Polish Journal of Radiology
www.PolRadiol.com

REVIEW ARTICLE

Imaging Spectrum of Cerebellar Pathologies: A Pictorial Essay

Richa Arora[ABCDEF]

Department of Radiology, Nizams Institute of Medical Sciences, Hyderabad, India

Author's address: Richa Arora, Department of Radiology, Nizams Institute of Medical Sciences, Hyderabad, India, e-mail: dr.richaarora@gmail.com

Summary

The cerebellum is a crucial structure of hindbrain which helps in maintaining motor tone, posture, gait and also coordinates skilled voluntary movements including eye movements. Cerebellar abnormalities have different spectrum, presenting symptoms and prognosis as compared to supratentorial structures and brainstem. This article intends to review the various pathological processes involving the cerebellum along with their imaging features on MR, which are must to know for all radiologists, neurologists and neurosurgeons for their prompt diagnosis and management.

Background

The 'Cerebellum' is a latin word which means small brain and it has almost the same number of neurons as in the cerebral cortex, although, weight and volume of the former are approximately one-tenth of the latter. It lies in the posterior fossa posterior to the pons & medulla and consists of two cerebellar hemispheres laterally and a narrow vermis medially. Three pairs of dense fibre bundles (Superior, middle and inferior cerebellar peduncles) connect cerebellum with the brainstem [1].

There are certain pathologies unique to the cerebellum and it can also be involved by some nonspecific diseases that affect other areas of brain as well. These abnormalities are being identified with increasing frequency because of high resolution of infratentorial structures with MR imaging [2]. This article intends to review the various pathological processes involving the cerebellum along with their imaging features.

These are divided into Cerebellar malformations, vascular pathologies, infective & inflammatory conditions, toxic & metabolic processes, demyelinating diseases, neurodegenerative disorders and neoplasms and are elaborated in subsequent sections.

Cerebellar Malformations

Developmental malformations of the cerebellum are diverse and can be secondary to genetic mutations (as in joubert syndrome), due to mechanical compression (like in chiari 2 malformation due to small posterior fossa) or pre,peri & postnatal insults. They may or may not be associated with involvement of other structures like pons, corpus callosum and cerebral hemispheres [1]. The important ones are discussed below.

Dandy-Walker continuum

It is a sporadic disorder which occurs due to chromosomal abnormalities or single gene disorder or teratogen exposure resulting in developmental arrest of hindbrain formation. It consists of a group of anomalies in which there is cerebellar vermis hypoplasia and cystic dilatation of the fourth ventricle, which can be well appreciated on MR. Dandy-Walker malformation(which forms the most severe end of the spectrum) can be differentiated from the Dandy-Walker variant (Figure 1) by the presence of enlarged posterior fossa, elevation of tentorium and torcula herophilli, absent vermis (in contrast to hypoplastic vermis in Dandy-walker variant). The cerebellar hemispheres can be hypoplastic as well. Obstructive hydrocephalus is present in 75% of the cases due to associated aqueduct stenosis [2].
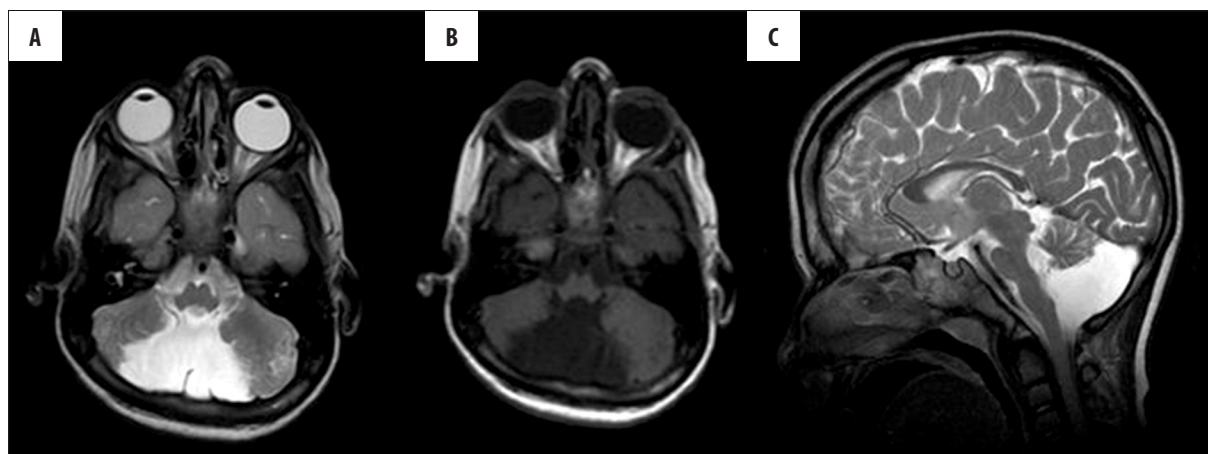
Figure 1. 5-year old male patient with Dandy-Walker variant. Axial T2 weighted (**A**), T1 weighted (**B**) and sagittal T2 weighted (**C**) MR images showing widely separated & hypoplastic cerebellar hemispheres (**A, B**) and hypoplastic vermis (**C**).
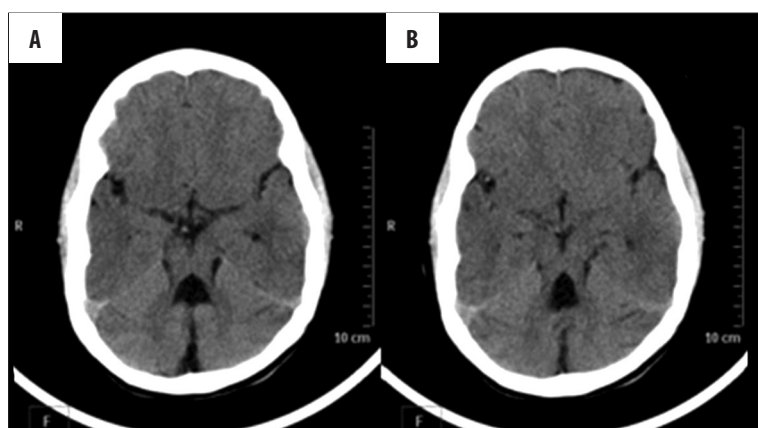

Figure 2. 6-year old boy with joubert syndrome. Axial non-contrast CT images of the brain showing dilated batwing-shaped fourth ventricle (**A**), hypoplastic vermis (**A, B**) in addition to thickened and elongated superior cerebellar peduncles with deep interpeduncular fossa (**A, B**) giving molar-tooth appearance.

The various associations are: nervous system malformations(neuronal migrational anomalies and agenesis of corpus callosum) and extracranial anomalies(cardiac anomalies, skeletal dysplasias). Early diagnosis and management with ventriculoperitoneal or cystoperitoneal shunting is essential for improved survival in these patients [1].

Mega cistern magna and arachnoid cyst constitute the major differentials on imaging. Mega cisterna magna can be differentiated by the presence of normal cerebellar vermis and hemispheres. Arachnoid cyst mimicks mega cisterna magna radiologically, but it causes mass effect on cerebellum, fourth ventricle and sometimes scalloping of inner cortex of occipital bone. However, the exact differentiation between the two is possible with ventriculogram or cisternogram to show the communication of mega cisterna magna with the perimedullary subarachnoid space [3]. Blake's pouch cyst is another entity which mimics Dandy-walker continuum on imaging and is characterized by ballooning of superior medullary velum into the cisterna magna, however, it shows normal vermis and cistern.

Cerebellar hypoplasia

It was previously categorized as chiari4 malformation, but this term is obsolete now. It refers to generalized hypoplasia of cerebellar hemispheres and vermis (with no associated cyst or enlargement of the posterior fossa). It may be unilateral or bilateral. The etiological factors include exposure to drugs such as phenytoin, infection with cytomegalovirus, ionizing radiation and genetic defects(trisomies 21,18 and 13). These patients often present with cognitive impairment in memory, behavior, language, social functioning and often autism [1,2,4].

Joubert syndrome

It is a rare autosomal recessive disorder associated with varying degrees of vermian dysplasia and lack of decussation of fibres in the superior cerebellar peduncles and pyramidal tracts. It is characterized clinically by neonatal breathing dysregulation, developmental delay, hypotonia, ataxia, nystagmus, intellectual disability and abnormal facies [5]. The pathognomonic features on MR imaging include vermian hypoplasia(or sometimes complete agenesis) causing 'batwing' or triangular shape of the fourth ventricle and 'molar tooth sign' (Figure 2), which is produced by the thickened, horizontally oriented superior cerebellar peduncles, thinned ponto-mesencephalic junction and the deep interpeduncular fossa [6,7].

Its associations include brain anomalies (corpus callosum agenesis, grey matter heterotopias, cortical dysplasias & ventriculomegaly) and systemic abnormalities (like congenital hepatic fibrosis, multicystic dysplastic kidney, retinal dystrophy, polydactyly) [5–7].
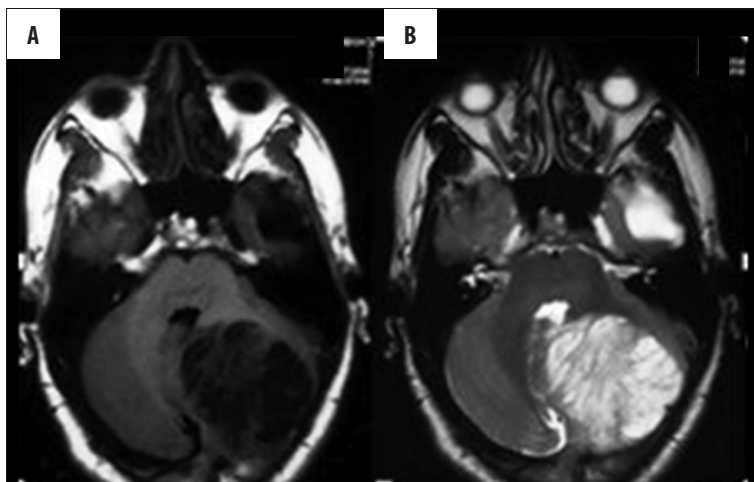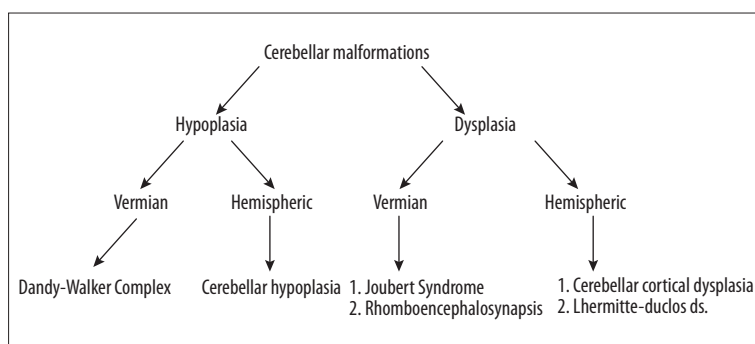
Figure 3. 40-year old man with Lhermitte-duclos disease. Axial T1 (**A**) & T2 weighted (**B**) MR images showing a well defined mass in left cerebellar hemisphere with hypointense signal on T1 and hyperintense signal on T2 weighted sequences along with isointense and hyperintense striations within it. There is evidence of mass effect on the fourth ventricle.

Supplement Figure 1. Approach to diagnose cerebellar malformations.

Rhomboencephalosynapsis

It is a congenital abnormality of the cerebellum with absent or small vermis along with the fusion of cerebellar hemispheres, cerebellar peduncles with or without dentate nuclei [1,2].

Other rare syndromes associated with hypoplastic vermis include Oro-facial-digital syndrome type4, COACH syndrome and Arima syndrome(cerebro-oculo-hepato-renal syndrome) [1].

Cerebellar cortical dysplasia/heterotopias

It is focal area of disorganized architecture in cerebellar hemispheres. Although its pathogenesis is still unclear, it is seen in association with chromosomal trisomies, intrauterine infections and congenital muscular dystrophies. Most common sites of involvement are the nodulus, flocculus and tonsils. The MR imaging features include folial thickening, irregular grey-white interface with defective, large, abnormal fissures, heterotopias and cortical cystic lesions (in severe cases) with no contrast enhancement [8].

Lhermitte-Duclos-Disease

It is also known as dysplastic cerebellar gangliocytoma and presents clinically with macrocephaly and seizures. It results from derangement of normal laminar cellular organization of cerebellum resulting in thickened folia. MR typically shows non-enhancing focal enlargement of cerebellum with prolonged T1 and T2 relaxation times with curvilinear stripes in it (that are isointense to cortex giving striated/tigroid appearance) (Figure 3). It has a strong association with visceral hamartomas [2].

The approach to diagnose cerebellar malformations is well illustrated in Supplement Figure 1.

Chiari malformations

These are common hindbrain malformations associated with the caudal displacement of the cerebellum and the brainstem [1].

Chiari 1 is the commonest out of all and is characterized by herniation of peg like cerebellar tonsils more than 5 mm below the foramen magnum. Syringohydromyelia is a common association. It is often asymptomatic or patients present in early adolescence with ataxia, neck pain and headache [9].

Chiari 2 consists of caudal displacement of cerebellar vermis, medulla and fourth ventricle through the foramen magnum and often associated with spina bifida and lumbosacral myelomeningocele [9,10] (Figure 4). Clinical symptoms depend on the age and range from myelomeningocele, cranial nerve palsies in the neonatal period to features of raised intracranial pressure during childhood and scoliosis in adults.

Chiari 3 is quite uncommon and is characterized by combination of Chiari 2 malformation along with occipital or high cervical encephalocele [9].
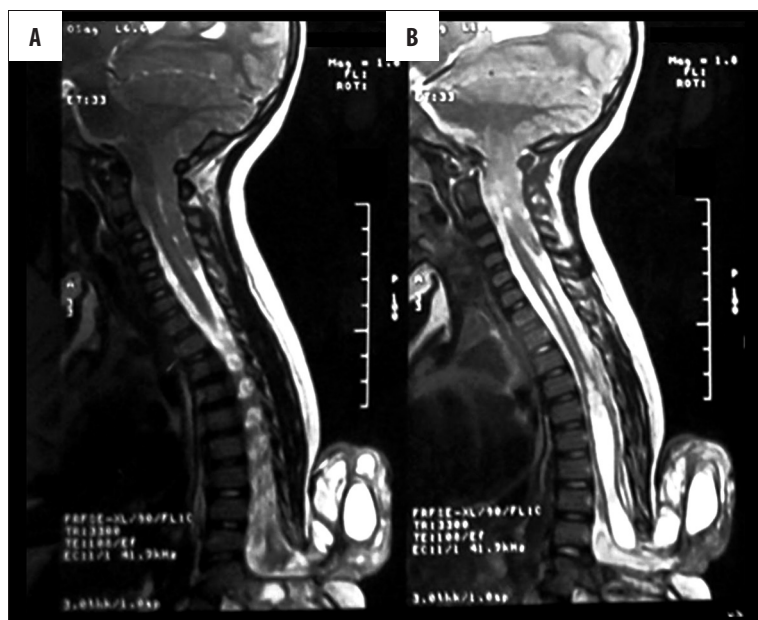
Figure 4. (**A, B**) One and a half year old male child with chiari-2 malformation. Sagittal T2 weighted images of the cervicodorsal spine including craniocervical junction showing evidence of caudal displacement of the cerebellar vermis and medulla with spina bifida and meningomyelocele in the lower dorsal spine. There is presence of syringohydromyelia as well in the dorsal spinal cord.
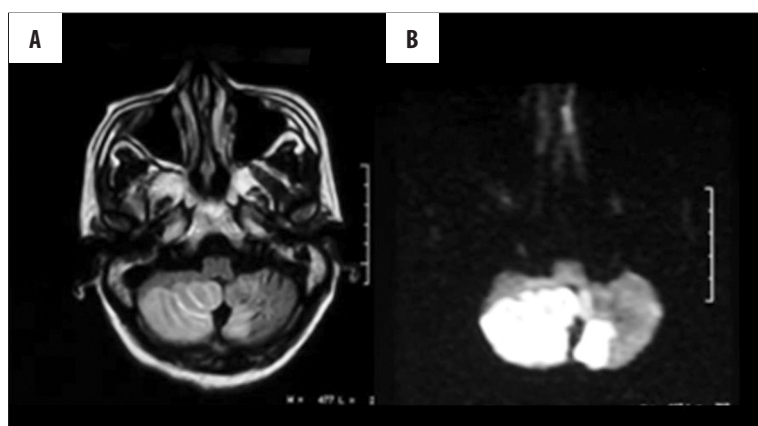

Figure 5. 60-year old man with bilateral PICA infarcts. Axial FLAIR sequence (**A**) showing hyperintensity involving bilateral posterior inferior cerebellar hemispheres corresponding to territory of posterior inferior cerebellar artery which shows acute diffusion restriction on diffusion weighted sequence (**B**).

Chiari 4 is obsolete term now and was previously used for severe cerebellar hypoplasia without its herniation [9].

Vascular Pathologies

These consist of infarcts and hemorrhage.

Cerebellar infarcts

Although cerebellar infarcts are more common than hemorrhage, they constitute only 1.5–2.3% of all strokes. Infarcts may occur in territories of any of the three arteries supplying the cerebellum namely: superior cerebellar artery (SCA), anterior inferior cerebellar artery (AICA), posterior inferior cerebellar artery (PICA). SCA supplies superior surface of cerebellar hemispheres upto great horizontal fissure, superior vermis, dentate nucleus and parts of midbrain. AICA perfuses anteroinferior surface of cerebellum with middle cerebellar peduncle,flocculus and inferolateral pons. Lastly, PICA supplies posteroinferior cerebellar hemispheres, inferior vermis, tonsils and lower medulla. On MR, wedge shaped area of altered signal intensity (hypointense on T1 and hyperintense on T2 & FLAIR) is seen involving both grey and white matter of a particular

arterial distribution along with the evidence of mass effect and diffusion restriction (Figure 5). The mortality rate of infarcts in this region is quite high with figures of about 20-50% and early correct diagnosis is crucial to prevent potentially fatal complications like brainstem compression and obstructive hydrocephalus [11].

Cerebellar hemorrhage

Cerebellar hemorrhages comprise only 10% of all brain hemorrhages. The most common etiological factor is uncontrolled hypertension, other causes being vascular malformations and neoplastic bleed. MR signal intensity characteristics depend on the stage of bleed. They carry a good prognosis if managed timely with evacuation and control of hydrocephalus, which is essentially required in cases of bleeds larger than 3 cm in diameter or with evidence of brainstem compression. Remote cerebellar hemorrhage is a rare entity seen in the clinical setting of prior intracranial surgery or supratentorial craniotomies and has self limiting course. Although its exact pathogenesis is still not clear, nevertheless, male sex, perioperative hypertension and CSF loss, preoperative use of anticoagulants are the risk factors [12].
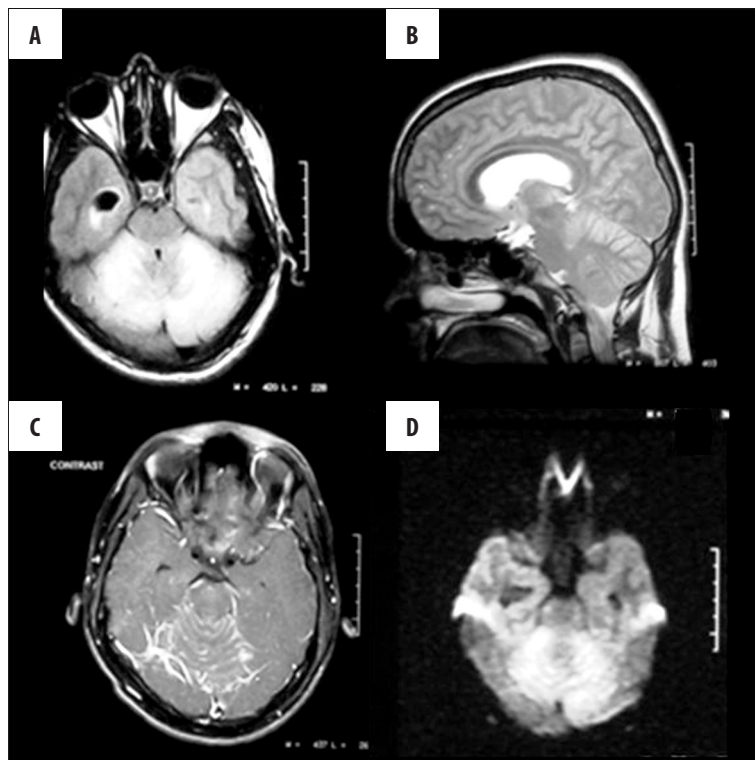
Figure 6. 20-year old male patient with scrub typhus cerebellitis. Axial FLAIR sequence (**A**) of MRI of the brain showing diffuse hyperintensities involving both cerebellar hemispheres with compression of fourth ventricle leading to obstructive hydrocephalus. Parasagittal T2 weighted sequence (**B**) showing hyperintensities involving grey matter of cerebellar hemisphere with compressed fourth ventricle and tonsillar herniation. Axial contrast enhanced T1 weighted MR (**C**) showing diffuse leptomeningeal enhancement in bilateral cerebellum and axial diffusion weighted sequence (**D**) showing restricted diffusion in the same location.

Infective and Inflammatory Conditions

Acute cerebellitis(acute cerebellar ataxia) is a rare inflammatory disorder characterized by isolated inflammation of cerebellum which can be either infectious, post-infectious or post-vaccination. The usual etiological factor being viral agents which include Varicella zoster, Epstein barr, measles, mumps, rubella, herpes simplex and coxsackie viruses. It is more common in children, who present with acute onset of ataxia, nystagmus and dysarthria. Neuroimaging is usually normal and recovery often occurs over weeks, except in severe cases, in which there are features of raised intracranial pressure, hydrocephalus and brain herniation. These cases also show positive findings on MR. The most common findings are diffuse swelling of bilateral cerebellar hemispheres involving both grey and white matter with T2 hyperintensities, mild diffusion restriction and contrast enhancement in the involved cerebellar cortex and leptomeninges (Figure 6). Unilateral involvement of cerebellum is uncommon and involvement of vermis and cerebellar peduncles is variable [13].

Parenchymal granulomas like tuberculomas, neurocysticercosis can also occur in the cerebellum as in other areas of the brain.

Toxic/Metabolic Disorders

The cerebellar abnormalities due to various inborn errors of metabolism can be divided into four groups namely: cerebellar hypoplasia(in adenylsuccinase deficiency), cerebellar atrophy, patchy or diffuse white matter abnormalities and involvement of dentate nuclei and cerebellar cortex [14].

Cerebellar atrophy can be differentiated from cerebellar hypoplasia(discussed earlier) by the presence of enlarged fissures in comparison to normal foliae secondary to loss of tissue in the former, but differentiation can be difficult at times or atrophy can be superimposed on hypoplasia (in congenital disorder of glycosylation). Atrophy can be due to a number of toxic causes like alcoholic cerebellar degeneration, drugs like phenytoin along with genetic neurodegenerative disorders(discussed later) [14,15].

Symmetric involvement of dentate nuclei is seen in metabolic disorders like leigh disease, maple syrup urine disease (also involves cerebellar white matter), wernicke encephalopathy, and toxic causes like metronidazole induced neurotoxicity (Figure 7), methyl bromide intoxication or some organic solvents. It is seen as hyperintensity on T2 weighted and FLAIR sequence with or without diffusion restriction [16]. The cerebellar cortex involvement is a feature of infantile neuroaxonal dystrophy [14,15].

Genetic Neurodegenerative Disorders

The inherited degenerations comprise a complex group of chronic disorders characterized by progressive ataxia and dysarthria. Familiarity with these entities and their imaging features is helpful for the differential diagnosis of ataxias of uncertain clinical type. The important ones are discussed below.

Ataxia telangiectasia

It is a rare autosomal recessive neurocutaneous disorder which consists of progressive cerebellar ataxia (in all cases), oculomucocutaneous telangiectasias and predisposition to recurrent brocho-pulmonary infections and certain
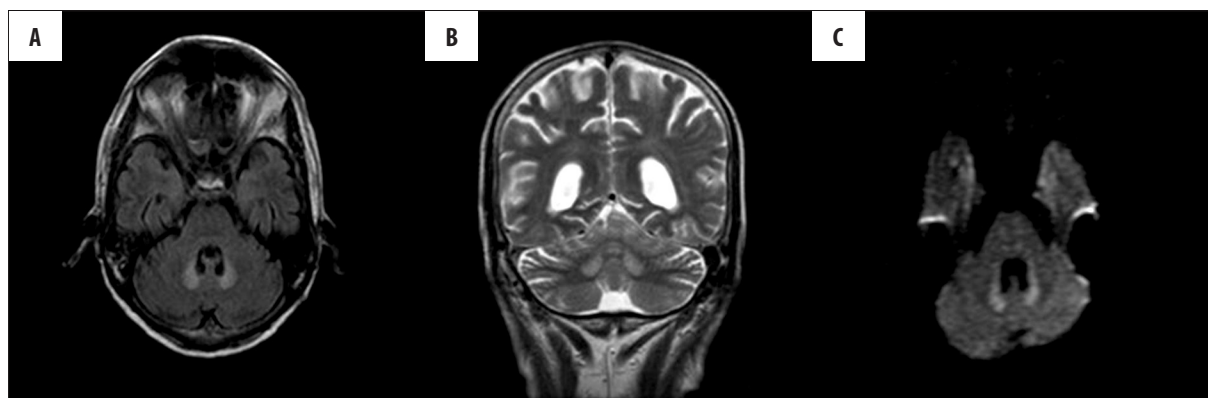
Figure 7. 40-year old man with metronidazole toxicity. Axial FLAIR (**A**) and coronal T2 (**B**) sequence of MRI of the brain showing symmetric hyperintensities involving bilateral dentate nuclei of the cerebellum. Axial diffusion weighted sequence (**C**) showing mild diffusion restriction in bilateral dentate nuclei.
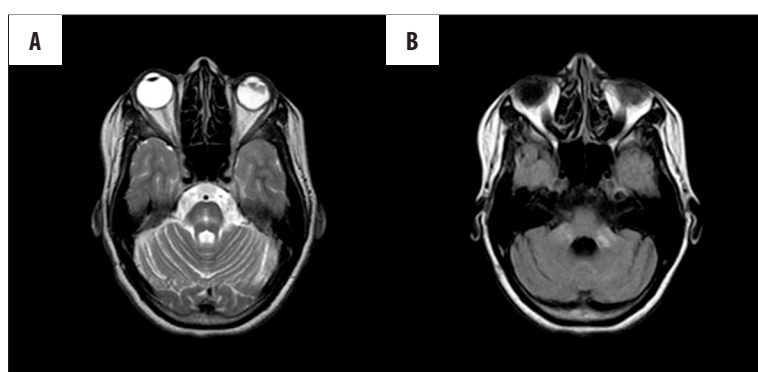

Figure 8. 40-year old female patient with olivopontocerebellar atrophy. Axial T2 weighted image (**A**) of the brain showing diffuse atrophy of the cerebellum and pons with hot cross bun sign in pons. Axial FLAIR sequence (**B**) showing hyperintensity in bilateral middle cerebellar peduncles with pontocerebellar atrophy.

malignancies(bowel & breast cancer, leukemias & lymphomas). It results from a defective gene located on chromosome 11q22-23. MRI shows marked cerebellar atrophy involving both vermis and hemispheres. Additionally, low signal intensity foci can be seen on T2 gradient echo images through out the brain representing capillary telangiectasias. Sometimes hemorrhages can be seen due to rupture of telangiectatic vessels. Wallis et al. reported increased choline signal in the cerebellum in these patients which can be a helpful differentiating feature from other forms of ataxias [17].

Freidreich ataxia

It is the most common inherited progressive ataxia with autosomal recessive inheritance, which results due to repetition of unstable GAA trinucleotide in chromosome 9q. In addition, there is loss of myelinated fibres and gliosis in the posterior and lateral columns of the cervical spinal cord. MRI shows thinning and intramedullary signal changes in the cervical spinal cord. The other significant finding which correlates with the degree of neurological deficits is atrophy of the peridentate cerebellum and superior cerebellar peduncles. Moreover, there is atrophy of the central portion of the medulla and midbrain, dorsal part of upper pons and optic chiasma [18,19].

Olivopontocerebellar atrophy

Olivopontocerebellar atrophy (also known as multiple system atrophy-c type): It is a neurodegenerative disorder resulting from abnormalities of alpha-synuclein metabolism resulting in intracellular deposition both in neurons and oligodendroglia. Presentation is with ataxia and bulbar dysfunction and MR is the imaging modality of choice. On MRI, gross atrophy and T2 hyperintensities are seen in pons, middle cerebellar peduncles and cerebellum with hot cross bun sign in pons (selective involvement of pontocerebellar tracts leads to formation of a cross on T2 weighted axial images through the pons) [20] (Figure 8).

Fahr disease

It is an uncommon familial neurodegenerative disorder which is characterized by abnormal cell loss and calcium deposition in basal ganglia (mainly globus pallidus), internal capsule, dentate nucleus, thalamus, cerebellar and cerebral white matter. The etiology is still not clear [21].

Fragile X-associated ataxia/tremor syndrome

It is a X-linked dominant neuropsychiatric degenerative disorder (with a reduced penetrance) and is the most common cause of inherited mental retardation. Clinically, patients present with cognitive impairment (recent episodic memory problems, difficulties in sustained attention and other executive problems), behavioural problems, ataxia, tremors and rigidity. Imaging features on MR include: symmetric hyperintensities in bilateral middle cerebellar peduncles (MCP sign – most characteristic) and in white matter of cerebellar & cerebral hemispheres on T2 weighted images along with the atrophy of pons, cerebellum and cerebrum [22].
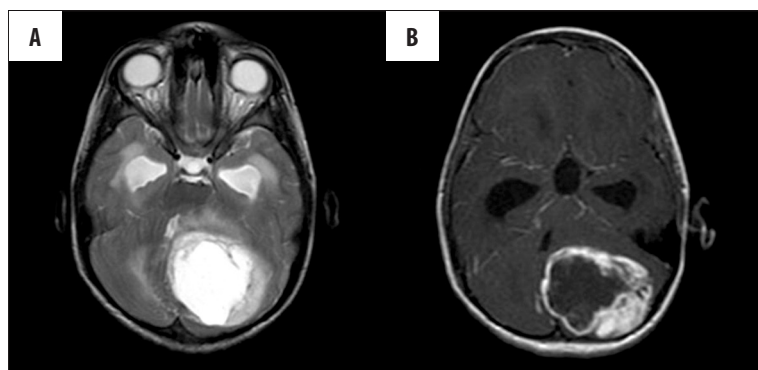
Figure 9. 9-year old boy with pilocytic astrocytoma. Axial T2 (**A**) and postcontrast T1 (**B**) weighted images of the brain showing a large well defined cystic lesion with mural nodule which is mildly hyperintense on T2 weighted image (**A**) seen involving left cerebellar hemisphere and vermis with thick irregular intense enhancement of the wall of the cyst and the mural nodule.
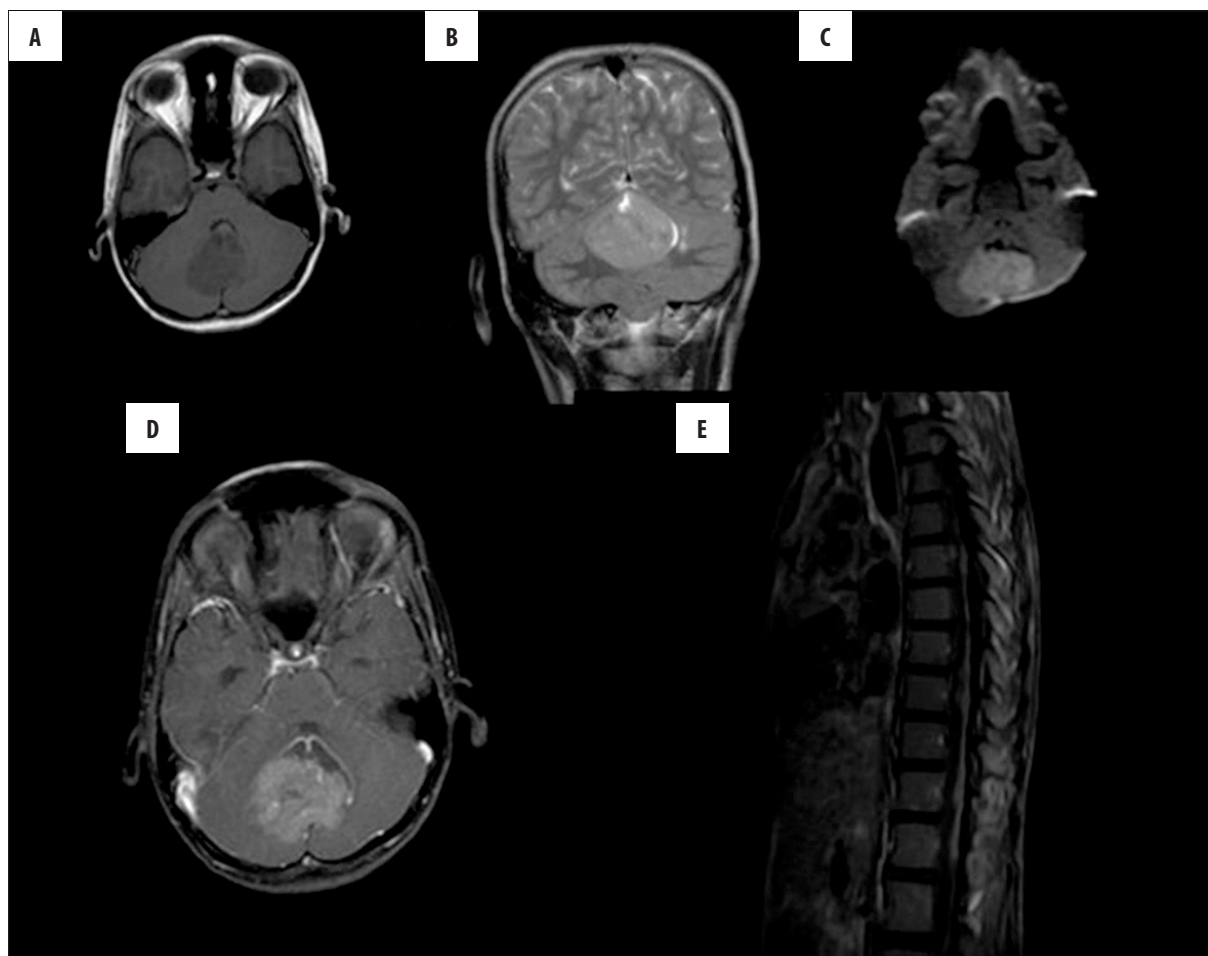

Figure 10. 7-year old boy with medulloblastoma. Axial T1 (**A**) and coronal T2 (**B**) weighted image of the brain showing a well defined homogenous mass with hypointense signal on T1 (**A**) and mildly hyperintense signal on T2 (**B**) weighted sequences in the vermis with mass effect on fourth ventricle causing obstructive hydrocephalus. Axial diffusion weighted sequence (**C**) showing restricted diffusion within the mass. Axial post contrast T1 weighted sequence (**D**) showing moderate enhancement of the lesion with nonenhancing necrotic area anteriorly at the periphery. Sagittal post contrast T1 weighted image (**E**) of the dorsolumbar spine showing diffuse leptomeningeal spread.

Demyelinating Disorders

Various demyelinating diseases (like multiple sclerosis, acute disseminated encephalomyelitis and progressive multifocal encephalopathy etc.) may involve cerebellum. On imaging, they vary from small punctate lesions to large tumefactive lesions with less mass effect as compared to their size. Enhancement is variable and lesions may be non-enhancing or may show open ring type of enhancement along the leading edge of inflammation, which is a characteristic pattern of these disorders. They are not discussed in detail in this article.

Neoplasms

The tumors are described in two groups: childhood tumors and tumors in adults.

Childhood tumors

The commonest cerebellar neoplasm in this age group is pilocytic astrocytoma followed by medulloblastoma. Other less common tumors include atypical teratoid/rhabdoid tumor, teratoma(in infants) and haemangioblastoma(in patients of Von-Hippel-Lindau syndrome [VHL syndrome]).

Juvenile pilocytic astrocytoma

These are WHO grade 1 tumors which tend to occur in first two decades of life. They most commonly arise in the cerebellum (with most tumors involving both the vermis and the hemisphere) and comprise 85% of cerebellar astrocytomas. On imaging, most tumors demonstrate large cystic lesion with an intensely enhancing mural nodule. The wall of the cyst may or may not show enhancement (Figure 9). Less commonly, they are seen as mixed solid, cystic tumors and completely solid tumors are extremely rare. Solid component is hypointense on T1 and hyperintense on T2 weighted sequences. Surrounding vasogenic edema is extremely uncommon. Neurofibromatosis-1 is commonly associated with this tumor and the presence of eosinophillic rosenthal fibres is a pathognomonic feature on histopathology. The prognosis is good and complete surgical resection is usually curative. Recurrence and disseminated disease are extremely uncommon [23].

Medulloblastoma

They are WHO grade 4,extremely cellular, small round blue cell tumors and are also known as CNS Primitive neuroectodermal tumors. The median age of occurrence is 9 years and most of them occur in males with a M: F ratio of 2–4:1. Their most common location is cerebellum with three-fourths arising in the vermis. Obstructive hydrocephalus is common due to protrusion into the fourth ventricle from its roof. They may also involve brainstem. As they are densely cellular tumors, these are hyperdense on CT, isointense on T1 and T2 weighted sequences with restricted diffusion on diffusion weighted sequence. Cyst formation and necrosis is common and is seen in 40–50% and calcification is seen in 10–20% of cases. It is important to acquire contrast enhanced MR of the entire neuraxis to screen for drop metastasis and leptomeningeal spread, as CSF seeding is common in these patients [24,25] (Figure 10).

Atypical teratoid/rhabdoid tumor

They are uncommon tumors that mostly occur in cerebellum in children less than 2 years of age. They are extremely aggressive, WHO grade 4 tumors with high potential for dissemination. The presence of sheets of rhabdoid cells distinguishes these tumors from medulloblastomas or primitive neuroectodermal tumors. MRI shows these lesions as iso to slightly hyperintense on T1 and hyperintense on T2 weighted sequences with areas of necrosis, cyst formation,calcification and hemorrhage with heterogenous enhancement on postcontrast scan. Leptomeningeal spread is common like medulloblastomas, which further worsens the prognosis [26].

Tumors in adults

Metastases comprise the commonest cerebellar neoplasms with most common malignancies to metastasise being lung and breast cancer. Other less common primaries include melanoma, thyroid cancer and renal cell carcinoma. They are sharply demarcated lesions and usually peritumoral edema is extensive and disproportionate to the size of the tumor. Almost 50% of them are multiple and hemorrhage can be a feature in mets from all the above mentioned primaries. Most of them are isointense on T1 and hyperintense on T2 weighted and FLAIR sequences, though, hemorrhage may produce varied signal in them. These features may help, but the complete differentiation from primary brain tumors may be difficult. MR spectroscopy shows almost depleted n-acetyl aspartate (NAA) in addition to elevated choline, in contrast to primary tumors (especially glioblastoma) where NAA is decreased less profoundly [27].

Haemangioblastoma

Haemangioblastomas are tumors of vascular origin that can occur both in the central nervous system and extraneural sites like kidneys, liver and pancreas. They come under WHO grade 1 tumors. Sporadic cases constitute 75–80% of all tumors and peak age group involved in these patients is between 30–60 years. The remaining cases occur in patients of VHL syndrome in which onset is earlier and lesions are usually multiple. Apart from features of cerebellar dysfunction and obstructive hydrocephalus, 5–40% of them have polycythemia due to erythropoietin production. Their most common location is cerebellar hemisphere (85% of cases) followed by vermis (10%). On MR imaging, almost 60% of these tumors are seen as well defined cystic lesions (with non-enhancing wall) with a vividly enhancing mural nodule. Mural nodule is hypointense on T1 weighted and hyperintense on T2 weighted sequence and has prominent serpentine flow voids. Increased age, absence of calcification and non-enhancing wall are the features that differentiate them from pilocytic astrocytomas. The rest 40% of these tumors are solid with or without cystic spaces within it. Preoperative embolisation has a role in the large lesions and surgery is curative in most cases [28].

Very rarely, medulloblastomas and astrocytomas occur in adults (less than 1% of all tumors).

Conclusions

Cerebellum is a vital structure of the brain. Its dysfunction could lead to incoordination of the movements, dysarthria, dysmetria, hypotonia, disequilibrium & vertigo, delays in initiation and termination of the movements and nonmotor deficits(defects in higher functions like verbal and nonverbal intelligence, memory etc.). Certain cerebellar pathologies can have mass effect and cause compression of fourth ventricle and brainstem and thus can be mortal. It is therefore important to be aware of imaging features of cerebellar abnormalities to facilitate their earlier diagnosis.

References:

1. Michael SS, Bernard LM: Brainstem and cerebellar malformations. In: Maria BLIII (ed.), Current management in child neurology. Hamilton, Ontario: BC Decker Inc,, 2005: 479–85

2. Patel S, Barkovich AJ: Analysis and classification of cerebellar malformations. Am J Neuroradiol, 2002; 23: 1074–87

3. Kollias SS, Ball WS, Prenger EC: Cystic malformations of the posterior fossa: Differential diagnosis clarified through Embryologic Analysis. Radiographics, 1993; 13: 1211–31

4. Brancati F, Dallapiccola B, Valente EM: Joubert Syndrome and related disorders. Orphanet J Rare Dis, 2010; 5: 20

5. Ross ME, Swanson K, Dobyns WB. Lisencephaly with cerebellar hypoplasia: a heterogenous group of cortical malformations. Neuropediatrics, 2001; 32: 256–63

6. Sarikaya B, Akpinar E, Karli-Oguz K, Cil B: Joubert Syndrome: Magnetic resonance imaging findings. Neuroanatomy, 2004; 3: 30–31

7. Poretti A, Huisman TAGM, Scheer I, Boltshauser E: Joubert Syndrome and related disorders: Spectrum of Neuroimaging findings in 75 patients. Am J Neuroradiol, 2011; 32: 1459–63

8. Chou TH, Huang PY, Kuo TL: Magnetic resonance imaging diagnosis of focal cerebellar cortical dysplasia. J Radiol Sci, 2011; 36: 187–90

9. Pearce JM: Arnold chiari, or "CruveilhierclelandChiari" malformation. J Neurol Neurosurg Psychiatr, 2000; 68(1): 13

10. Geerdink N, Vliet TV, Mullaart R: Essential features of chiari 2 malformation in MR imaging: an interobserver reliability study – part 1. Child Nerv Syst, 2012; 28(7): 977–85

11. Koh MG, Phan TG, Atkinson JL et al: Neuroimaging in deteriorating patients with cerebellar infarcts and mass effect. Stroke, 2000; 31(9): 2062–67

12. Amini A, Osborn AG, McCall TD, Couldwell WT: Remote cerebellar hemorrhage. Am J Neuroradiol, 2006; 27: 387–90

13. Cheung KH, Fung HT: An unusual case of dizziness presenting to emergency department: acute cerebelllitis. Hong Kong Journal of Emergency Medicine, 2012; 19: 133–37

14. Steinlin M, Blaser S, Boltshauser E: Cerebellar involvement in metabolic disorders: a pattern-recognition approach. Neuroradiology, 1998; 40(6): 347–54

15. Poretti A, Wolf NI, Boltshauser E: Differential diagnosis of cerebellar atrophy in childhood. Eur J Pediatr Neurol, 2008; 12: 155–67

16. Lekhra OP, Jain S, Maheshwari A, Rathore Y: Reversible hyperintense dentate lesions. Journal of Evolution of Medical and Dentate Sciences, 2013; 2(16): 2763–66

17. Wallis LI, Griffiths PD, RitchieSJ et al: Proton spectroscopy and imaging at 3T in ataxia telangiectasia. Am J Neuroradiol, 2007; 28: 79–83

18. Pagani E, Ginestroni A, Nave RD et al: Assessment of brain white matter fiber bundle atrophy in patients with friedreich ataxia. Radiology, 2010; 255(3): 882–89

19. Mascalchi M, Salvi F, Piacentini S, Bartolozzi C: Friedreich ataxia: MR findings involving the cervical portion of the spinal cord. Am J Roentgenol, 1994; 163(1): 187–91

20. Gulati A, Virmani V, Singh P, Khandelwal N: The hot cross bun sign. Neurology India, 2009; 57(1): 104–5

21. Malik R, Pandya VK, Naik D: FAHR disease: A rare neurodegenerative disorder. Indian J Radiol Imaging, 2004; 14: 383–84

22. Brunberg JA, Jacquemont S, Hagerman RJ et al: Fragile X Premutation Carriers: Characteristic MR Imaging Findings of Adult Male Patients with Progressive Cerebellar and Cognitive Dysfunction. Am J Neuroradiol, 2002; 23: 1757–66

23. Koeller KK, Rushing EJ: Pilocytic astrocytoma: Radiologic-Pathologic correlation. Radiographics, 2004; 24(6): 1693–708

24. Fonte MVM, Amaral RPG, Costa MOR et al: Medulloblastoma: correlation among findings of conventional magnetic resonance imaging, diffusion-weighted imaging and proton MR spectroscopy. Radiologia Brasileira, 2008; 41(6): 373–78

25. Porreti A, Meoded A, Huisman TAGM: Neuroimaging of pediatric posterior fossa tumors including review of the literature. J Magn Reson Imaging, 2012; 35: 32–47

26. Meyers SP, Khademian ZP, Biegel JA et al: Primary intracranial atypical teratoid/rhabdoid tumors of infancy and childhood: MRI features and patient outcomes. Am J Neuroradiol, 2006; 27: 962–71

27. Ghods AJ, Munoz L, Byrne R: Surgical treatment of cerebellar metastases. Surg Neurol Int, 2011; 2: 159

28. Bishop FS, Lui JK, Chin SS, Fults DW: Recurrent cerebellar hemangioblastoma with enhancing tumor in the cyst wall: case report. Neurosurgery, 2008; 62(6): 1378–79