A 8 year old male patient was referred to our centre with a 2 months history of bilateral tinnitus, headache, nausea, and mild deafness. He had no family history of neurofibromatosis type 2. Otoneurologic examination revealed moderate sensorineural hearing loss. Magnetic resonance imaging (MRI) demonstrated two extra-axial low signal enhancing masses on the T1- and predominantly high signal on the T2-weighted images in the vicinity of the right and the left vestibular nerves (Fig. 1-3). These findings were evaluated as bilateral acoustic neurofibromatosis. The case was considered as neurofibromatosis type 2 without any concomitant abnormality in the central nervous system. Symptomatic medical treatment was initiated and the patient was referred to the neurosurgery department.

Discussion:

Neurofibromatosis 2 (NF2) is an autosomal dominant disease predisposing to the formation of multiple tumours in the central and peripheral nervous system. Vestibular schwannomas (VS) are considered to be the hallmark of the disease, but other tumours and ocular findings occur as well [1-4]. They are often bilateral and become symptomatic in the third or fourth decade of life. It should be noted that VS usually occur in the fifth or sixth decade of life in patients that do not carry the NF2 mutation. Schwannomas, meningiomas, ependymomas in the central nervous system, schwannomas of the peripheral nerves, cutaneous and ophthalmological manifestations are the other features of NF2 [1, 2, 4]. Disease phenotype is variable among patients. High degree of suspicion is mandatory for the diagnosis of NF2 in the paediatric age group. It should be considered in any child with multiple central nervous system or skin tumours without cafe-au-lait spots or Lisch nodules [1-3]. Given that the VS rarely produce the signs seen at the time of admission, careful examination of the skin and eyes following by gadolinium-enhanced MRI of the brain and spine should be carried out. Moreover, first-degree relatives should be examined as well [1-3, 5, 6]. Treatment and follow of NF2 patients up require oto-neurosurgical teams experienced in NF2 [4-6]. A clinical screening protocol and yearly and life time surveillance is suggested. Classically, only symptomatic lesions should be treated. Early proactive strategy against vestibular schwannoma has been advocate by some authors in order to preserve hearing. If the treatment is advisable, surgery remains the treatment of choice for tumours. Auditory brainstem implant must be taken into account in hearing rehabilitation [5, 6]. Main negative prognostic factors
include the young age at onset of symptoms and a high number of tumours at diagnosis.

**Differential Diagnosis List:** Neurofibromatosis type 2

**Final Diagnosis:** Neurofibromatosis type 2

**References:**


Figure 1

Description: demonstrates two hyposignal extraaxial masses locating at the cerebellopontine angles.
Origin:
Figure 2

Description: shows the enhancement of these two masses after the administration of the gadolinium.
Origin:
Description: discloses two predominantly hypersignal masses of the CPA. Origin: