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Case Report

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BILATERAL VESTIBULAR SCHWANNOMA, ABOUT A CASE

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INTRODUCTION

Vestibular schwannoma is a slow-growing benign tumor that develops in the vestibulocochlear nerve. It was first described by E. Sandifort in 1777. It affects 1 per 100,000 people, is more frequent between the fourth and sixth decade of life, occupies between 80 and 90% of all tumors of the cerebellopontine angle. In cases of bilateral schwannomas, its prevalence ranges from 1: 210,000. In the event that the tumor is presented bilaterally, it is consitence with neurofibromatosis type II.

CASE PRESENTATION

Female of 33 years, with a history of hearing loss predominantly of the left side with six months in progress, accompanied by headache, tinnitus and gait disturbances, so she goes to a doctor who, after evaluation, indicates to perform an audiometry and magnetic resonance imaging study, that report left audiometric anacusia and right sensorineural hearing loss. Magnetic resonance imaging is requested that reports bilateral hypointense angle lesions in T1 and hyperintense sequences in T2 with marked reinforcement after contrast administration.

The lesion of the right side with measures of 3.2 x 3.0 x 2.7 cm and that of the left side with measures of 2.9 x 2.7 x 2.3 cm; both lesions compress the intracisternal path from the VI to the IX cranial nerve and displace the V pair. The lesion of the left cerebellopontine angle extends to the internal auditory canal, compresses the intracanalicular path of the acoustic state package and contacts the cochlea, findings compatible with bilateral vestibular Schwannoma. (Figure 1.)

It is referred to a neurosurgeon and is scheduled for surgery. A right retrosigmoid approach is performed with a biopsy result that confirms the diagnosis of acoustic schwannoma. However, the complete extraction of the same is not performed, so he decides to perform stereostatic brain radiosurgery with gamma Knife rays, initiating treatment to the lesion of the right side and three months after the left side, without complications during these.

The patient continued under evaluation for a period of 3 years, where they underwent brain magnetic resonance studies every year, which reported cessation of the growth of the lesions and in the last study carried out there was a slight reduction of the lesions, said lesions had measures 2.8 x 2.7 cm so it was concluded as a successful treatment. (Figure 2.).

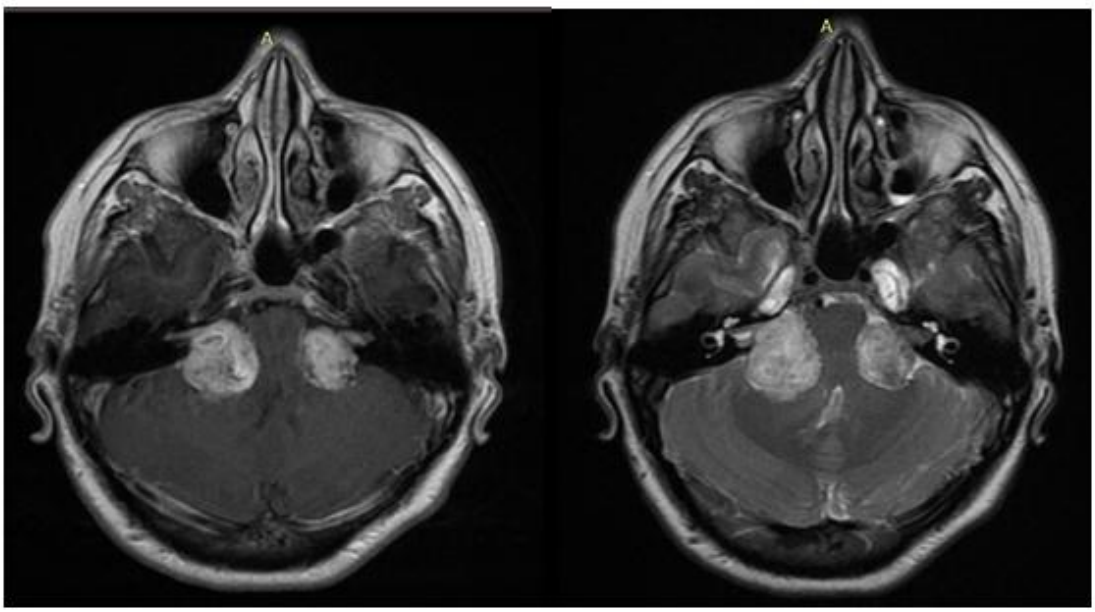

Fig. 1: An image is seen in axial section weighted in T1 with contrast (A) and T2 (B) showing a lesion of the cerebellopontine angle with bilateral intracanalicular component, findings in relation to acoustic schwannoma.

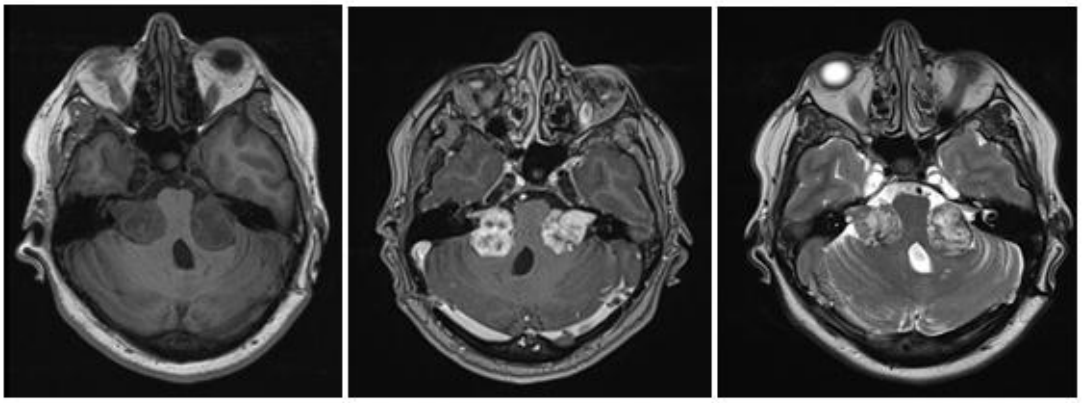

Fig. 2: Tumor is observed at the level of the cerebellar ponto angle after treatment with gamma knife that extends to the eighth cranial nerve, causes compression on both cerebellar peduncles and deforms the IV ventricle, the lesions enhance post contrast administration.

DISCUSSION

The acoustic neurinoma or vestibular schwannoma is the most frequent tumor of the internal auditory canal and the cerebellopontine angle. Vestibular schwannomas constitute approximately 10% of all intracranial neoplasms and about 60 to 90% of all tumors of the pontocerebellar angle. It has an approximate incidence of 1 in 100,000, with a peak incidence between the fourth and sixth decade of life, although cases have been reported in young patients.[1,3]

A high percentage of cases are unilateral and in cases where they occur bilaterally, a rare autosomal dominant disease is associated caused by a mutation of chromosome 22 called Neurofibromatosis type 2, in which other cranial, spinal and tumors occur in peripheral nerves, in addition to corneal opacity and skin abnormalities.[2,3,5]

The 95% of schwannomas are unilateral; Only 5% of cases are bilateral. As for age, the maximum frequency is between 35 and 40 years and there is a slight preference for the female sex.[9]

These tumors originate from the transition zone of the central myelin and the peripheral myelin (Obersteiner-Redlich zone). This transition zone is in the internal auditory canal. Schwannomas do not contain nerve fibers. They are benign nerve sheath tumors, typically encapsulated and well differentiated schwann cell compounds.[6]

The 80% of shwannomas originate in the internal auditory canal (IAC) and grow medially towards the cistocerebellar angle cistern (APC), expanding the acoustic pore. The remaining 20% originate in the cisternal portion of the eighth cranial nerve.[9]

The initial symptoms of vestibular Schwannoma include tinnitus, hearing loss and impaired balance, which can lead to compression of the cerebellum, hydrocephalus and facial paralysis.[8]

The diagnosis is made with the help of an adequate medical history, neurological examination and audiological tests such as tonal audiometry and tympanometry.[3.8]

Radiological studies include computed tomography of the skull, simple and contrasted, with parenchymal window and bone window in order to determine the specific characteristics of the tumor and bone at the level of the internal auditory canal, as well as its relationship with the sigmoid sinus.[7]

The test with the greatest impact corresponds to the diagnosis through magnetic resonance imaging (MRI) with gadolinium, currently constituting the gold standard in the diagnosis, also allowing the pre-surgical planning of the tumor.[1]

The most common findings in Computed Axial Tomography is that they have a spherical or ovoid shape and form an acute angle in the bone. CT shows a higher incidence of non-tumor calcifications, not detected in MRI in patients with neurofibromatosis type 2.[3,8]

In the MRI it is visualized in the T1 sequence isointense or slightly hypointense to the bridge. In T2 slightly hyperintense to the bridge and reinforce in an intense way with the contrast medium.[3]

Based on medical treatment, there are three therapeutic options for the patient with a vestibular schwannoma: observation with serial magnetic resonance imaging, surgery, radiotherapy and chemotherapy.[6,9]

CONCLUSION

Bilateral vestibular acoustic schwannomas are very rare tumors, which occur only in patients with type 2 neurofibromatosis.

It is important to emphasize that an early diagnosis of this type of lesions added to a timely treatment and a multidisciplinary medical management accompanied by a good imaging control, will lead us to offer our patients not only a better quality of life, but also to contribute to the development of a favorable prognosis in the evolution of the disease.

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