearance and without changes at follow-up. The latter, causes them to be considered benign, non-aggressive lesions. We present the case of a 21-year-old patient, who presented an incidentally found lesion in the central region of the vermis. MRI showed a multicystic-looking mass, that had no changes at follow-up in the last 3 years and without obvious differential diagnosis, suggestive of MV PLUS.

Resumen

Existen múltiples lesiones quísticas que se pueden encontrar en el encéfalo y en la fosa posterior. Entre estas, una nueva entidad denominada MV PLUS (por su sigla en inglés de *multinodular and vacuolating posterior fossa lesions of unknown significance*) tiene características imagenológicas similares al tumor multinodular vacuolante, pero como lo indican su sigla en inglés, está localizado en la fosa posterior. Estos tumores se definen como un conjunto de lesiones subcorticales nodulares pequeñas, de alta señal en secuencias con informaciónT2-FLAIR, con o sin realce al administrar medio de contraste. Pueden diferenciarse de otras entidades quísticas porque son lesiones agrupadas, sin cambios inflamatorios reactivos, presentes en una corteza de apariencia normal y sin evolución en el seguimiento. Esto último hace que se consideren lesiones benignas, no agresivas. Se presenta el caso de un paciente de 21 años de edad, a quien de manera incidental se le encontró en la región central del vermis una lesión de aspecto multiquístico, sin cambios durante el seguimiento en los últimos 3 años, con diagnóstico probable de MV PLUS.

Introduction

Multinodular and vacuolating posterior fossa lesions of unknown significance (MV-PLUS) (1) is a new tumor (2) that consists of clustered subcortical nodular lesions, with high signal in T2-FLAIR sequences, with or without contrast medium enhancement, without edema or inflammatory changes and that is located in the posterior fossa (1, 2). It is a type of vacuolating and nodular neuronal tumor, but, as the acronym indicates, it has a location in the posterior fossa. Recently, in a retrospective study of patients with suspected vacuolating multinodular neuronal tumors, Lecler and collaborators found that when this entity is located in the posterior fossa, in 82% of the patients there was a central area of low signal in T2 that they called "the sign of the central point", which could be useful for the identification of this new form of lesion (2).

Patients presenting this entity have been followed for a minimum of 24 months without any imaging changes, so this condition is considered to have a benign behavior (1-3).

Case presentation

This is a 21 year-old man, of mixed race, with a personal history of central hypogonadism and anosmia, who at the age of 18 had a nuclear magnetic resonance (NMR) scan of the Turkish chair due to the described history, with a diagnosis of Kallman syndrome: dysmorphia of the straight girdles, with markedly hypoplastic olfactory grooves. Incidentally, a lesion was found in the vermis.

When practicing cerebral MRI to visualize the lesion in the vermis, it was found in the central region an image of multi-cystic aspect conformed by millimetric,

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multinodular, rounded lesions, with high signal in T2-FLAIR sequence (figures 1 and 2), of low signal in sequences with T1 information, without edema, without contrast medium enhancement (figure 3), and without compressive effect on the fourth ventricle. In the spectroscopy -Multivoxel CSI 3D (TE 35 and TE 135) and Single Voxel SVS with TE 30 - carried out on the lesion, no abnormal metabolites were identified. In particular, there was no elevation of the choline; neither was there diffusion restriction or susceptibility effect. Although images were performed with high resolution T1 and T2 information, "the sign of the central point" was not identified. No supratentorial commitment was found. Periodic follow-up was recommended and in subsequent studies the lesion has remained stable, both in size and characteristics, for the last 3 years.

Discussion

The differential diagnoses of cystic lesions in the posterior fossa are multiple, among which we highlight:

- **Dysplastic cerebral gangliocytoma (Lhermitte-Duclos).** It has a striated pattern with alternating signal intensities, which correspond to the thickened folia. It can generate mass effect in the fourth ventricle and generally is a dispersed lesion with cerebellar hemisphere involvement, for which it was excluded as a diagnosis (4).
- **DNET (Dysembryoplastic neuroepithelial tumors).** Although it may have a multi-cystic aspect, it was discarded, since it is generally supratentorial and intracortical; it occasionally presents some degree of enhancement with the contrast medium and may have calcifications in up to 20% of the cases (4).
- **Parasitic cysts.** They are usually multiple and with a diffuse involvement (5). The lack of evolution towards other stages and the absence of reactive inflammatory changes make this diagnosis unlikely in the case presented.
- **Prominent Virchow Robin spaces.** Typically exhibit a signal similar to cerebrospinal fluid in all the sequences, while MVNTs are not suppressed in FLAIR sequence with T2 information as is the case in these peri-vascular spaces (1).
- **Hamartomas**. Also called nodular neuronal heterotopias, in MRI they typically exhibit the same normal parenchymal signal -although occasionally they may appear with high signal in sequences with T2-FLAIR information-, but they have a common location (6), which rules out the possibility of this being the case in the patient in this case.
- **Cerebellar Astrocytoma.** It is a frequent lesion in this age group, cystic, cerebellar, the edge of the cyst may or may not be enhanced with the contrast medium and is occasionally associated with a wall nodule. It has an eccentric location and involves a cerebellar hemisphere, which does not agree with the image of the patient in this case, so it was excluded as a diagnosis (7).

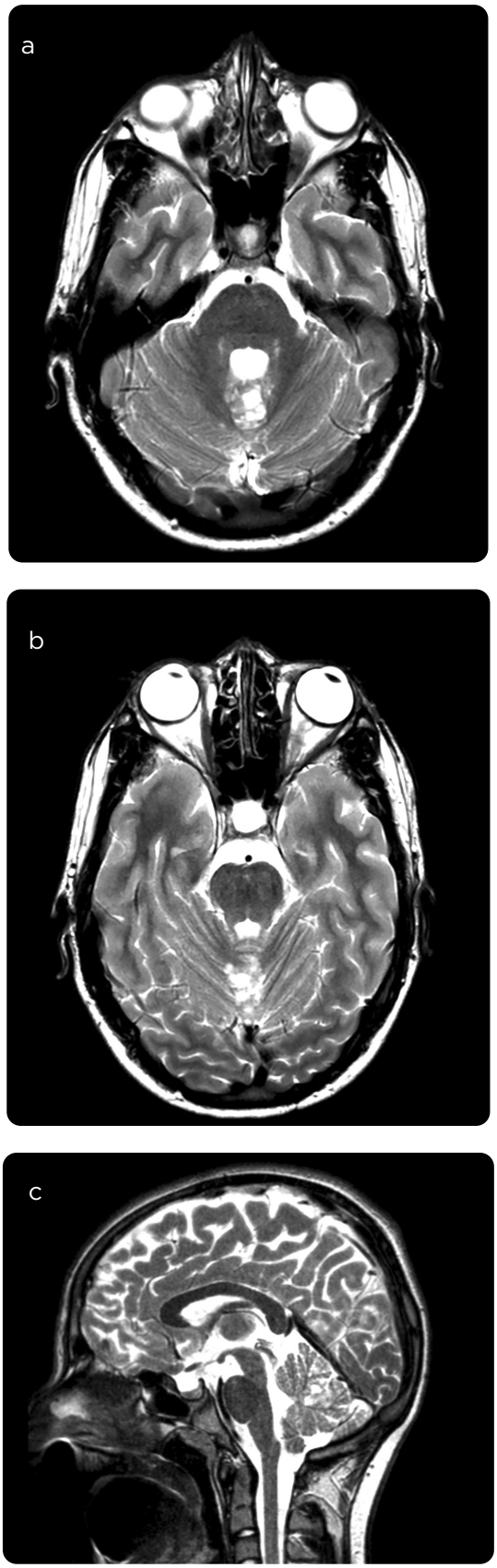

Figure 1. Multinodular images, coalescent images in the central region of the vermis, with a multi-cystic aspect, with high signal intensity in TSE sequence with T2 information.

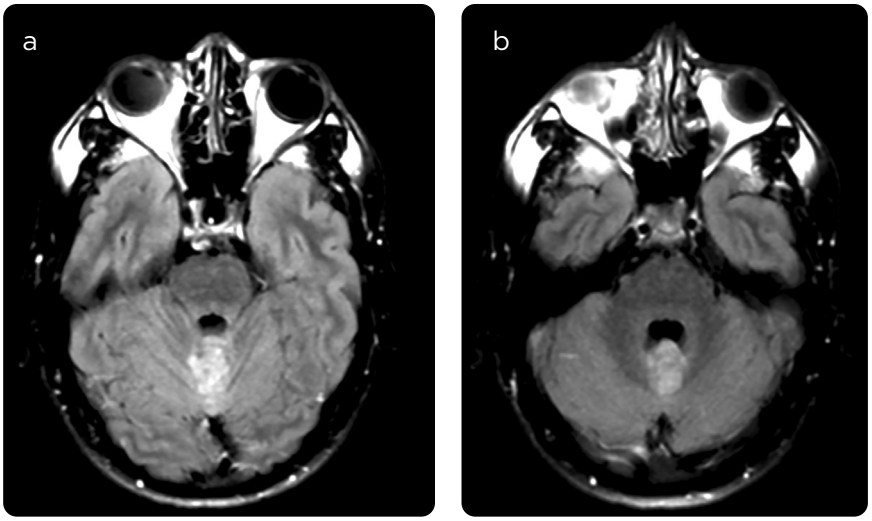

Figure 2. Multinodular images, coalescing images in the central region of multi-cystic appearance, with high signal in T2-FLAIR sequence.

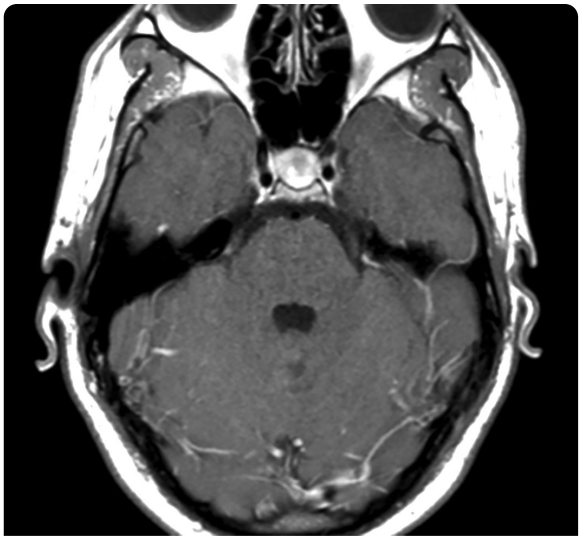

Figure 3. TSE sequence with T1 information with contrast medium. The multinodular lesion in the central region of the vermis is not enhanced with the administration of contrast medium.

In patients with congenital muscular dystrophies, subcortical cerebellar cystic lesions can be observed; in these cases the alterations are related to polymicrogiria and a variable pachygiric aspect, more in the frontal area. These patients generally present cerebellar cortical dysplasia; none of the latter are related to the patient's images (8).

Once the previous diagnoses were ruled out, it was concluded that the lesion visualized in our patient most probably corresponds to an MV-PLUS.

As it was exposed, within the differential diagnosis of cystic masses in the posterior fossa, the MV-PLUS should be included. Specifically, in nodular grouped lesions, of high signal in the T2-FLAIR, which do not generate mass effect, without enhancement with the contrast medium, without alterations in the metabolites in the spectroscopy and which are invariable in time.

The description of more cases, the follow-up of the same and the constant updating of the specialists in radiology, allows the adequate classification of lesions that are not so common and to know what type of follow-up and intervention the affected patients require. The study of this type of neoplasms and the description of a greater number of series should be continued, since it is considered that the lack of reports on this entity leads to under-registration in the medical literature due to reporting bias for lack of knowledge and because most lesions of this type probably do not dry out (1).

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