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# Cranial Computed Tomographic Findings of Neurofibromatosis Type 2

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**Abstract:**

Neurofibromatosis type 2 (NF-2) is a rare inherited autosomal dominant disorder, characterized by multiple neoplasms of the central and peripheral nervous system associated with ocular abnormalities. NF-2 is associated with tumors of Schwann cells and meninges. Intracranial schwannoma most frequently involve the vestibulocochlear nerve (eighth cranial nerve). Bilateral eighth cranial nerve schwannomas are the hallmark of NF-2 and diagnostic of this condition. Intracranial meningiomas are also common in this disorder and are often multiple.We report a case of bilateral vestibulocochlear schwannoma and a solitary meningioma in a 17-year-old female who presented with headache and bilateral hearing impairment for last three months. Based on the clinical and computed tomographic (CT) findings, the diagnosis of NF-2 was made.

**Key words:** Tomography, Meningioma, Neurofibromatosis, Vestibulocochlear Nerve, Hearing Loss.

## Introduction

Neurofibromatosis is not a single entity but is actually a group of heterogeneous diseases [1]. Until 1987 neurofibromatosis type 1 and 2 were considered one single disease, but it was then demonstrated that the two disorders arose from different mutations in different chromosomes [2]. NF-2 is a rare disease with incidence of 1 in 33,000 to 40,000 and with approximately equal distribution in male and female [3,4,5]. It is an inherited autosomal dominant disorder associated with chromosome 22q12 [6]. It is suspected that approximately one-half of cases are inherited, and one-half are the result of new de novo mutations [7]. Spilberg *et al* reported a case of sporadic NF2, in which diagnosis was established based on medical history, clinical symptoms and image findings on magnetic resonance (MR) imaging [8]. The aim of this report is to present a case of NF-2 where diagnosis was established based on the clinical and image findings on cranial computed tomography.

## Case Report

A 17-year-old female presented with history of on & off headache followed by bilateral hearing impairment for last three months. There was no history of fever, earache, ear discharge, visual disturbance or trauma. There

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was no history suggestive of neurofibromatosis in her family. No cutaneous marker of the disease was found on examination. An audiometric exam showed bilateral sensorineural hearing loss, profound on the right and moderate on the left side. Her ocular findings were unremarkable.

Non enhanced CT (NECT) and contrast enhanced CT (CECT) scan of the cranium [Fig. 1, 2 & 3] showed well-circumscribed, ovoid, hypodense to isodense extra-axial masses in bilateral cerebellopontine (CP) angle cistern with its base on the posterior aspect of the petrous temporal bone in the region of the internal auditory meatus showing strong heterogeneous contrast enhancement with presence of small non enhancing intratumoral cystic areas. Image displayed in a bone window showed widening of the left internal auditory canal with erosion of its bony margins suggesting intracanalicular component of the tumor [Fig. 4]. The CT findings were consistent

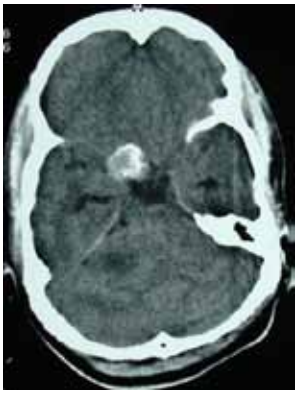

**Fig.1:** NECT showing an iso to hypodense mass in right C-P angle cistern; subtle isodense mass in left C-P angle cistern with presence of CSF cleft between the mass and cerebellum and partly calcified meningioma in relation to the sphenoidal ridge on right side.

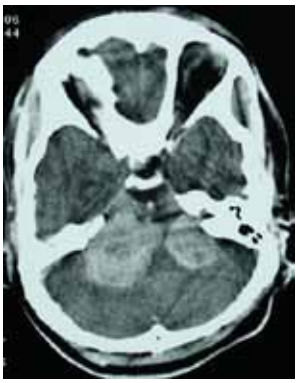

**Fig.2:** CECT shows strongly enhancing bilateral vestibulocochlear schwannomas with small intra-tumoral cystic areas.

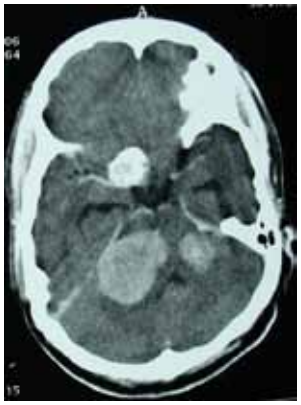

**Fig.3:** CECT shows moderate enhancement in the non-calcified portion of the meningioma seen in relation to the sphenoidal ridge on right side.

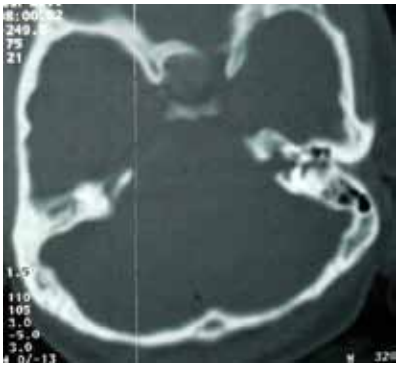

**Fig.4:** CT Image displayed in bone window shows widening of the left internal auditory canal with erosion of its bony margins suggesting intracanalicular component of the tumor.

with bilateral vestibulocochlear schwannoma. There was compression of fourth ventricle and mild dilatation of bilateral lateral & third ventricles. There was another well circumscribed partly calcified moderately enhancing round to oval mass in relation to the sphenoidal ridge on right side, consistent with a calcified meningioma. Based on the imaging & clinical findings, diagnosis of NF-2 was made and the patient was referred to Neurosurgery Center for surgical treatment.

## Discussion

Neurofibromatosis type 2, also known as NF-2 or "bilateral acoustic schwannoma," is a distinct form of the disease that must be separated clinically and radiographically from NF-1 [9]. The term MISME has been proposed to the NF-2 syndrome, due to multiple inherited schwannomas (MIS), meningiomas (M) and ependymomas (E). In addition to the neoplasms, posterior subcapsular lenticular opacity (juvenile cortical cataract) is often present. Patients with NF-2 may have cutaneous schwannomas that resemble skin tags, but they rarely have café -au-lait spotsand do not demonstrate the cutaneous neurofibromas like NF-1 [8].

The National Institute of Health (NIH) Consensus Committee has defined clinical criteria for NF-2. Bilateral masses of the eighth cranial nerve are diagnostic. A patient is also considered to have NF-2 if there is a first degree relative with NF-2 plus either a single eighth nerve mass or any two of the following: schwannoma, neurofibroma, meningioma, glioma, or juvenile subcapsular lens opacity [10]. According to Gutmann *et al*, presence of bilateral eighth cranial nerve schwannomas on MRI or CT scan fulfills the criteria for definite diagnosis of NF-2 and no biopsy is necessary [11]. Cutaneous markers are rarer in NF-2 as compared to NF-1. CNS lesions are seen in virtually all cases and include neoplasms (cranial nerves and meninges), non neoplastic intracranial calcifications and spinal cord nerve root tumors [10].

Several authors have studied series of cases to define incidence of the tumors in NF-2. Mautner *et al* studied 48 patients with NF-2, in which the prevalence of findings were: vestibular schwannomas (CN VIII) in 46 (96%), spinal tumors in 43 (90%), posterior subcapsular cataracts in 30 (63%), meningiomas in 28 (58%), and trigeminal schwannomas in 14 (29%) [12]. Aoki *et al* [13] reported cranial MR of 11 patients in which all patients had acoustic schwannomas, 8 had other cranial nerve tumors (5 multiple and 3 single) and 6 hadmeningiomas (4 multiple and 2 single).

Approximately 30-45% of patients with NF-2 are diagnosed because of symptoms resulting from eighth cranial nerve schwannomas, such as hearing loss, tinnitus, balance impairment, and weakness in seventh cranial nerve distribution. Often the first clinical sign of NF-2 is a sudden loss of hearing due to the development of bi- or unilateral vestibular schwannomas. The tumor causes symptoms by compressing or stretching the cochlear nerve, compressing the blood supply to the nerve or to the cochlea, or causing hemorrhage into the nerve or cochlea [8,14,15].

Our patient had bilateral eighth cranial nerve schwannomas and one intracranial meningioma with mild hydrocephalus. The schwannomas were the cause of her bilateral hearing loss and the headache was probably due to increased intracranial pressure. Although she had no family history, finding of bilateral schwannomas was diagnostic for the syndrome without the need of a biopsy.

## Conclusion

This rare case has been presented to highlight the clinical presentation and cranial CT findings of NF-2 where presence of bilateral vestibulocochlear schwannoma and meningioma fulfilled the diagnostic criteria, without the need of a biopsy.

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