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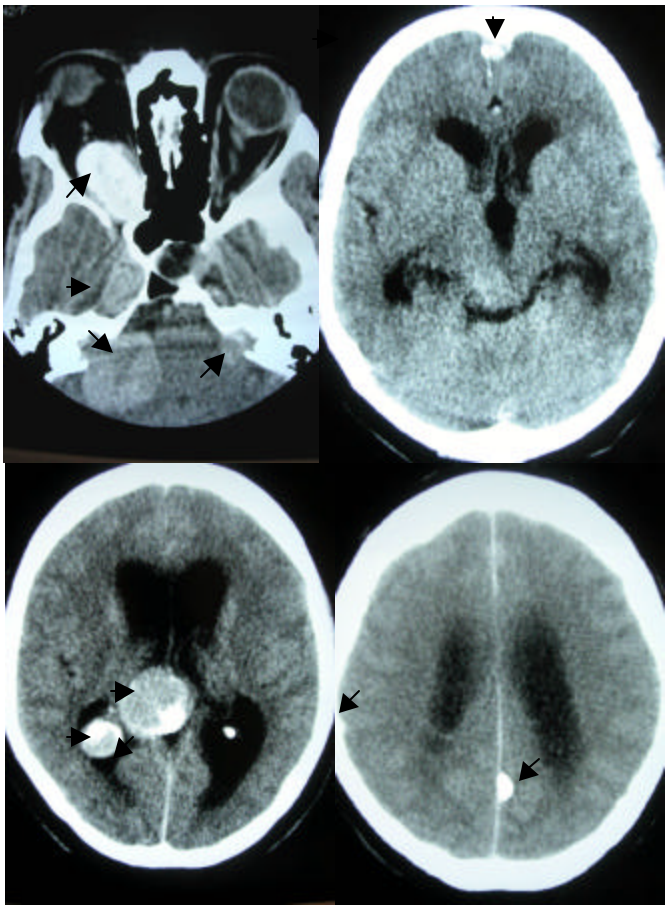
Neurofibromatosis type 2



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This 42-year-old woman presented with progressive loss of vision in the right eye, gradual diminution of vision in the left eye accompanied by gait ataxia and headache. On examination she had pale disc on the right side, and florid papilledema on the left side. Axial post contrast computerized tomography (CT) scans show (arrows show the various tumors in multiple locations) bilateral acoustic schwannomas, which are seen as enhancing large mass in the right cerebellopontine angle expanding the right internal auditory canal and smaller mass with similar characteristics on the left side. Enhancing mass in the right Meckel's cave area extending to the cavernous sinus region suggests trigeminal schwannoma. A calcified mass is seen encasing the right optic nerve and represents a right optic nerve sheath meningioma. Enhancing dural base masses in the convexity and parafalcine area suggest the presence of multiple meningiomas. Two partially calcified and enhancing intraventricular meningiomas are also seen in the right lateral ventricle. Hydrocephalus is also noted. In total 8 intracranial masses are seen.

Neurofibromatosis type 2 (NF2) is an autosomal dominant disease with an abnormality in the long arm of chromosome 22. The estimated incidence is 1 in every 50,000 people. NF2 is associated with tumors of Schwann cells and meninges. Bilateral vestibular Schwannomas, the hallmark of NF2, is the major finding in all individuals with NF2 and is diagnostic. Meningiomas (often multiple), glioma, neurofibroma, schwannoma and cerebral calcifications are other common associated findings.

References

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