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Case report

Surgical management of cerebellopontine angle teratomas with total and subtotal approach in adults: A case report

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ABSTRACT

Introduction: Germ cell tumors (GCTs) are neoplasms originating from either gonadal or extragonadal tissues. Primary central nervous system (CNS) GCTs are rare and categorized into germinomas and non-germinomatous germ cell tumors (NGGCTs), the latter including teratomas. Intracranial teratomas represent a small fraction (0.1–1.5 %) of all brain tumors, predominantly occurring in the pineal and sellar regions. Posterior fossa teratomas in adults are exceptionally rare.

Case presentations: This study presents two cases of cerebellopontine angle (CPA) mature teratomas in adult patients. The first case involves a 26-year-old female with severe headaches, diagnosed via MRI with a left CPA mass extending to the C2 level, confirmed as a mature teratoma post-surgical resection. The second case features a 37-year-old male with an incidental finding of a right CPA mass, leading to subtotal resection due to proximity to critical brainstem structures.

Discussion: CNS teratomas constitute 0.5–1 % of intracranial neoplasms.Typical locations include midline structures; however, CPA localization is rare. Diagnosis primarily relies on imaging modalities like MRI, supplemented by histopathological confirmation. Surgical resection remains the mainstay of treatment, with the extent of resection balancing tumor removal and preservation of neurological function.

Conclusion: CPA teratomas are rare but should be considered in the differential diagnosis of CPA masses. Effective management requires advanced imaging for accurate diagnosis and careful surgical planning to balance tumor removal with preservation of neurological function. These cases illustrate successful outcomes with tailored approaches, emphasizing the need for awareness and expertise in handling such uncommon tumors.

1. Introduction

Germ cell tumors (GCTs) are neoplasms, originating in either gonades (both female and male) or could be extragonadal, known as - extragonadal germ cell tumors (EGGCTs). EGGCTs may have various locations in the body, from the brain to the coccyx [1].

Primary central nervous system (CNS) germ cell tumors (GCT) are rare neoplasms that are divided into two groups: germinomas and non-germinomatous germ cell tumors (NGGCTs). NGGCTs include: teratomas, embryonal carcinomas, choriocarcinomas and yolk sac tumors [2].

Teratomas represent a small percentage (0,1–1.5 %) of intracranial tumors. Frequently occupying regions in the brain are pineal gland (51 %) and sellar area (30 %). Age distribution peaks for intracranial teratomas are infancy/neonatal period and between 5 and 14 years. As for posterior fossa teratoma in adulthood, they are extremely rare. Only a few cases have been reported in the literature so far. The clinical presentation of cerebellopontine angle mature teratomas is heterogeneous, with symptoms varying widely based on tumor size, location, and the extent of cranial nerve compression [3].

Here, we present two patients, 26 years old women and 37 years old men diagnosed with cerebellopontine angle mature teratoma. Mature teratomas made up of highly differentiated, adult-type tissues displayed in a variable organized manner. Commonly, all three germ cell layers (endoderm, mesoderm, ectoderm) are represented [4].

Based on fact that teratomas constitute more than 50 % of fetal brain

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tumors, 33 % of neonatal intracranial tumors, 2–4 % of intracranial tumors in people less than 15 years of age, presented cases are atypical not only because of its location, but also for the age of patients [5].

Primary treatment for intracranial teratomas is surgical resection. Total tumor excision is treatment of choice [6] but also can be successfully managed with subtotal resection to reduce the risk of neurological morbidity [7]. Despite successful surgical removal, teratomas can recur due to the potential presence of residual immature or malignant components within the tumor.

The work has been reported in line with the SCARE criteria [8].

2. Case presentation 1

A 26-year-old woman was admitted to the neurosurgery department for a planned operation. The patient complained of severe headache, which started about 2 weeks ago and were progressing in time.

Neurological status on admission: The patient was alert and oriented to person, place, and time. Memory and thought process were intact with normal mood and affect, normal cranial nerve function, intact light touch, pain sensation, vibration sense, and proprioception. Muscle strength 5/5 throughout, no focal weakness. Muscle tone normal, no spasticity or rigidity. Coordination intact, normal finger-to-nose and rapid alternating movements. Gait normal, walks without difficulty. Normal deep tendon and plantar reflexes. It is noteworthy that the patient does not report any family history of brain teratoma or any disease.

According to the findings from the MRI scan of the head and cervical spine, a volumetric formation was identified in the left cerebellopontine angle, extending down to the C2 level of the cervical spine. The posterior fossa mass exhibited hyperintense signal characteristics on T1-weighted images (Fig. 1A, B) heterogeneity with mostly hyperintense signal on T2-weighted images (Fig. 2A, B). Diffusion tensor imaging (DTI) was carried out to detect any hidden spinal abnormalities (Fig. 3). The sagittal FSE T1–weighted MR image with RD map fusion shows a focal increase in RD values at the C1-C2 level (Fig. 4). This finding suggests loss of myelin integrity due to long-standing spinal cord compression. These changes correspond to the early stages of myelopathy. This finding prompted us to expedite surgery, with the goal of complete tumor excision.

Surgery was performed to remove a bulky brain tumor. Surgical procedure: Intracranial lesion extirpation; Anesthesia: General balanced anesthesia; Duration of operation: 6 h; Patient positioning: Prone, with the head securely fixed on the table. A 16 cm incision was made in the occipital region extending to the spinous processes of the C2 vertebra. A left-sided far-lateral craniotomy was performed. The dura mater was opened in a C shape, and the basal cisterns were subsequently opened to relax the brain. Gentle traction of the left cerebellar hemisphere was applied. Intraoperative neuromonitoring (ION) was utilized during the surgical procedure to enhance precision and safety. The tumor was extirpated without compromising brain substance or cranial nerves.

According to the pathological changes detected during the operation, a white bulky tumor is visualized to the left of the medulla oblongata, also occupying the cerebellopontine angle, extending down to the C2 Level, which causes gross deformation of the brain stem and dislocation of the 6th, 7th, 8th, 9th, 10th, 11th cranial nerves. The tumor was embedded in the mentioned cranial nerves. Tumor had a thin capsule. A total resection of the tumor was performed. The case yielded a positive outcome. The treatment decision depends on the possibility of complete resection with minimal damage to healthy tissue. If possible, complete resection is preferred.

As a result of the micromorphological study of the biopsy material, mature multi-layered squamous cell fragments with reticular hyperkeratosis and orthokeratosis are revealed in the fragments of the formation. A cystic structure lined with a multi-layered squamous epithelium, with a granular layer, filled with keratinous masses is also shown. Fragments of mature cartilage tissue with reactive changes of chondrocytes are distinguished. The tissue exhibited glandular structures consistent with gastrointestinal epithelium, including goblet cells (darkly stained nuclei in a columnar arrangement) interspersed with connective tissue. Adjacent to these structures, there were densely packed basophilic cells, possibly indicating immature neural or epithelial components. The overall morphology suggested differentiation toward multiple germ layers, primarily ectodermal and endodermal derivatives. The mentioned histomorphology corresponds to a mature teratoma (Fig. 5A, B).

After the procedure, the patient was transferred to the neurosurgical unit. Cortical and cranial nerve functions were intact. She remained afebrile, and her vital signs were within normal limits. The patient was discharged after 9 days without any complaints. A follow-up MRI after 2 years revealed post-trepanation and postoperative changes, with visualized dorsal craniospinal deformities. The cisterna magna appeared moderately constricted, while no other significant changes were detected. The patient has fully recovered and resumed her normal lifestyle.

3. Case presentation 2

A 37-year-old man was admitted to the neurosurgery department for elective surgery. The patient's general condition is stable. The patient was alert, oriented, and coherent. He reported severe headache, dizziness, and weakness. Cranial nerve function was intact. Normal muscle strength and intact sensation to touch and pain. Coordination was intact.

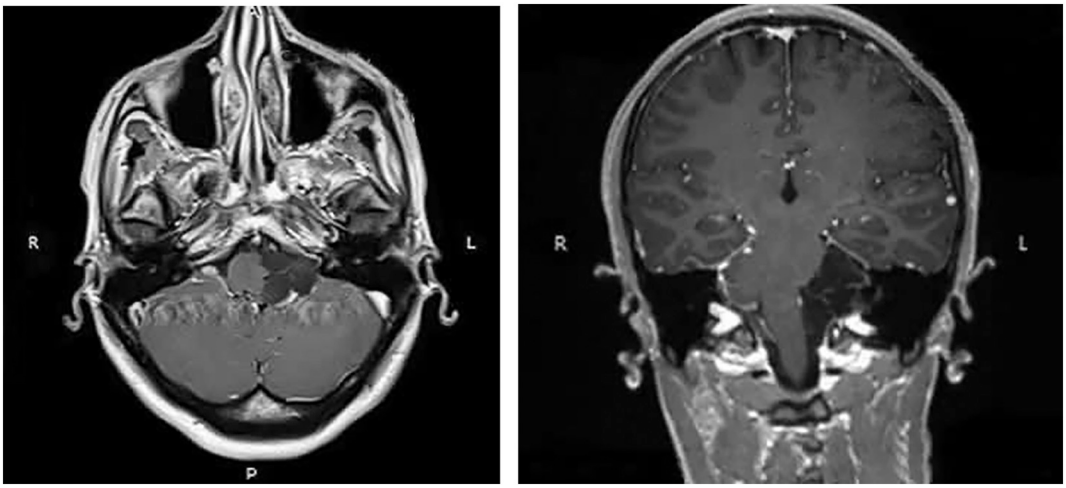


Fig. 1. Left cerebellopontine angle teratoma T1 weighted image-axial (A) and coronal (B) view.

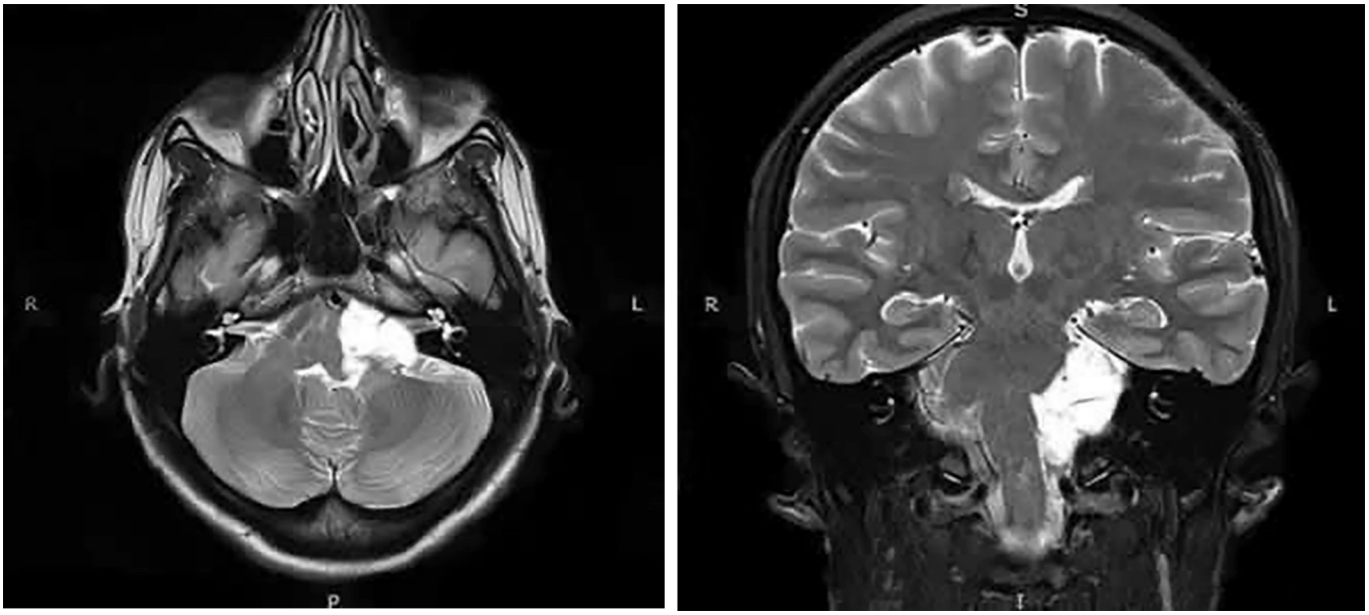


Fig. 2. Left cerebellopontine angle teratoma - T1 weighted image-axial (A) and coronal (B) view.

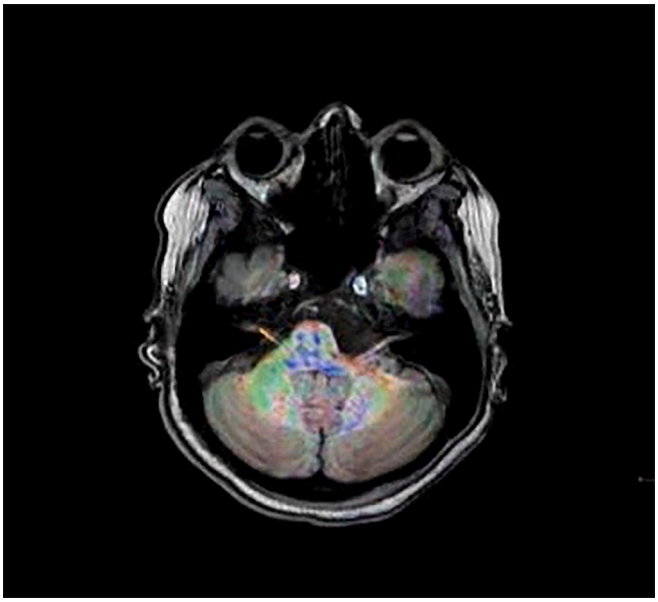


Fig. 3. DTI (Diffusion Tensor Imaging) of mature teratoma – case 1.



Fig. 4. Left cerebellopontine angle teratoma - sagittal view - T1 image.

Deep tendon and plantar reflexes were normal. He reports a family history of hypertension. He does not disclose a family history of cancer or other serious illness.

An incidental finding of a right cerebellopontine angle tumor was revealed, according to MRI (Fig. 6A, B). The tumor caused compression and misalignment of the stem structures. Cerebellopontine angle cistern - pressed. The right cerebellar hemisphere and cerebral crus deviated dorsally, and the brain stem deviated medially. The left lateral ventricle was severely deformed and narrowed.

Surgery was performed to remove a brain tumor. Surgical procedure: Intracranial lesion extirpation; Anesthesia: General balanced anesthesia; Duration of operation: 3.5 h; Patient positioning: Prone, with the head securely fixed on the table. A suboccipital right-sided craniectomy of the occipital bone was performed with a size of 5/5 cm. The dura mater was opened in a Y shape manner, the basal cisterns were opened, and traction of the right cerebellar hemisphere was performed gently. ION was utilized during the procedure. Intraoperatively, a whitish multi-chamber volumetric mass was visualized to the right of the medulla oblongata, including the cerebellopontine angle, causing gross deformation and dislocation of the brainstem along with the 5th, 7th, and 8th cranial nerves. A voluminous tumor was embedded in the mentioned cranial nerves. Since the bulky tumor was located close to the brain stem, approaching the stem during the operation caused the patient's arterial pulse to decrease and the tumor could not be completely excised safely. Subtotal tumor resection was performed. Following the procedure, the patient was transferred to the neurosurgical unit. She remained afebrile but experienced trigeminal neuralgia and paresthesia during the initial postoperative days. However, she was discharged after 5 days without any further complaints.

The histomorphological description was consistent with a mature teratoma. The biopsy highlighted keratinized squamous epithelium with distinct layers, including a stratum corneum, indicative of ectodermal

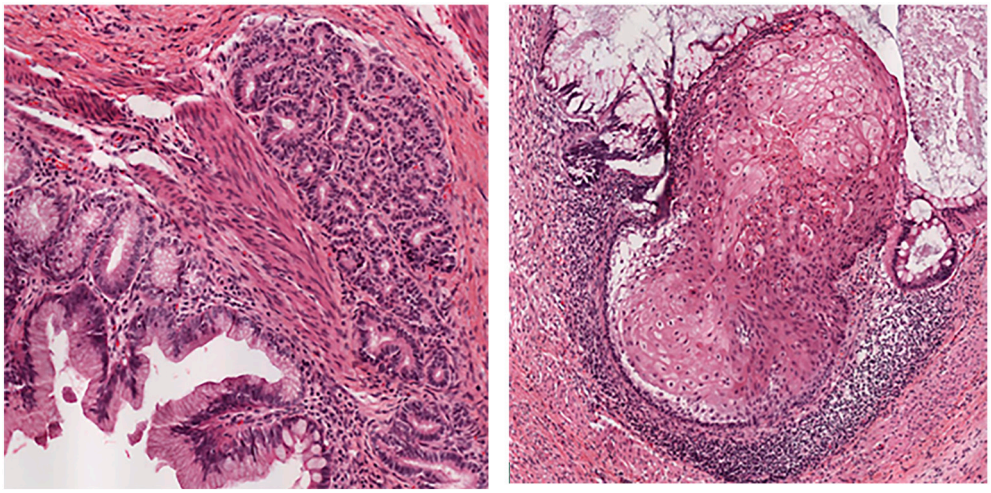


Fig. 5. Histomorphological image of mature teratoma – case 1 – A, B.

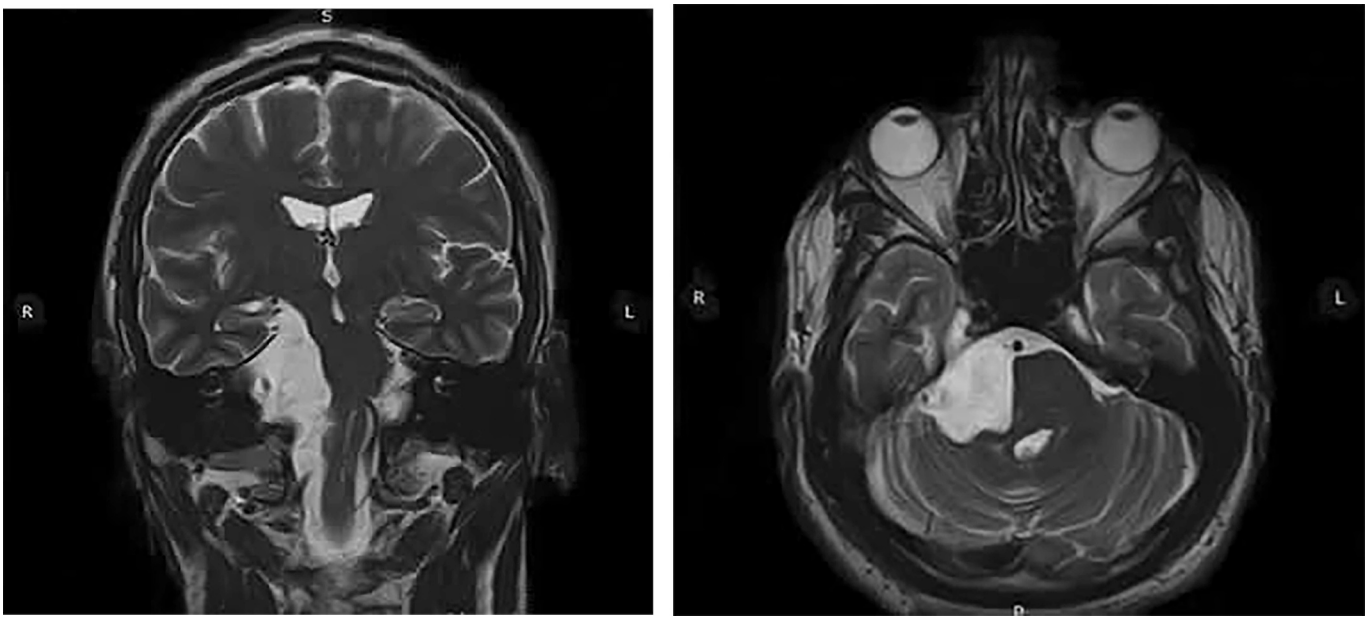


Fig. 6. Right cerebellopontine angle teratoma - T2 weighted image - coronal view - (A) and axial (B) view.

differentiation. Beneath the epithelium, there appeared to be loosely arranged connective tissue and keratin debris. The presence of skin-like structures, including keratinizing epithelium, strongly supported the diagnosis of a mature teratoma (Fig. 7A, B).

The postoperative period was without complications. At the time of discharge, the patient's overall condition was satisfactory.

According to the MRI study performed two months after surgery, compression on the cerebellopontine angle cistern, the right cerebellar hemisphere, and the cerebral crus was decreased. The brainstem was displaced somewhat laterally to the left (Fig. 8A, B). One year later, no negative radiological dynamics were detected in the MRI scan compared to the previous study (Fig. 9A, B). The patient has no neurological deficits and has resumed normal activities.

	Case presentation 1	Case presentation 2
Demographics	26-year-old white woman	37-year-old white man
Clinical presentation	Progressive, severe headache and dizziness for 2 weeks	Stable patient with normal physical examination findings
Imaging findings	Left-sided tumor of the medulla oblongata, continued	Right-sided tumor of the medulla oblongata, continued
		(*continued on next column*)

(*continued*)

	Case presentation 1	Case presentation 2
	on the left side of the cerebellopontine angle, causing dislocation of the CN 6–11	on the right side of the cerebellopontine angle, causing dislocation of the CN 5, 7, 8 and adhered to them
Surgical approach	Total resection	Subtotal resection
Outcomes	Fully recovered, resumed normal lifestyle	Without any deficits, resumed normal lifestyle

4. Discussion

Central nervous system (CNS) teratomas are uncommon neoplasms that represent only 0,5–1 % of intracranial neoplasms. Occurrence is higher in children than in adults (1.5 %-5 %-in adults, up to 15 % in children) and certain countries like Japan, Korea, Taiwan [2,3]. Typically, CNS teratomas originate in midline locations. Common sites are: pineal region, suprasellar cistern, basal ganglia and thalamus. However,

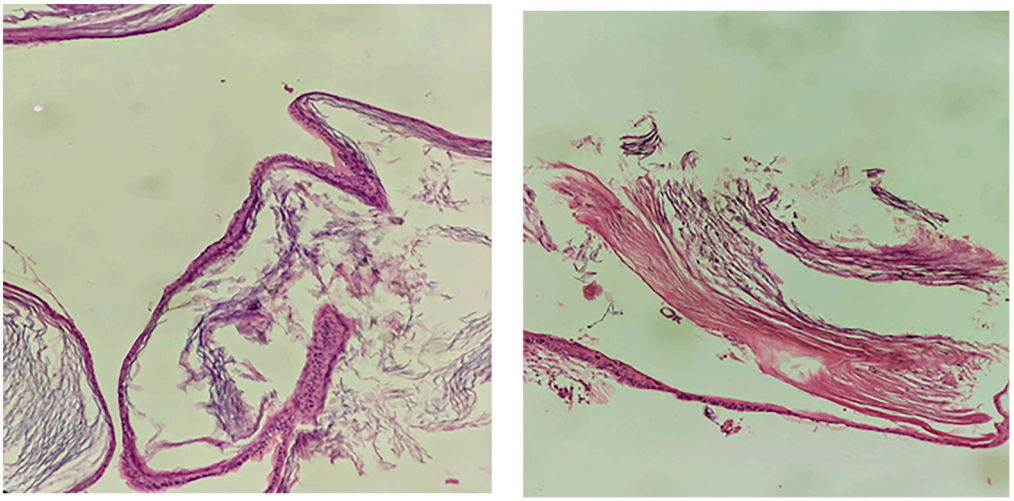


Fig. 7. Histomorphological image of mature teratoma – case 2 – A, B.

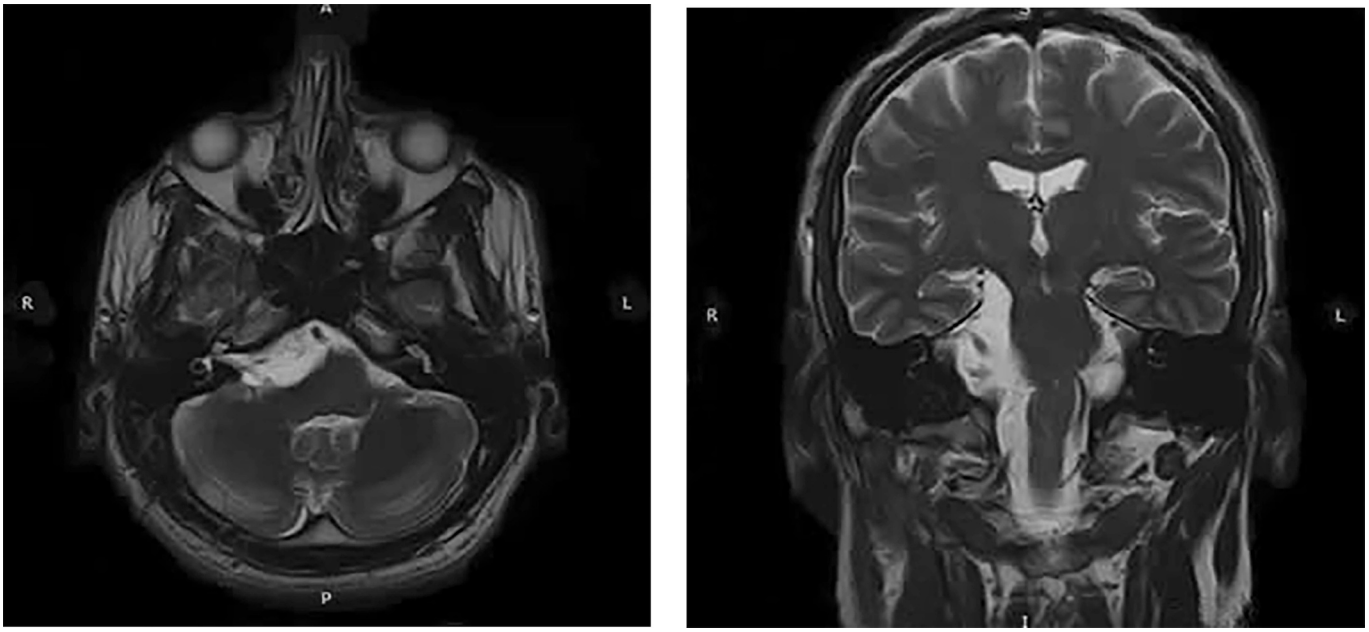


Fig. 8. Post operational right cerebellopontine angle teratoma-T2 weighted image-axial (A) and coronal (B) view.

there are also very rare sites, such as the temporal lobe, cerebellum, lateral ventricles or posterior fossa. Due to their rarity, there are few studies of CNS teratomas in the medical literature [5]. The presented cases of cerebellopontine angle mature teratoma are atypical not only because of its location, but also for the age of the patients.

The clinical presentation of these tumors is highly variable and largely dependent on the tumor's size, location, and the extent of cranial nerve involvement, although headache is consistently reported as the most common initial symptom. Auditory disturbances, including hearing loss and tinnitus, are frequently observed due to compression of the vestibulocochlear nerve. Vestibular dysfunction, manifested as vertigo and dizziness, is also common. Other potential presenting symptoms may include facial weakness or numbness, dysphagia, and visual disturbances [3]. Our literature search identified a case of a 70-year-old female with a cerebellopontine angle mature teratoma who presented with symptoms of headache, vomiting, and gait disturbance [9].

Imaging tests such as X-rays, CT (computed tomography), magnetic resonance imaging (MRI), and ultrasound are used to determine the size and location of a teratoma [10,11]. MRI is considered the radiological study of choice in the diagnosis of brain tumors, although visualization of intracranial mature teratomas is often dependent on their contents and requires postoperative histological examination [10,11]. In our considered cases, it is noted that we used a DTI with a woman. It gives precise information about the involvement and integrity of the white matter tracts in the immediate region surrounding tumors [10]. Also, in our case with a 26-year-old woman, an MRI scan was performed at the first stage, as a result of which a bulky tumor was revealed on the left cerebellopontine region. However, a diagnosis of teratoma was made immediately after a histopathological examination of the material obtained from the biopsy performed during the operation [12].

Mature teratomas of the brain are characterized histopathologically by well-differentiated tissues derived from all three germ layers, with a notable presence of neural tissue. Ectodermal derivatives frequently dominate, including mature neural components such as glial tissue (astrocytes and oligodendrocytes) and neurons [13]. Occasionally, more specialized neural derivatives, such as retinal tissue or ependymal cells, are observed, reflecting differentiation along central nervous system lines [14]. These neural elements are often integrated with other

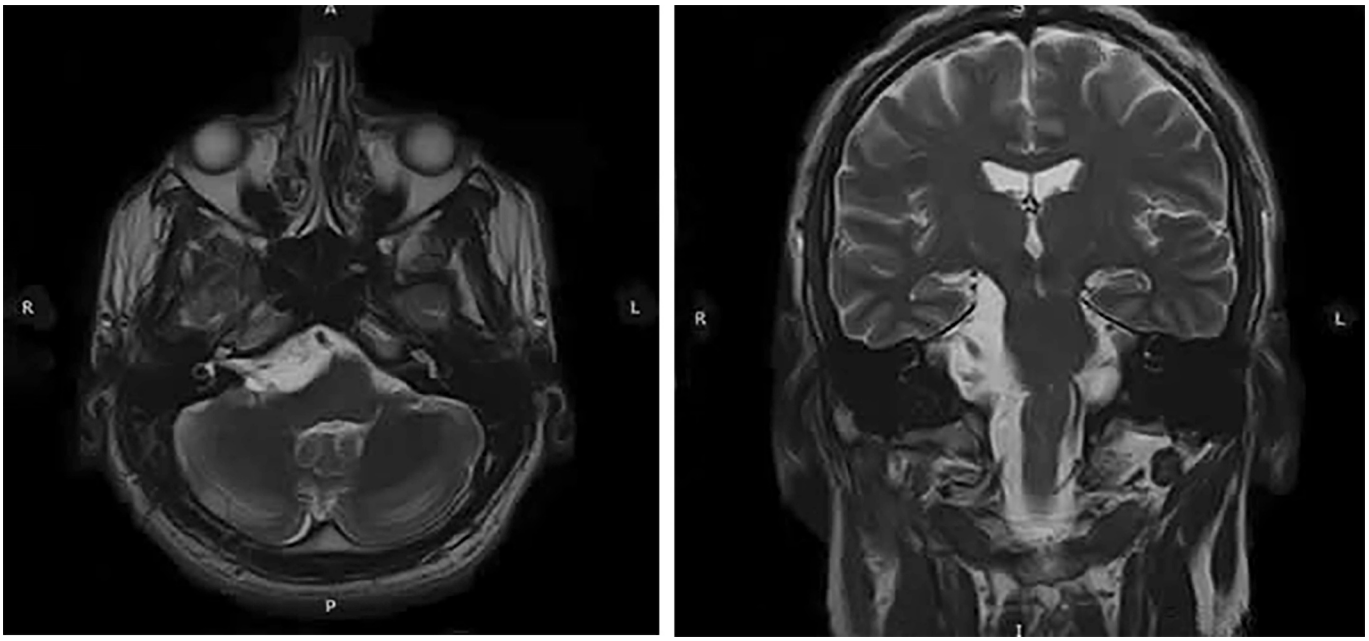


Fig. 9. Post operational right cerebellopontine angle teratoma-T2 weighted image-axial (A) and coronal (B) view.

ectodermal components, such as keratinized squamous epithelium, sebaceous glands, and hair follicles [14]. Mesodermal elements, including connective tissue, vascular structures, and adipose tissue, provide a supporting framework, while endodermal components, such as glandular epithelium resembling respiratory or gastrointestinal mucosa, are less prominent [15]. The neural tissue within brain teratomas is typically mature and lacks evidence of immaturity or malignancy, which is critical for distinguishing these tumors from immature teratomas or malignant transformations. Hematoxylin and Eosin (H&E) staining is the primary method for assessing these tumors, with immunohistochemical techniques used to confirm neural differentiation when necessary [16]. The prominence of neural tissue in brain teratomas underscores their unique histopathological profile and highlights their relationship to the intracranial environment, providing key diagnostic and therapeutic insights.

Intraoperative complications include hemorrhage and iatrogenic brain injury. If we consider the general complications, we can mention neurological dysfunction, such as sensory changes, coordination problems [12]. It should be noted that in the case of the first patient, increased intracranial pressure was manifested by severe headache, nausea, vomiting [12,17]. Obstructive hydrocephalus is also caused by a blockage in the flow of cerebrospinal fluid. Also, pressure fluctuations can complicate the postoperative condition. Intraoperative complications include bleeding, shock. It should be noted that during the operation, mature teratoma may be completely removed, as in the case of a 26-year-old woman, or partially, as in the case of a 37-year-old man. The man's teratoma was not completely removed because it was located close to the brain stem, the patient's pulse rate was decreasing, and only part of it was removed to keep him alive. Brain teratoma is also characterized by convulsions, hormonal imbalance, which is explained by a violation of the pituitary gland [12].

An expected postoperative complication is wound infection. Wound complications include infection, dehiscence, necrosis, and fistulas. The outcome is aseptic-antiseptic measures. Intracranial complications include cerebrospinal fluid (CSF) leak, meningitis, encephalitis, cerebral edema, abscess, pneumocephalus, hematoma, stroke, cranial nerve injury, and seizures and the solution is continuous observation and screening [12]. Also postoperative complication is the neurological deficit. Surgery causes physical changes in brain tissues and can result in diffuse cognitive deficits, including problems with attention, memory, executive functioning, and information processing. For example, patients report feeling that their ability to process information has slowed and that they are more vulnerable to distraction. These symptoms can be addressed through cognitive remediation [12]. Acupuncture is also a potential rehabilitative approach to improving neurological deficits. This addition provides a more comprehensive perspective on post-operative recovery options for cerebellopontine angle teratomas. Insertion of several filiform needles with stimulation at designated sites of nerve distribution induces neural stimulation. This is thought to result in local biochemical and neural stimulation, followed by the release of mediators. Acupuncture may improve neurological deficits associated with conditions such as ptosis and diplopia following surgery [18].

The primary treatment for CPA (cerebellopontine angle) teratomas is surgical intervention. The choice of surgical approach depends on the tumor's size, location, and relation to surrounding structures. Common approaches include: Retrosigmoid (Suboccipital), translabyrinthine and middle fossa Approaches [6,19]. Retrosigmoid (Suboccipital) is the most commonly used route, providing good access to the CPA. The patient is positioned laterally, and a craniotomy is performed behind the ear. This approach allows the surgeon to visualize the CPA contents, including the cranial nerves and blood vessels, facilitating careful dissection and removal of the tumor. Translabyrinthine Approach is chosen when the tumor involves or is very close to the internal auditory canal. It involves removing part of the temporal bone, including the labyrinth, which results in complete hearing loss on the affected side. However, it provides direct access to the CPA with minimal brain retraction. Middle Fossa Approach is used for tumors primarily located in the anterior part of the CPA. It provides good access to the internal auditory canal and the anterior CPA structures, preserving hearing in some cases.

Teratomas differ from other CP corner masses in that mature teratomas are benign tumors, most often consisting of two or three germ cell derivatives. Only in rare cases is the transition to a malignant tumor observed. Mature teratomas can occur at any age, but they are most common in the reproductive years. Whereas other corner CP masses can arise at any age and are malignant tumors [12].

The extent of resection—whether full or incomplete—can significantly influence patient outcomes, potential complications, and the need for additional treatments. Complete removal of the teratoma eliminate the tumor entirely to prevent recurrence and minimize the risk of neurological deficits by carefully preserving critical structures [7,20].

But attempts to achieve complete resection might result in significant functional deficits, if the tumor is closely adherent to vital structures. Incomplete resection can significantly alleviate symptoms caused by compression of nearby structures, but Residual tumor tissue can continue to grow, because of these patients require regular follow-up with imaging studies to monitor for tumor regrowth or progression. Several reported cases showed that 18 patients with posterior fossa teratomas received surgical resection as their primary treatment, and a majority of these studies observed no recurrence following surgery [3].

Mature teratomas are typically benign and do not require additional therapies beyond surgery, in cases where the tumor is incompletely resected or shows signs of aggressive behavior, further treatment might be considered. Options include: Radiotherapy-reserved for cases where there is residual tumor that cannot be safely removed, Chemotherapy-generally not indicated for mature teratomas due to their benign nature, but may be considered in mixed germ cell tumors with malignant components [1,21]. In one case we identified, a 70-year-old female with a cerebellopontine angle mature teratoma underwent successful total resection without the need for postoperative radiotherapy or chemotherapy. The patient experienced an uneventful recovery and remained disease-free at 1-year follow-up [19].

In conclusion, the treatment of mature teratomas in the CPA is complex, requiring a highly coordinated, multidisciplinary approach. Surgical resection remains the cornerstone of treatment, with the primary goal of maximal safe resection. Advances in imaging, surgical techniques, and intraoperative monitoring have significantly improved outcomes for patients with this rare condition. Long-term follow-up with periodic MRI scans is essential to detect any recurrence early. Also, from the patient's perspective, a healthy lifestyle and constant health monitoring are important for prevention. As well as light physical activity and sleep regulation. In both cases, the patients' quality of life gradually recovered after surgery. They were able to relieve their symptoms and return to their life.

5. Conclusion

CNS mature teratomas are rare in adults. We report two cases of brain mature teratomas diagnosed using DTI for preoperative planning. The first case underwent total resection, while the second had subtotal resection. Both cases yielded positive outcomes. Treatment decisions depend on the feasibility of complete resection with minimal healthy tissue damage. If achievable, total resection is preferred; however, if there is a high risk of significant functional impairment, subtotal resection is recommended.

Author contribution

Sopho Nakhutsrishvili (SN): Writing-original draft, Validation, Supervision, Methodology, Data curation, Conceptualization.

Otar Gakhokia (OG): Writing – original draft, Visualization, Data curation, Conceptualization, Supervision.

Mia Metreveli (MM): Writing – review & editing, Validation, Methodology, Data curation, Conceptualization.

Revaz Shalibashvili (RSH): Writing – review & editing, Validation, Methodology, Data curation, Conceptualization.

Anna Basilashvili (AB): Writing – review & editing, Validation, Methodology, Data curation, Conceptualization.

Elene Alphaidze (EA): Writing – review & editing, Validation, Methodology, Data curation, Conceptualization.

Shota Ingorokva was the neurosurgeon who performed the operation.

Alexander Najarov and Otar Gakhokia were the neurosurgeons who performed the clinical and neurological evaluation, also performed the operation.

Consent

Written informed consent was obtained from the patients for publication and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Ethical approval for this study was provided by The Bioethics International Committee of the Petre Shotadze Tbilisi Medical Academy (IRB0310202401/1), on 3 October 2024.

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Guarantor

Dr. Otar Gakhokia.

Research registration number

This case reports are not a first time of reporting, because of this Research Registry Unique identifying number is not necessary.

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Conflict of interest statement

The authors disclose no conflicts of interest.

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