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# Post viral unilateral isolated hypoglossal nerve palsy with cervical lymphadenitis: a case report.

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Keyword 1: unilateral hypoglossal nerve palsy.
Keyword 2: viral infections.
Keyword 3: magnetic resonance imaging.
Keyword 4: computerised tomography scan.

## Abstract:

Background/objectives: Unilateral hypoglossal nerve palsy is not an uncommon finding in neurological diseases. It is a rare condition in childhood where the mononeuropathies of the facial nerve are the commonest reported.

Setting: Department of Head and Neck oncosurgery, Kidwai Memorial Institute of Oncology , Bangalore.

Case report: A 12yr old boy was referred to our outpatient clinic with history of deviation of the tongue to the right and small enlarged lymph nodes in the right neck since 3 years.The deviation was seen after an episode of viral infection with rashes. Contrast enhanced CT and MRI scans were negative for any organic pathology. Excision biopsy revealed reactive lymphadenitis in the lymph nodes.

Conclusion: Isolated unilateral hypoglossal nerve palsy represents a formidable challenge and indepth patient history accompanied with good knowledge of hypoglossal nerve anatomy and a rational selection of diagnostic tests is necessary for making the diagnosis.

## Introduction:

*Unilateral isolated hypoglossal nerve palsy (IHNP)is not an uncommon finding in neurological diseases.[1] It provides a diagnostic and management enigma when the hypoglossal nerve palsy presents as an isolated entity as very few cases are reported in world literature.[2] Unilateral isolated hypoglossal nerve palsy is a rare condition in childhood where the mononeuropathies of the facial nerve are the commonest reported.[3,4]*

*Common etiologies in adult include intracranial and extracranial space occupying lesions, head and neck injuries, vascular abnormalities, infections, autoimmune diseases and neuropathies.[3] In pediatric age group the common etiologies include vaccination, aneurysms, trauma, dislocation of the vertebrae, intracranial tumors or viral infections like infectioious mononucleosis.[5] Only a few cases of idiopathic IHNP are reported and majority of these cases are reversible.[3]*

*We report a case of unilateral IHNP where all the other causes were ruled out and the diagnosis was narrowed down to a exathematous rash viral infection causing the palsy. An attempt to trace the nerve which is purely motor and innervates the intrinsic and extrinsic muscles of the tongue was made. Lesions which may include any of the five segments i.e, medullary(nuclear), cisternal (extramedullary intracranial), skull base( hypoglossal nerve canal), nasopharyngeal, oropharyngeal, carotid and sublingual space where it terminates supplying the lingual muscles were studied. Localizing the site of lesion along the course was done using contrast enhance CT scanning and MRI.*

## Case report:

*A 12-year-old boy with history of deviation of the tongue to the right and small enlarged lymph nodes in the right neck was referred to our head and neck outpatient department.(fig 1) The boy was aapparently alright 4 years back when he noticed small swellings on the right side of the neck. They were lymph node swellings and were painless. Over a period of time they increased in size and were discrete 4-5 in number.*

*An year after noticing the lymph nodes enlargement , a deviation in the tongue to the right was noticed. The patient and the mother noticed the deviation of the tongue after the episode of skin rashes and fever. Since then they have consulted many local doctors while the deviation of the tongue to the right remained unchanged. The deviation was associated with mild slurring of speech and chewing difficulty with evident muscle fasciculations.*

*No history of visual and hearing disturbance, deviation of the angle of the mouth and nasal regurgitation was noticed in the patient. Also complains of ocassional tinnitus in both ears , holocranial nonthrobbing headache, associated with cough and cold were seen. He gives a history of chicken pox one year back and otomycosis , wax impaction a year back. His medical history was unremarkable and there was no family history of IHPN or any other neurological disorders.*

*History of immunization not known, mother had tuberculous pleural effusion and was treated before marriage. His younger sister and brother were normal. On examination cervical adenopathy was seen in the right level II,III,IV & V with mild enlargement of the thyroid. The patient was concious and oriented to time and place. Optic fundus examination was normal, extraocular movements were normal, no nystagmus and tests for trigeminal and facial nerve were normal. Palatal movements were decreased on the left side. The masticatory muscles were all normal. On protrusion*

*of the tongue deviation and wasting with atrophy and fasciculations were seen on the right side. The tone of the upper and lower limbs were normal. The tone, power, reflexes, sensation and co-ordination in upper and lower limbs were normal. Also the deep tendon reflexes and plantars were normal. Oral and maxillofacial cause of any local structural disease was excluded. Tests for cranial nerves I–XI were intact except for decreased sensation in the distribution of the right lingual nerve. No signs of cerebellar disease were present.*

*Complete blood picture revealed Hb% b-11.6 g/dl, WBC 6.4x$10^9$/l, platelets 352x$10^9$/l. Random blood glucose was 87mg/dl, bloond urea nitrogen was 11.1mg/dl, serum creatinine was 0.4mg/dl. A normal liver function test revealed total bilirubin 0.2mg/dl, total protein 7.7g/dl, serum albumin 4.4g/dl, alkaline phosphatise 202U/l,AST 25U/l,ALT-17U/l, LDH 210U/l.*
*CSF analysis was in normal limits with slight elevation of proteins but no abnormal cells. No radiological features of tuberculosis or sarcoidosis was seen on chest x-ray. Echocardiography revealed a normal cardiac valves and chambers, with normal left ventricular function and detected shunt lesions.*
*Thyoid profile reported a bit raised TSH( T3-1.34ng/ml, T4- 8.5µ g/dl, TSH-5.57µIU/ml). Her liver and renal function tests were normal and non reactive VDRL.*

*FNAC done revealed non specific lymphadenitis in the nodes, and lymphocytic thyroiditis in the thyroid. High resolution sonography revealed multiple cervical lymphadenopathy bilaterally in level I,II and in right level V with loss of central hilum and hypervascularity with rest of the neck structures being normal.*
*Contrast enhanced CT of the skull base and the neck revealed multiple variable sized discrete lymph nodes at levels I-V on the right side and level I on the left side with minimal homogeneous enhancement and rest of the neck structures and carotids were normal. (fig 2,3) A radiological provisional diagnosis of lymphoma was made. Rapid ELISA test for HIV I,II and HBsAg was negative. Prothrombin time was 17.7 secs nd INR was 1.3 and partial thromboplastin time was 28secs. Lymph node biopsy was done under general anaesthesia.*
*MRI brain done in a multiplanar , multisequential pattern revealed normal supra and infratentorial brain parenchyma with no brain parenchymal focal lesions and normal ventricular systems. No obvious abnormality in the lower cranial nerves seen. Cisterns and the extra-axial spaces appear normal, with structures in the posterior fossa ,the cerebellum, brainstem and 4$^{th}$ ventricle appearing normal.(fig 5-8)*

*Commonest causes of isolated unilateral hypoglossal nerve palsy like the local vasculitic lesion or the cerebrovascular accident were ruled out. He was referred to the speech pathologist and asessment was done for speech and language therapy.*

## *Discussion:*

*The complex anatomy of the hypoglossal nerve nucleus is completely understood with the nucleus consisting of four topograpically distinct subnuclear columns.[1] The peripheral lesions can be classified topograpically into extramedullary intracervical lesions, hypoglossal foramen lesions, extracranial lesions at the base of the skull, and cervical lesions.[6]*

*After the exit from the hypoglossal canal, the nerve enters the carotid segement lying deep to the internal carotid artery, internal jugular vein , the 9$^{th}$ and 10$^{th}$ cranial nerve.[7] Thereafter, it lies in between the internal jugular vein and the internal carotid artery.[7] The lingual segment of the hypoglossal nerve, at the angle of the mandible, loops around the occipital artery to lie superficially, and at the level of the hyoid bone it lies over the hyoglossus after crossing the lingual vessels, in the sublingual space.[7]*

*Common lesions in the carotid space and lingual segment include schwannoma of the hypoglossal nerve, vascular anomaly of the vertebral or basilar artery, massive traumatic haematoma in the deep spaces, occipital condyle fracture, following extraction of third molar, occipital condyle fracture and head and neck injury.* [8,9]

*Isolated hypoglossal nerve palsy is rare due to its complex course and close proximity to other cranial nerves and vessels.*[10] *Isolated nerve palsy may denote a serious underlying pathology which shoud be meticulously investigated.*[3] *Freedman et al. has very rightly proposed that isolated hypoglossal nerve palsy is rare but should be regarded with suspicion.*[7] *With the positive history of a post viral infection isolated HNP we ruled out other causes of nerve palsies like skull base metastasis,retropharyngeal infections and surgical procedures in the neck.* [3] *Using multiplanar MRI conditions like Arnold-Chiari malformation, dural arteriovenous fistula of the transverse sinus, syringobulbia, carotid artery dissection or aneurysm and periostitis of hypoglossal canal were ruled out.*[3] *In dural arteriovenous fistula of the transverse sinus the patient may have hypoglossal palsy resulting from ischaemia in the region of the hypoglossal nucleus, as would thrombosis to the median branches of the vertebral artery.*[3]

*Keane et al in his study on 100 cases, the largest case series of hypoglossal nerve palsy, found neoplastic etiology in 49% of his cases, gunshot wound trauma in 12% cases, stroke in 6%, hysteria in 6% multiple sclerosis in 6%, post operatve in 5%,Guillain Barre neuropathy in 4% and infection in the rest 4% of the cases.* [1] *Combarros et al in their study in their study on 9 cases reported 3 cases to be metastatic origin, 1 arnold chiari malformation, i dural AV fistula and the rest 3 to be idiopathic etiology.*[2] *Marina Boban et al in their study on 4 cases found pseudo aneurysm of internal carotid artery, metastasis, clival tumor and acute disseminated encephalomyelitis as etiologies.*[11]

*Sharma et al, reported nasopharyngeal carcinomas, skull base metastasis, carcinomatous meningitis, trauma, dolichoectasia of the vertebral artery, dissection of the extracranial internal carotid artery and hypoglossal schwannoma as the most commonest causes.*[12,13] *In their 12 case study on isolated unilateral hypoglossal nerve palsies 4 had tubercular etiology and were successfully treated, one basal pachymeningitis, one aberrant ecstatic vessel compressing the nerve at the skull base, one rheumatoid arthritis and in the rest no cause was elicited.* [12,13]

*Idiopathic isolated hypoglossal nerve palsy has been reported by 3 authors and almost all recover over a period of years.* [14,15]
*Lee et. al, believe that self-limiting idiopathic hypoglossal nerve mimics Bell's palsy of the VIIth cranial nerve.* [14,15]

*Mujgan et. al, reported complete or partial recovery in only 15% of cases with with hypoglossal paralysis and recovery is very less in rapid onset paralysis without specific diagnosis or treatment.* [16,17,18,19]

## *Conclusion:*

*Isolated unilateral hypoglossal nerve palsy represents a formidable challenge and indepth patient history accompanied with good knowledge of hypoglossal nerve anatomy and a rational selection of diagnostic tests is necessary for making the diagnosis.*

*The possible diagnoses, may only reveal themselves after a number of investigations correlated with clinical findings. MRI is the diagnostic procedure of first choice to localize the lesion intracranially as well as skull base. MR angiography, CT angiography or digital subtraction angiography might add up additional diagnosis. CT skull base can be usefull initial procedure in a suspected expansile lesions. All patients in pediatric age group with hypoglossal nerve palsy must be carefully evaluated for systemic disorders and viral infections.*

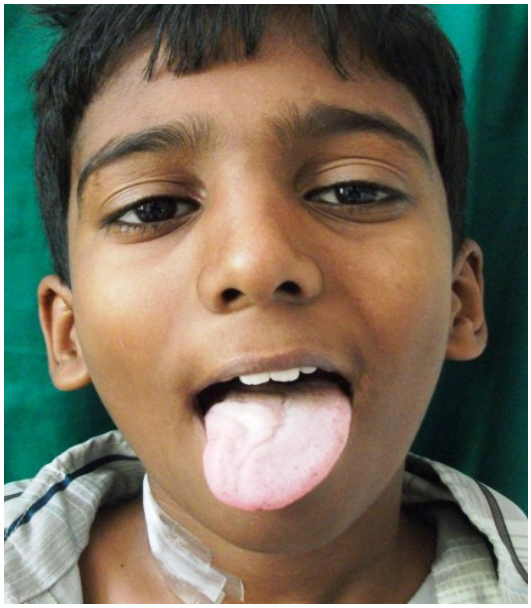

*Fig 1 Tongue deviation to the right seen on protrusion*

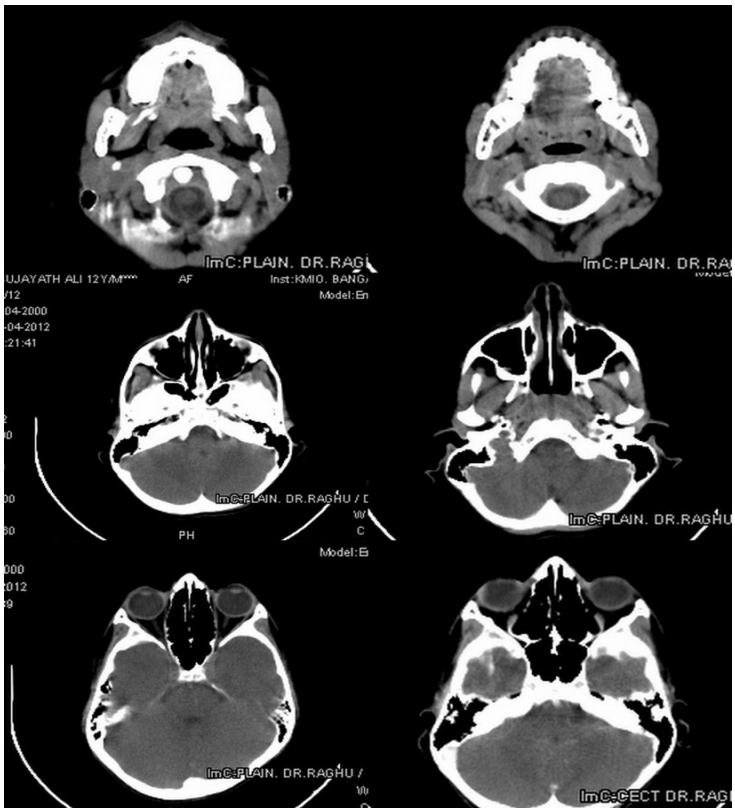


*Fig 2 Axial Contrast CT images showing normal structures*

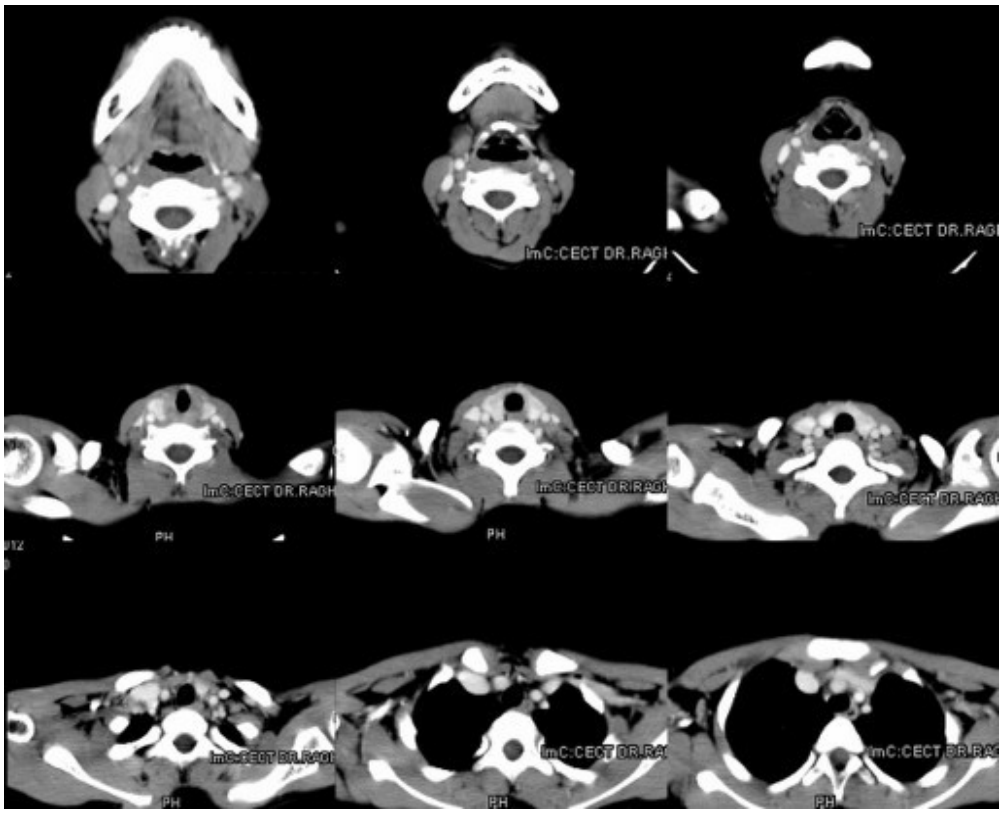

*Fig 3 Axial cuts of skull base, neck and upper thorax with contrast enhancement showing normal structures*

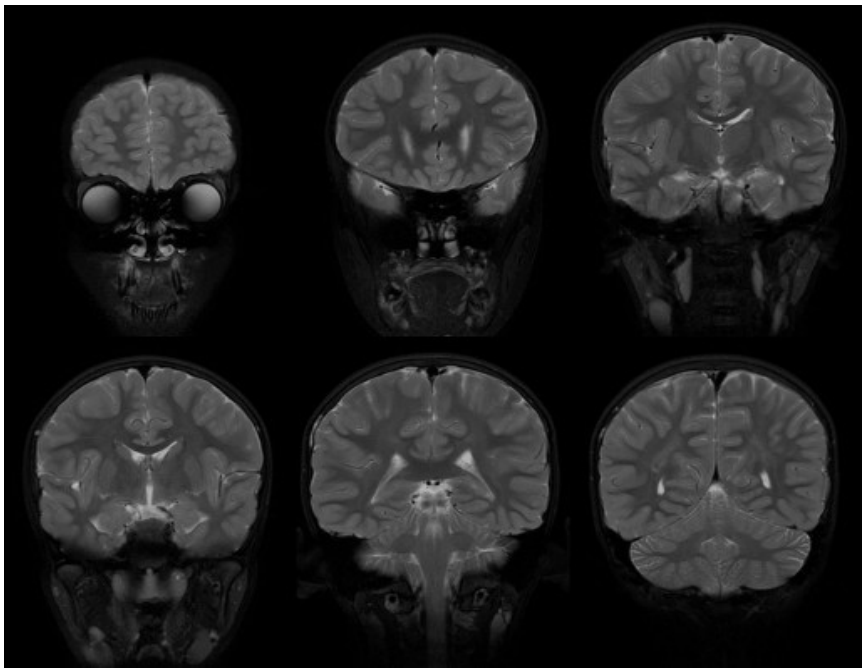

*Fig 4 Normal cerebral parenchyma seen on coronal Contrast enhanced MRI*

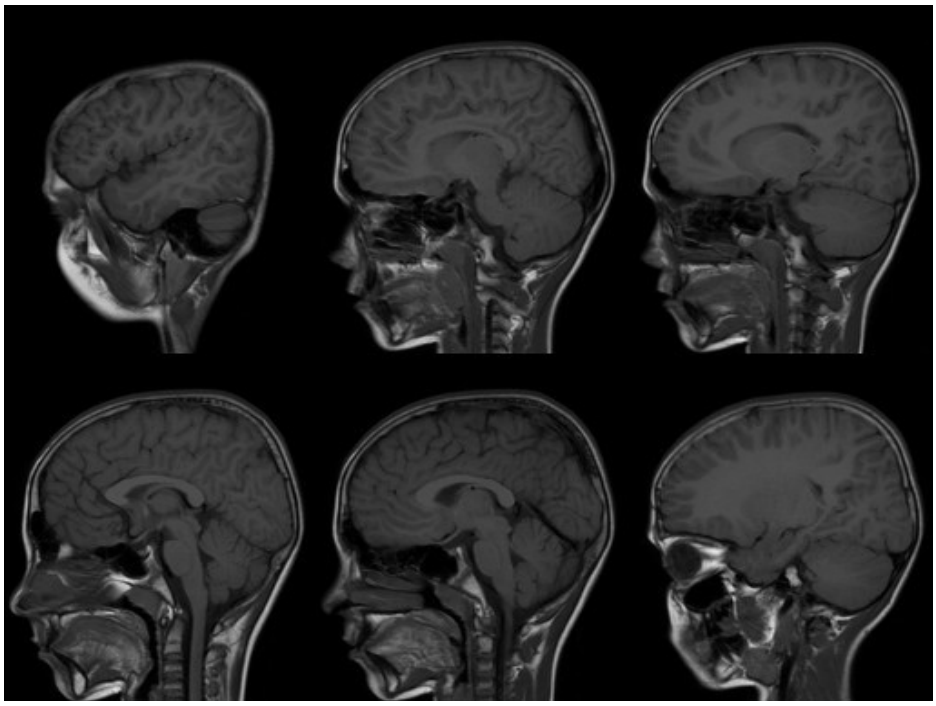

*Fig 5 Normal ventricular system seen on sagittal contrast enhanced MRI*

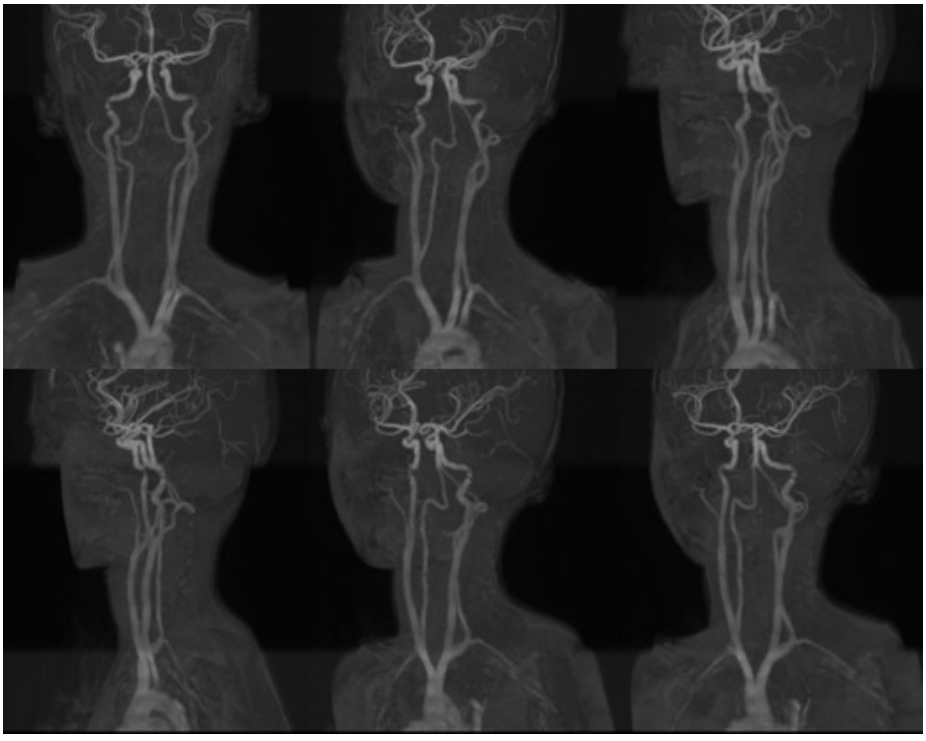

*Fig 6 Normal carotid and basilar system seen on  contrast MRI angiography*

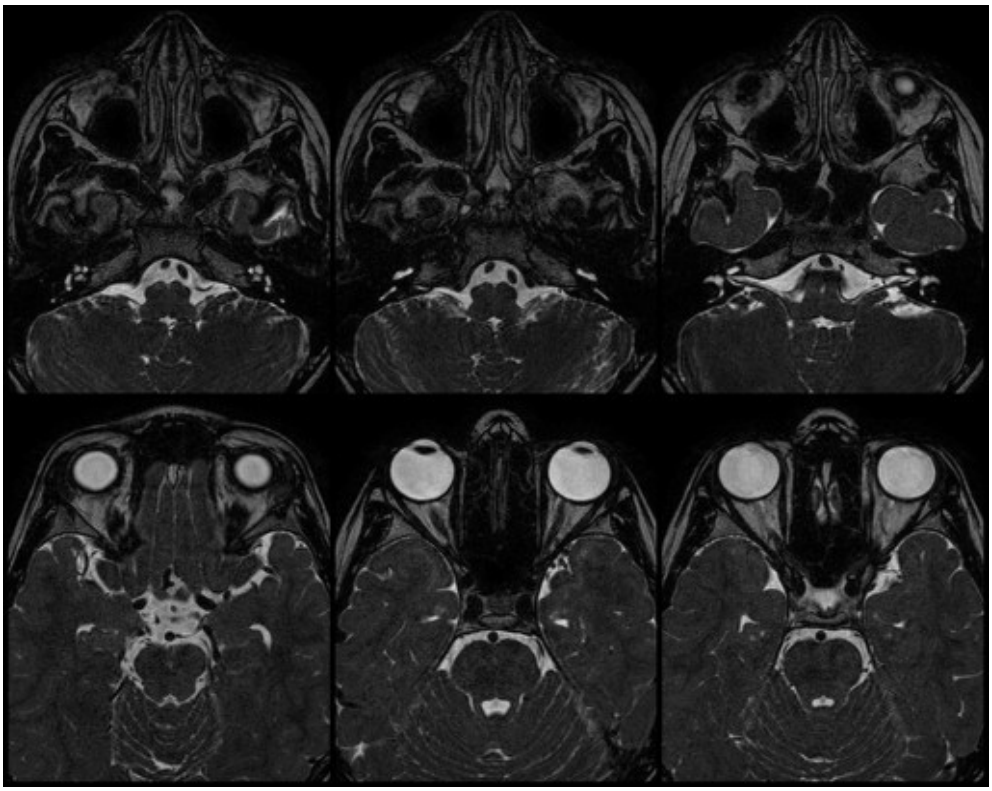

*Fig 7 Axial cuts of cranial fossae and the skull base showing normal anatomy*

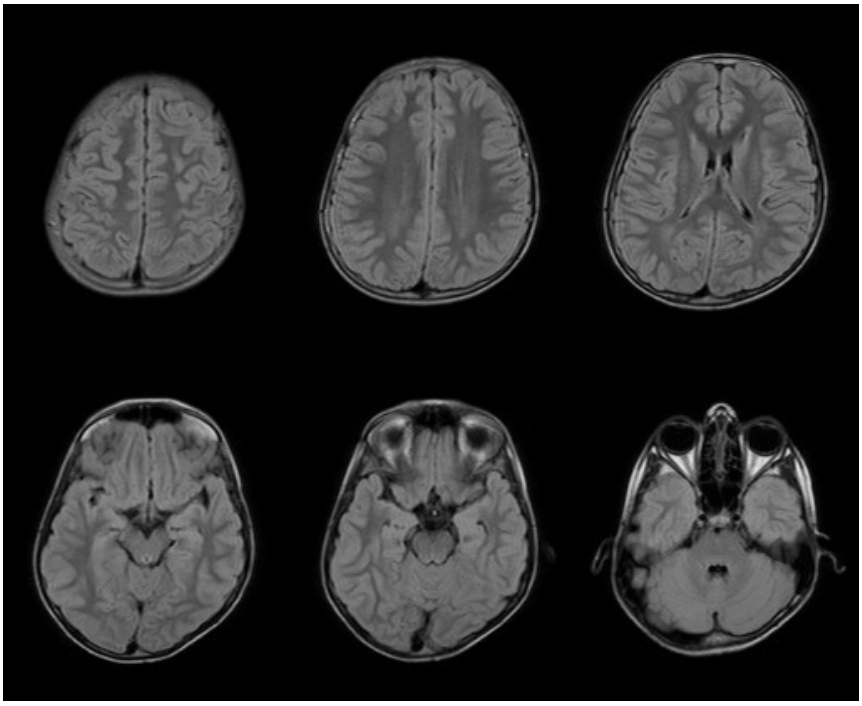

*Fig 8 Axial cuts of brain parenchyma showing normal structures*

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