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Inpatient Rehabilitation of Cerebellopontine Angle Epidermoid Cysts: Report of 3 Cases

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Abstract

Objective—Demonstrate the inpatient rehabilitation potential of cerebellopontine angle epidermoid cyst patients. Due to their location, symptoms may present with a complex combination of headache, cerebellar dysfunction, and cranial nerve deficits affecting functional status.

Methods—This report describes the cases of 3 patients with cerebellopontine angle epidermoid cysts who underwent neurosurgical resection followed by inpatient rehabilitation. All 3 patients experienced gait instability and cranial nerve deficits before surgery, and 2 of the patients had mild cognitive deficits. A customized rehabilitation program for these patients can address these deficits.

Results—Each patient showed demonstrable gains in function with inpatient rehabilitation and good outcomes at discharge.

Conclusion—When rehabilitating epidermoid cyst patients, the clinician must be aware of a higher likelihood of cranial neuropathies, need for increased psychosocial support, and the need for more vigilant long term medical monitoring to detect recurrence.

Keywords

Epidermoid Cyst; Cerebellopontine Angle; Rehabilitation

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Introduction

Intracranial epidermoid cysts, also known as primary cholesteatomas, are uncommon brain tumors that account for 0.2-1.8% of all intracranial tumors.¹ Approximately half of these cysts are located at the cerebellopontine angle (CPA)², although they can also be found in the parasellar regions, 4th ventricle, cerebellum, cerebral hemispheres, brainstem, and lateral ventricles³. These tumors are congenital and arise from displaced epithelial tissue between the 3rd and 5th weeks of gestation, when neural tube closure occurs⁴. Despite their early origination, symptoms appear decades later due to the tumor's slow and gradual growth as the epithelial cells accumulate. These brain lesions have an epithelial lining that encapsulates the tumor, which consists of desquamated epidermal cell debris. Most lesions are solid, but because the desquamated cells contain cholesterol some may have a liquid cystic center. The rate of tumor growth is gradual and linear, resembling the rate of human epidermis turnover, which is unlike the exponential growth of neoplastic lesions.⁵

The growth of epidermoid cysts follows the natural planes across the basal surface of the brain, and the cysts can wrap around and adhere to important neurovascular structures.⁶ CPA epidermoid cysts often are found compressing the surrounding cranial nerves (CN). Patients typically present with a combination of ataxia, gait abnormality, and cranial nerve deficits. Because these neurologic symptoms significantly impair function, rehabilitation after complete or partial resection of the tumor is a critical part of the patient's recovery.

Epidermoid cysts account for 3-6% of CPA tumors. Comparatively, acoustic neuromas or schwannomas, the most common CPA angle tumor, account for 85%.⁷ The rehabilitation process for acoustic neuromas focuses on vestibular rehabilitation, facial nerve rehabilitation, and swallowing deficits.⁸⁻¹¹ Balance difficulties can result from the tumor burden as well as from the required surgical resection, and compensation for vestibular dysfunction can take weeks to months. A literature review of patients with acoustic neuromas reports the majority have balance difficulties both pre-operatively and post-operatively, and in turn, quality of life (QoL) can be adversely affected.¹² Due to the similar location of epidermoid cysts in the CPA, one could surmise the same negative effects for these patients, such as balance impairment and reduced QoL, reinforcing the need for early rehabilitation intervention.

Despite this importance of rehabilitation for patients with resected intracranial epidermoid cysts, a recent PubMed literature search revealed no publications regarding rehabilitation for this patient population. This article helps address this gap in information by reporting the benefits of an inpatient rehabilitation program for 3 patients with neurologic effects from epidermoid cysts treated at the same cancer center. It is imperative for physiatrists to have an understanding of the natural course of epidermoid cysts, the potential side effects of the tumor and its treatment, and clinical symptoms to be watchful for in the long-term should recurrence occur in the future.

Patients

Between 1997 and 2011, 3 patients with epidermoid cysts were admitted to the inpatient rehabilitation unit at a tertiary referral-based cancer center.

Case 1

An 18-year-old right-hand-dominant female presented at the cancer center with a several-month history of worsening left-sided headache that was unrelieved with pain medication and exacerbated by moving her head forward or backward. She complained of a mild gait imbalance, ringing in her left ear, and blurry vision in her left eye. Neurologic examination at the time revealed normal sensory, motor, reflex, and cerebellar findings for all 4 extremities. The extraocular muscles were intact and no facial droop was noted. The only abnormal neurologic finding pre-operatively was left V2 and V3 facial numbness. Magnetic resonance imaging revealed a 1.4 cm × 2.7 cm × 2.1 cm left CPA hypointense lesion that was bright on T2 imaging, consistent with an epidermoid cyst, and there was mass effect on the pons and displacement of cranial nerves VII and VIII.

This patient received a left retromastoid craniotomy with tumor resection and results of the pathologic examination were consistent with an epidermoid cyst. A post-operative MRI revealed no evidence of residual tumor. Physical therapy, occupational therapy, and speech therapy followed the patient post-operatively, and the consult physiatrist performed an evaluation on post-operative day 4. The patient was transferred to the inpatient rehabilitation unit on post-operative day 5 to further address gait impairment, cognitive deficits, and cranial nerve impairments.

Post-operatively, this patient demonstrated ataxia, requiring durable medical equipment for safety. Diplopia was present due to a left cranial nerve VI palsy, which also contributed to her gait impairment and her rehabilitation included an eye patch to be used during all functional tasks. A facial droop accompanied her continued facial numbness on the left side of her face. She denied hearing abnormalities but had symptoms of vertigo. Mild cognitive deficits with respect to short-term memory, attention, and problem-solving skills were addressed. There was some concern of dysphagia; however, the findings from a modified barium swallow test on post-operative day 7 were negative for dysphagia and a regular diet with thin liquids was ordered and tolerated. These symptoms were addressed during her 5-day inpatient rehabilitation stay, and she made significant improvement with physical, occupational, and speech therapy, reducing the amount of assistance she required at home once discharged to home with her family. Her functional independence measure (FIM) scores at the time of admittance to inpatient rehabilitation and at the time of discharge are listed in table I. Her brain imaging is presented in Figure I. The patient was followed up in the neurosurgery clinic 3 months post-operatively. By that time she had returned to work and findings from her neurologic examination, which included the cranial nerves, were normal.

Case 2

A 72-year-old right-hand-dominant male had a right epidermoid cyst removed 30 years prior at a different facility after he had complained of right upper extremity ataxia. Post-operatively, the patient was informed that he had received a subtotal resection. His post-operative course had been complicated by aseptic meningitis, continued right upper extremity incoordination, and complete right-sided hearing loss.

He presented 30 years after his original resection with right shoulder droop, dysarthria, and worsening gait with 2 falls reported in 5 months. On pre-operative physical examination, the patient demonstrated significant right upper extremity dysmetria, a pronator drift with a positive Romberg sign, inability to tandem walk, slight tongue deviation to the left, and right cranial nerve VI and VII weakness with facial droop. Cranial nerve V sensation was intact. Magnetic resonance imaging revealed a large right CPA tumor measuring 7 cm in diameter with significant brainstem displacement and hydrocephalus. The mass extended through the foramen magnum and through the foramen of Luschka. The patient underwent right retromastoid craniotomy, C1 laminectomy, right frontal ventriculostomy, and microdissection of tumor from cranial nerves VII, VIII, IX, and X. Pathology results confirmed an epidermoid cyst. CT revealed a large resection cavity on post-operative day 1. Determination of the amount of residual tumor was difficult to determine without MRI which was not performed post-operatively.

Post-operatively the patient had dysphagia requiring Dobhoff tube placement, an unsteady gait, and hoarseness. A flexible nasal pharyngoscopy revealed right vocal cord paresis. Follow-up as an outpatient was recommended for possible vocal cord collagen injection. Audiology evaluated the patient on post-operatively day 15 and found no measurable hearing in his right ear.

Physiatry was consulted on post-operative day 9. The patient was able to walk 10 feet with an intravenous pole and to perform sit-to-stand transfers with minimal assistance. He demonstrated balance difficulty with veering to the right, and this was reported to be a significant factor limiting his independence post-surgically. He was admitted to inpatient rehabilitation on post-operative day 13, where he stayed for 8 days, and received speech, physical, and occupational therapy. His ambulation progressed to 300 feet with a rolling walker at supervision level. His swallow improved, and he was advanced to a soft diet with thin liquids using aspiration precautions. Once his caloric intake was sufficient, the Dobhoff tube was removed and tube feeding stopped. He was discharged from inpatient rehabilitation to home under the care of his wife. His FIM scores are listed in table II. Brain imaging is presented in Figure II.

The patient lived outside of the state and was lost to outpatient follow-up.

Case 3

An 18-year-old right-hand-dominant female had been undergoing boot camp after joining the military. During an obstacle course, she had fallen and sustained a concussion. A computed tomography scan of the head had revealed an incidental finding of a right CPA mass with severe brainstem compression. She was referred to the cancer center for surgical

resection, where a pre-operative physical examination revealed only two neurologic abnormalities: slight tongue deviation to the left and left upper extremity dysmetria. The patient had 5/5 strength in all 4 extremities with intact sensation. A right retromastoid craniotomy was performed with excision of the tumor. Pathology results confirmed an epidermoid cyst. MRI on post-operative day 3 revealed complete resection of the tumor.

After her operation, extubation was difficult. The patient suffered respiratory stridor and required reintubation. Later she was diagnosed with mild to moderate dysphagia with intermittent aspiration and bilateral vocal cord dysfunction. The patient's voice quality continued to improve during her hospitalization. She was placed on a mechanical soft diet and received daily speech therapy.

Post-operatively, her tongue deviation and left upper extremity dysmetria resolved. However, she had evidence of a right cranial nerve IV paresis, right CN VI palsy, and right CN V2 and V3 numbness. She denied any hearing difficulty. Diplopia was an especially frustrating symptom for this patient. Neuro-ophthalmology evaluated the patient on post-operative day 18. Her visual fields were intact despite her impaired right extraocular movements, and no acute interventions were recommended besides taping glasses lens for compensation. The patient was to follow-up with a pediatric ophthalmologist as an outpatient.

On post-operative day 10, the patient was evaluated by a consult physiatrist. She had 5/5 strength, normal sensation in all 4 extremities, and a mild left upper extremity dysmetria. Due to her continued gait instability, diplopia affecting overall functional status, and dysphagia, the patient was transferred to the inpatient rehabilitation service on post-operative day 12. She progressed from walking 350 feet with a rolling walker at minimal assist to walking the same distance with a rolling walker at supervision assist. Although her appetite was poor, she tolerated a mechanical soft diet and was educated on progressing her diet as tolerated. The patient made modest gains and was discharged after 7 days on the inpatient rehabilitation unit. She went home under the care of her parents. Her FIM scores are listed in table III. Her brain imaging is presented in Figure 3.

The patient presented for follow-up in clinic 11 weeks post-discharge. She no longer had facial droop, dysphagia, or headaches. Her voice was more robust although not at her baseline. Her right CN V1 and V2 distribution numbness had completely resolved, and the remaining abnormal sensations only involved a small area just lateral to her mouth. Unfortunately, she continued to have a right cranial nerve VI palsy and diplopia with no apparent improvement, and she continued taping one lens of her glasses for comfort. Physically, she was independent without assistive device needs to perform basic activities of daily living, gait and transfers. She began jogging 2 miles approximately 5 weeks post-discharge. Due to her visual deficits she did not drive. The patient and her mother had not noticed any cognitive deficits but were considering a neuropsychological evaluation for completeness and due to her future plans of attending community college. Because she was from out of state, she was instructed to follow-up with local physicians.

Discussion

DIAGNOSIS AND TREATMENT

CPA epidermoid cysts can compress the surrounding cranial nerves, brainstem, and cerebellum and the brainstem. Ataxia and cranial nerve palsies often result¹. Men and women are equally affected and the symptoms usually arise between the mid-20's and early 50's⁶, with a mean age of 38.8 years at presentation⁴. Patients have a mean duration of 3.1 years from onset of symptoms to diagnosis and surgical intervention¹³. On computed tomography images the lesions are hypodense with attenuation values slightly above zero. Sometimes they have small areas of calcification.¹ Cyst rupture is an uncommon complication that can cause aseptic meningitis.¹⁴

Treatment of CPA epidermoid cysts is purely surgical. Surgery can be challenging because of their location near the brainstem and because they often wrap around cranial nerves and vascular structures. The capsules encompassing these tumors often adhere to neighboring vital areas of brain tissue, increasing the difficulty of resection. Surgeons typically perform a suboccipital craniectomy with intent of complete resection, but subtotal resections are frequently required¹⁵ because a more aggressive, total resection may lead to a worse neurologic picture for the patient.¹³ These are amongst the most difficult to remove CPA tumors. Schwannoma resections are typically easier due to a cleavage plane and less adherence to the nerves and vessels. This typically makes cranial nerve deficits, in particular the lower cranial nerves, amongst the most profound symptoms of epidermoid cyst patients.³ While schwannomas and epidermoid cysts can cause similar symptoms, the neurologic deficits of the epidermoid cyst may result in more cranial nerve abnormalities than schwannomas. Epidermoid cysts can recur from the residual unresected tumor capsule; however, because of the slow rate of growth it may take up to 30-40 years for symptoms to reappear. 94% of schwannomas are totally resected. Recurrence rates are less than 2% for totally resected schwannomas.² In comparison, reports of epidermoid cyst total resection vary from 25-57% of epidermoid cysts.¹⁵ An epidermoid cyst recurrence rate of 35% after subtotal resection and 5% after total resection at 13 years has been reported. The mean period for recurrence was 8.1 years after initial resection.²⁰ Surgical resection is also used to treat recurrence. Neither radiotherapy nor chemotherapy has been found to be effective.^{13,16}

REHABILITATION PROCESS

CPA epidermoid cysts and their surgical treatment can lead to damage of surrounding structures. Multiple cranial nerves and the cerebellum reside in the CPA. Patients can present with cognitive deficits, ataxia, gait instability, vertigo, diplopia, facial numbness, facial weakness, dysarthria, hearing changes, dysphagia, voice hoarseness and gastrointestinal dysfunction. The resulting devastating neurologic deficits can impact a patient's function and quality of life. Because of the complex neurologic presentation of these tumors, a rehabilitation plan should be customized around the known cyst location, the symptoms of the patient and the cranial nerves affected. The customized interdisciplinary rehabilitation plan may require the services of speech therapists, audiologists, physical therapists, occupational therapists, neuropsychologists, physiatrists, otolaryngologists, ophthalmologists and optometrists.

While there are past publications on epidermoid cysts, none have addressed their rehabilitation. There is also a considerable amount of publications on the rehabilitation of other CPA tumors, namely schwannoma patients. However, this is the first publication to describe the rehabilitation process of CPA epidermoid cyst patients. All 3 patients had improved FIM scores while on inpatient rehabilitation. These reports support the use of inpatient rehabilitation in this patient population. Much of the rehabilitation process would be similar to CPA schwannomas. However, there are also some important differences from other CPA tumors that should be noted by the treating clinician.

These tumors engulf cranial nerves and adhere to them making them more difficult to resect neurosurgically than other CPA tumors. Versus other CPA tumors, one would expect cranial nerve deficits to be more prominent and cranial neuropathy symptoms such as diplopia and vertigo to be larger contributors to gait instability. Our patients had cranial nerve palsies ranging from cranial nerve IV through X and XII. Visual and vestibular rehabilitation was an important part of the rehabilitation for our patients and should be emphasized.

Because of the higher rate of recurrence, patients should maintain medical follow-up. Our patients did not follow closely with the treating team at our institution after discharge. This may be in part due to distance. Follow-up and tumor surveillance is important because delayed detection has been reported to occur in half of patient recurrences.²⁰ Patients and local physicians should be educated to look for clinical signs of recurrence which could happen at any time often decades after initial diagnosis. Serial MRI imaging is also recommended.³

Psychosocial support is also important for these patients. Like other CPA tumors, patients have to deal with permanent neurologic symptoms that can affect employment, function, and quality of life. However, due to the higher rate of recurrence of these tumors years and sometimes decades later, they also have uncertainty that their existing symptoms may worsen or new ones can occur. It has been reported that oncology health care professionals often underestimate the psychological distress of cancer patients and therefore underutilize mental health services.¹³ Referral to appropriate support services for these patients is recommended.

Conclusion

This is the initial publication describing the responses of patients with epidermoid cysts to inpatient rehabilitation. Patients may require a customized and coordinated rehabilitation plan. The epidermoid cyst rehabilitation process differs from other CPA tumors due to the higher likelihood of cranial neuropathies, need for increased psychosocial support, and the need for more vigilant long term medical monitoring to detect recurrence. Future studies with larger sample sizes are needed.

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Left: Pre-Operative T2 MRI Imaging; Right: Post-Operative T2 MRI Imaging

Figure 1.
Brain Imaging of Case 1

Left: Pre-Operative T2 MRI Imaging; Right: Post-Operative CT Imaging

Figure 2.
Brain Imaging of Case 2

Left: Pre-Operative T2 MRI Imaging; Right: Post-Operative T2 MRI Imaging

Figure 3.
Brain Imaging of Case 3

Table I

Functional Independence Measure (FIM) Scores

	At admission	At discharge
Survey item		
Eating	5	6
Grooming	4	6
Bathing	4	6
UE dressing	5	6
LE dressing	5	6
Toileting	5	6
Bladder management	5	6
Bowel management	5	6
Transfer: bed, w/c, chair	4	6
Transfer: toilet	5	6
Transfer: tub or shower	3	5
Locomotion: walker, w/c	4	5
Locomotion: stairs	4	5
Comprehension	7	7
Expression	7	7
Social interaction	7	7
Problem solving	5	5
Memory	5	5
Total FIM score	89	106
FIM gain		17
FIM efficiency		3.4

UE Upper Extremity, *LE* Lower Extremity, *w/c* Wheelchair

Table II

Functional Independence Measure (FIM) Scores for Case 2

	Admit	Discharge
Eating	7	7
Grooming	5	5
Bathing	4	5
UE Dressing	4	5
LE Dressing	5	5
Toileting	3	5
Bladder Management	3	3
Bowel Management	3	3
Transfer: Bed, w/c, chair	4	4
Transfer: Toilet	4	4
Transfer: Tub or Shower	4	4
Locomotion: Walker/WC	5	5
Locomotion: Stairs	1	1
Comprehension	6	6
Expression	6	6
Social Interaction	6	6
Problem Solving	5	6
Memory	5	6
Total FIM Score	80	86
FIM gain		6
FIM efficiency		0.75

UE Upper Extremity, *LE* Lower Extremity, *w/c* Wheelchair

Table III

Functional Independence Measure (FIM) Scores for Case 3

	Admit	Discharge
Eating	6	7
Grooming	5	6
Bathing	4	5
UE Dressing	5	6
LE Dressing	5	6
Toileting	6	6
Bladder Management	6	6
Bowel Management	5	5
Transfer: Bed, w/c, chair	5	5
Transfer: Toilet	5	6
Transfer: Tub or Shower	5	5
Locomotion: Walker/WC	4	5
Locomotion: Stairs	4	4
Comprehension	7	7
Expression	7	7
Social Interaction	7	7
Problem Solving	7	7
Memory	7	7
Total FIM Score	100	107
Change in FIM Score		7
FIM efficiency		1.0

UE Upper Extremity, *LE* Lower Extremity, *w/c* Wheelchair