

Gross and neurosurgical anatomy of the cerebellar tonsil

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Abstract

Introduction

The primary purpose of this review was to focus on the gross anatomy, including morphometric characteristics and arterial vasculature, of the human cerebellar tonsil. We also aimed to investigate the neurosurgical anatomy of this clinically important structure of the human brain.

Discussion

The cerebellar tonsil is a rounded lobule on the undersurface of each cerebellar hemisphere, continuous medially with the uvula of the cerebellar vermis and superiorly with the flocculonodular lobe. Arterial branches entering the tonsil originate not only from the posterior but from the anterior inferior cerebellar artery as well, often anastomosing with those from the posterior. The main clinical conditions able to affect the position of the cerebellar tonsil include raised intracranial pressure, myelomeningocele, Chiari malformations, posterior fossa hypoplasia and Idiopathic scoliosis. The main neurosurgical approaches related to the region of the cerebellar tonsil include the transcerebellomedullary fissure approach, the tonsilloveal fissure approach (telovelar approach), the lateral approach to the fourth ventricle, the supra- and subtonsillar approaches, the median and far-lateral suboccipital approaches.

Conclusion

The position of the cerebellar tonsil can be affected in some congenital and acquired disorders including Chiari malformations and raised intracranial pressure. The high incidence of tonsillar arterial supply of single origin, partially explained by vascular

anomalies such as arterial hypoplasticity, makes this structure potentially vulnerable to vascular accidents (mainly infarcts) of the inferior cerebellar arteries. A thorough understanding of the regional tonsillar anatomy, as well as its relations with the branches of the posterior inferior cerebellar artery is of paramount significance for neurosurgical interventions in this area.

Introduction

The human cerebellar tonsil is famous for its herniation through the foramen magnum either in congenital or in acquired conditions. Synonyms of the 'cerebellar tonsil' include 'tonsilla cerebella', 'ventral paraflocculus', 'cerebellar amygdala (= almond-shaped)' and 'amygdala cerebelli'^{1,2}. The tonsil belongs to the posterior cerebellar lobe¹. The uvula, the lower half of the diamond-shaped formation of the vermian surface, projects downward between the tonsils, thus mimicking the situation in the oropharynx³.

The primary purpose of this review was to focus on the gross anatomy, including morphometric characteristics and arterial vasculature, of the human cerebellar tonsil. It was also aimed to investigate the neurosurgical anatomy of this clinically important structure of the human brain.

The existing literature regarding the gross and clinical anatomy of the human cerebellar tonsil was reviewed with emphasis on its neurosurgical anatomy. Functional and imaging anatomy data are also presented. Furthermore, neuroanatomical comments on the tonsillar morphology and vasculature are provided.

Discussion

The author has referenced one of its own studies in this review. This referenced study has been conducted in accordance with the Declaration of Helsinki (1964) and the protocols of these studies have been approved by the relevant ethics committees related to the institution in which they were performed.

Gross anatomy

Morphology, locations and relations

The cerebellar tonsil is a rounded lobule on the undersurface of each cerebellar hemisphere, continuous medially with the uvula of the cerebellar vermis and superiorly with the flocculonodular lobe². The rostromedial margin of the tonsils borders the tapering edges of the uvula³. This ovoid structure is attached to the cerebellum along its superolateral border by a white matter bundle called the tonsillar peduncle. The remaining tonsillar surfaces are free surfaces³.

The inferior (caudal) pole and posterior surface of the cerebellar tonsil face the cisterna magna. The lateral surface of each tonsil is covered by, but is separated from the biventral lobule by a narrow cleft³ (the tonsillobiventral³ or postpyramidal fissure¹), except superiorly at the level of the tonsillar peduncle³. The medial, anterior, and superior surfaces all face other neural structures, but are separated from them by narrow fissures.

The anterior surface of each tonsil faces and is separated from the posterior surface of the medulla by the cerebellomedullary fissure. The medial surfaces of the tonsils face each other across a narrow cleft, the vallecula cerebelli, which leads into the fourth ventricle. The superior (rostral) pole is separated from the surrounding structures by a posterior extension of the cerebellomedullary fissure, called

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the telovelotonsillar cleft. The superior extension of this cleft over the superior pole of the tonsil is called the supratonsillar cleft³. The ventral aspect of the superior pole of each tonsil faces the three structures (tela choroidea, inferior medullary velum and nodule) forming the lower half of the roof of the fourth ventricle. The posterior aspect of the superior pole faces the uvula medially and the biventral lobule laterally³.

The tonsil is the most prominent structure blocking access to the caudal part of the fourth ventricle³. The medial segments of the choroid plexus of the fourth ventricle stretch from the level of the nodule anterior to the tonsils to the level of the foramen of Magendie. Each medial segment is subdivided into a rostral or nodular part and a caudal or tonsillar part. The tonsillar parts are anterior to the tonsils and extend inferiorly through the foramen of Magendie³.

The tonsillobiventral fissure is, to the author's anatomical experience, deeper and richer in vessels than the other nearby located sulci. The maximum tonsillar dimension (24 mm) reaches almost the half of the cerebellar hemisphere's height (50 mm). Tonsillar folia can be either major or minor and the latter can be occasionally incompletely formed. The mean number of tonsillar sulci is 24 (17-32) and respective number of major folia is 23 (16-31).

Vasculature

The posterior inferior cerebellar artery (PICA), arising from the vertebral artery, is divided into five segments. The last three are related to the cerebellar tonsil: the tonsillomedullary segment which courses around the caudal half of the cerebellar tonsil; the telovelotonsillar segment which courses in the telovelotonsillar cleft; and the cortical segment which is distributed to the cerebellar surface⁴. The PICA provides perforating, choroidal and cortical branches. The cortical branches are divided into vermian, tonsillar and

hemispheric groups. PICA branches supplying the cortical surface of the tonsil are usually emerging from its lateral trunk (when present)⁴. The left PICA usually courses around the lower pole of the tonsil while the right PICA descends well below the tonsil to the level of the foramen magnum before ascending along the medial tonsillar surface. The PICAs ascend between the tonsils and medulla to reach the interval between the tonsil and uvula and to supply the suboccipital surface³.

The cortical PICA segment begins where the trunks and branches leave the groove between the vermis medially and the tonsil and the hemisphere laterally, and includes the terminal cortical branches. The bifurcation of the PICA, into a medial and lateral trunk, often occurs near the origin of this segment. The cortical branches radiate outward from the superior and lateral borders of the tonsil to the remainder of the vermis and hemisphere⁵. The lateral trunk divides into a larger hemispheric trunk that gives off multiple branches to the hemisphere and smaller tonsillar branches that supply the posterior and inferior surfaces of the tonsil. This division of the lateral trunk into tonsillar and hemispheric branches may occur at various sites in relation to the tonsil, but is most commonly located near the posterior margin of the medial surface of the tonsil⁵.

The tonsillar PICA branches most commonly supply the medial, posterior, inferior and part of the anterior surfaces of the tonsil. If there are no branches directed predominately to the tonsil, the tonsil is supplied by the adjacent hemispheric and vermian branches. There is a reciprocal relationship with frequent overlap in the areas supplied by the tonsillar, hemispheric and vermian branches⁵. The most constant cortical areas supplied by the PICA include the anterior aspect of the tonsil. The smallest area supplied by the PICA is confined to the inferior

part of the ipsilateral cerebellar tonsil⁵. The anterior inferior cerebellar artery (AICA), usually originating from the basilar artery as a single vessel, bifurcates into a rostral and a caudal trunk⁵. The caudal trunk often enters the lateral portion of the cerebellomedullary fissure just below the lateral recess before turning laterally to supply the inferior part of the petrosal surface. The distal branches of the caudal trunk often anastomose with the PICA. The AICA gives rise to perforating arteries to the brainstem, choroidal branches to the lateral segment of the choroid plexus and nerve-related arteries⁵.

Arterial branches entering the tonsil originate not only from PICA but from AICA as well, often anastomosing with those from PICA. The number of cortical branches entering the tonsil ranges, to the author's gross anatomical experience, from two to seven regardless of their origin. The PICA supplies averagely 2.4 (0-7) while AICA 2.1 (0-7) tonsillar branches. Although PICA dominates the supply of cortical tonsillar branches, AICA seems to contribute substantially (46.7%) in this supply. Interestingly, in half of specimens the cortical tonsillar branches seem to have a single origin (either PICA or AICA). Vascular anomalies (such as arterial hypoplasticity) could only partially explain this phenomenon.

Functional data

The cerebellum plays an important role in coordinating movement. It receives sensory information and influences descending motor pathways to produce fine, smooth and coordinated motion. The cerebellar tonsil belongs to neocerebellum (also called the pontocerebellum) which is the largest portion of the cerebellum and coordinates the movement of the distal portions of the limbs. It receives input from the cerebral cortex and thus helps in the planning of ⁶.

There are data supporting that the cerebellar tonsil and other cerebellar structures, affected in horizontal gaze-evoked nystagmus, are part of a gaze-

holding neural integrator control system. Furthermore, gaze-evoked nystagmus might present a diagnostic sign pointing toward ipsilaterally located lesions of midline and lower cerebellar structures⁷. Recent neuroimaging and neurological data implicate cerebellum in non-motor sensory, cognitive, vegetative and affective functions. The cerebellar tonsil, among other cerebellar structures, was found to be activated by hypercapnia and consequent air hunger⁸.

Imaging data

Five millimetres sagittal (1.5 Tesla) magnetic resonance images (MRIs) of the cerebellar hemispheres display several structures including the tonsils. Surface features of the hemispheres including the deeper fissures and shallower sulci are best delineated on T1-weighted and T2-weighted sequences, which provide greatest contrast between cerebrospinal fluid (CSF) and parenchyma. MRI is useful in identifying, localising and quantifying cerebellar disease in patients with clinical deficits⁹.

The primary white matter tracts innervating several hemispheric (including tonsil) and vermian lobules are shown well on proton-density-weighted and T2-weighted spin-echo images of 5 mm coronal (1.5 Tesla) MRIs, which provide excellent contrast between grey and white matter. MRI in the coronal plane should be especially useful in identifying, localising, and quantifying normal and abnormal morphologic differences between the cerebellar hemispheres¹⁰.

Neurosurgical anatomy

Clinical conditions

The engagement of cerebellar tonsils into the foramen magnum is a well recognised consequence of increased intracranial pressure. Tonsillar herniation is not necessarily a terminal event, except, in cases in which it occurs in posterior fossa infarcts, acute subdural haematomas, or during a lumbar puncture. In such

cases, acute herniation is justifiably considered among the determinant factors of clinical outcome¹¹. Myelomeningocele, a congenital neural tube defect, is associated with tonsillar herniation and a smaller posterior fossa¹².

Any inferior displacement of a tonsil below the basion-opisthion reference line in adolescents should be regarded as abnormal¹³. Tonsillar ectopia is defined as any inferior displacement of the tonsils¹⁴ or such displacement with an extent within 5 mm when it is located below the foramen magnum¹⁵.

Tonsillar ectopia, encompassing slight descent of the cerebellar tonsils and Chiari type I malformation, is observed routinely in older children and adults and is believed to be an acquired form of the Chiari malformations. This entity is different from the other Chiari malformations in that hydrocephalus plays no role in its evolution. More likely it is a disorder of para-axial mesoderm, characterized by posterior fossa hypoplasia and content overcrowding, and not an embryologic anomaly of neuroectoderm¹⁶. Tonsillar ectopia is a potentially remediable anomaly which may first produce symptoms in adult life. In these cases diagnosis depends on radiological contrast studies; in particular it is important to examine the cervical canal in prone and supine positions¹⁷.

The incidence of Chiari type I malformation ranges from less than 1% to 3%. The occipital and exertional headache associated with this malformation can be observed in subjects who have new-onset tonsillar ectopia resulting from repeated lumbar puncture, idiopathic intracranial hypotension, lumboperitoneal shunting or spontaneous development. This new-onset headache can remit with return to normal tonsil positioning. The degree of posterior fossa hypoplasia and decrement of CSF flow velocity are important factors determining the clinical significance of tonsillar ectopia¹⁶.

Cousins and Haughton¹⁸ found 33% greater average total magnitude of tonsillar motion through the cardiac cycle in patients with Chiari type I malformation (0.57 mm) than in controls (0.43 mm). Tonsillar motion was 0.61 mm in patients with syringomyelia and 0.50 mm in those without it (22% difference). Suggested causal mechanisms of tonsillar herniation in patients with Chiari type I and type II malformations include cranial constriction, cranial settling, spinal cord tethering, intracranial hypertension and intraspinal hypotension¹⁹. Additionally, pre-existing structural abnormalities in the posterior fossa may constitute an important factor in the development of tonsillar herniation following supratentorial shunts²⁰.

In idiopathic scoliosis patients the position of the tonsil is significantly lower and also asymptomatic Chiari type I malformation is frequently reported in these patients¹³. Tonsillar ectopia with an extent >2 mm in adolescent idiopathic scoliosis patients should be regarded as abnormal¹⁵. These patients have a higher prevalence of tonsillar ectopia than controls¹⁵. Furthermore, scoliosis could be an important manifestation of subclinical tonsillar herniation¹³.

Posterior fossa tumours frequently present with raised intracranial pressure and may cause tonsillar herniation. Muzumdar and Ventureyra²¹ described an uncommon case of a pilocytic astrocytoma of the vermis in a 13-year-old girl who presented with clinical features of progressively worsening raised intracranial pressure, secondary tonsillar herniation and cervical syringomyelia. The cerebellar tonsils herniated down to the C2 level and there was a centrally located syrinx from C2-T1. The tumour was resected and at follow-up (three months later) MRI demonstrated total resolution of tonsillar herniation and syringomyelia. Further, Fujimoto et al.²² presented a case of dysembryoplastic neuroepithelial tumours (multiple cystic lesions on MRI, grey

multinodular gelatinous lesions in pathological examination) in the cerebellum and brainstem of a 44-year-old woman, including the right tonsil.

Hosomi et al.²³ reported a 66-year-old man with small multiple cerebellar infarcts affecting the tonsil and nodulus (detected by diffusion MRI), who complained of headache, vertigo, vomiting and chest oppression sensation. He could not walk veering to right and spontaneous contralateral horizontal nystagmus was noted. Tsuyuguchi²⁴ also reported a case of cerebellar tonsil infarction diagnosed with MRI. Table 1 summarises the main clinical conditions able to affect the position of the cerebellar tonsil.

Surgery

The tonsillomedullary and telovelotonsillar segments of the PICA are the most important vessels encountered in the transcerebellomedullary fissure approach to the fourth ventricle²⁵. Ucerler et al.²⁵ observed the passing of the tonsillomedullary PICA segment through the cerebellomedullary fissure to be placed superior to the tonsil in 5%, at the level of the upper pole of the tonsil in 17.5%, at the middle of the tonsil in 37.5% and at the level of the lower pole of the tonsil in 37.5% of specimens. A thorough understanding of the relationship of the PICA branches to the cerebellar tonsils is prerequisite for surgery in and around the fourth ventricle²⁵.

The tonsillouveal fissure approach, for tumours of the fourth ventricle, aims to avoid division of the inferior vermis. According to the median inferior suboccipital cerebellomedullary fissure approach (telovelar approach), the trajectory to the fourth ventricle is made through the vallecula and microsurgical opening of the arachnoidal layers allows the separation of the two tonsils and provides access to the tonsillouveal sulcus, which is located between the uvula and nodulus²⁶.

Shigeno et al.²⁷ reported the use of the lateral approach to the fourth

ventricle to remove an incidentally found arteriovenous malformation of the inferior medullary velum. A wide posterior fossa craniotomy was performed to move the cerebellar tonsil laterally with C1 laminectomy. The tela chroidea and inferior medullary velum, the two main sheets of tissue that form the lower half of the roof of the fourth ventricle can be exposed by gently displacing the tonsils laterally without splitting the vermis. Both the cerebellomedullary and tonsillouveal spaces were exposed. Because the lateral cerebellomedullary cistern was also exposed, the moving of the cerebellar tonsil in a lateral direction was easy to do without injuring the cerebellar tissues²⁷.

During an operation on the caudal part of the roof of the fourth ventricle, one should remember that the dentate nuclei are located just rostral to the superior pole of the tonsils, underlying the dentate tubercles in the posterolateral part of the roof, where they are wrapped around the superolateral recesses near the lateral edges of the inferior medullary velum³.

The definitive approach to the foramen of Luschka is subtonsillar, because this foramen is actually the end of the natural cleavage plane between the cerebellar tonsil and medulla²⁸. Jean et al.²⁸ described the operative technique for the subtonsillar approach to the foramen of Luschka region. After the cerebellar tonsil is freed from arachnoid adhesions, it can be retracted rostrorodorsally from the medulla,

to expose the cerebellomedullary fissure. By dissecting in a subtonsillar manner around it, the foramen of Luschka can be reached without traversing any neural tissue²⁸.

Lawton et al.²⁹ demonstrated the utility of the supratonsillar approach, an approach that traverses the tonsillobiventral fissure in a trajectory over the cerebellar tonsil to the inferior cerebellar peduncle, for resecting peduncular cavernous malformations. It uses wide splitting of the tonsillobiventral fissure. It differs from the transvermian and telovelar approaches to the fourth ventricle, with a more superolateral trajectory that leads instead to the inferior cerebellar peduncle. By splitting the tonsillobiventral fissure and mobilizing the tonsil inferomedially, the point of access to the lesion is deepened and transgression of normal cerebellar tissue is minimised²⁹.

Tatagiba et al.³⁰ described the surgical anatomy of the midline subtonsillar approach to the hypoglossal canal. This approach includes dorsal opening of the foramen magnum and elevation of the ipsilateral cerebellar tonsil to expose the hypoglossal nerve and its canal. It permits a straight primary intradural view to the hypoglossal canal with no necessity of condylar resections³⁰.

Reynier et al.³¹ reported a patient with a rare peripheral PICA aneurysm located in the tonsillomedullary segment of the right PICA and formed a caudal or infratonsillar loop in the cisterna magna close to the inferior part of the tonsil. The operation was performed with the patient in the sitting position using a median suboccipital approach through a C1 laminectomy. Interestingly, the authors suggested that aneurysms affecting the telovelotonsillar segment of a PICA, forming cranial or supratonsillar loops, and those involving its cortical segment can, be effectively operated using the same median suboccipital approach³¹.

Table 1: Main clinical conditions able to affect the position of the cerebellar tonsil

1	Raised intracranial pressure (e.g.: tumours, haematomas, infarcts)
2	Myelomeningocele
3	Chiari malformations
4	Posterior fossa hypoplasia
5	Idiopathic scoliosis

Table 2: Main neurosurgical approaches related to the region of the cerebellar tonsil

1	Transcerebellomedullary fissure approach
2	Tonsilloveval fissure approach (telovelar approach)
3	Lateral approach to the fourth ventricle
4	Subtonsillar approach
5	Supratonsillar approach
6	Median suboccipital approach
7	Far-lateral suboccipital approach

Stoniewski et al.³² described a case involving a ruptured intradural aneurysm of the meningeal branch of the occipital artery arising from the external carotid artery and connecting with the caudal loop of the PICA by a dural fistula. The aneurysm was located intracranially below the tonsil, at the site of the connection of the caudal loop of the PICA with an anastomosis of the meningeal branch of the occipital artery, compressing the lateral surface of the medulla at the level of the foramen magnum. They applied a far-lateral suboccipital approach without removing the arch of the C1³².

Finally, in PICA-PICA bypass, the PICA is isolated in its lateral medullary or tonsillar loop and dissected free of arachnoid. The technique of anastomosis is similar to that of the superior temporal artery-middle cerebral artery bypass. If the AICA is the recipient vessel, the lateral branch of the AICA posterior to the VIIIth cranial nerve is isolated and used for the anastomosis³³.

Quiñones-Hinojosa et al.³⁴ reported a 30-year-old man with subarachnoid haemorrhage due to a right vertebral artery dissecting aneurysm. The patient underwent a suboccipital craniotomy, where the PICAs were brought together between the tonsils with a side-to-side anastomosis to allow the contralateral PICA to supply the ipsilateral PICA in a retrograde

fashion³⁴. Table 2 summarises the neurosurgical approaches related to the region of the cerebellar tonsil.

Conclusion

The position of the cerebellar tonsil (Figure 1) can be affected in some congenital and acquired disorders including Chiari malformations and raised intracranial pressure. The high incidence of tonsillar arterial supply of single origin, partially explained by vascular anomalies such as arterial hypoplasticity, makes this structure potentially vulnerable to vascular accidents (mainly infarcts) of the inferior cerebellar arteries. A thorough understanding of the regional tonsillar anatomy, as well as its relations with the PICA branches is of paramount significance for neurosurgical interventions in this area.

Abbreviations list

AICA, anterior inferior cerebellar artery; CSF, cerebrospinal fluid; MRIs, magnetic resonance images; PICA, posterior inferior cerebellar artery.

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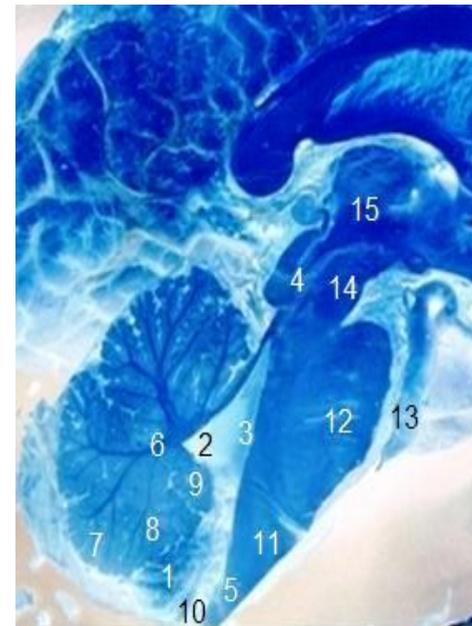


Figure 1: Midsagittal section of brainstem and cerebellum (human brain, left hemisphere). 1: tonsil, 2: roof of the fourth ventricle, 3: floor of the fourth ventricle, 4: cerebral aqueduct (of Sylvius), 5: foramen of Magendie, 6: arbour vitae, 7: pyramid of the vermis, 8: uvula, 9: nodulus, 10: cerebellomedullary fissure, 11: medulla oblongata, 12: pons, 13: basilar artery, 14: midbrain, pyramid, 15: third ventricle (modified from Mavridis³⁵).

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