# Diagnostic and Surgical Implications of Ventral Vertebrobasilar Displacement by Posterior Fossa Neurenteric Cysts

*Timothy Uschold, David S. Xu, David A. Wilson, Adib A. Abla, Peter Nakaji, Robert F. Spetzler, Steve W. Chang*

**Key words**
- Cerebellopontine angle
- MRI
- Neurenteric cyst
- Posterior fossa tumor

**Abbreviations and Acronyms**
**BA**: Basilar artery
**CPA**: Cerebellopontine angle
**NEC**: Neurenteric cyst


*Division of Neurological Surgery, Barrow Neurological Institute, St. Joseph's Hospital and Medical Center, Phoenix, Arizona, USA*

*To whom correspondence should be addressed: Steve W. Chang, M.D. [E-mail: Neuropub@dignityhealth.org]*

*Citation: World Neurosurg. (2014) 82, 3/4:480-484. http://dx.doi.org/10.1016/j.wneu.2013.10.037*

*Journal homepage: www.WORLDNEUROSURGERY.org*

*Available online: www.sciencedirect.com*



OBJECTIVE: **Neurenteric cysts (NECs) are uncommonly encountered lesions of the central nervous system with heterogeneous imaging characteristics. The object of this study was to review the preoperative imaging findings represented among a cohort of surgically treated posterior fossa NECs. These findings are considered in the context of surgical technique, and inform an understanding of aberrant neuroembryological development associated with NECs.**

METHODS: **A single-institution, multisurgeon series of 7 consecutive patients (5 female and 2 male patients, mean age 36 years, range 19 to 57 years) treated surgically for histopathologically confirmed posterior fossa NECs was retrospectively reviewed. Lesion imaging and anatomic characteristics were noted on preoperative magnetic resonance imaging (MRI). Imaging comparisons were made against an additional cohort of 266 consecutive surgically treated posterior fossa masses to validate unique anatomic findings.**

RESULTS: **T1 and T2 MRI signal characteristics were variable when compared across lesions. All NECs were found to be anteriorly located within the posterior fossa, but always situated between the brainstem pial surface and the vertebrobasilar system, causing ventral displacement of vertebrobasilar vessels.**

CONCLUSIONS: **Posterior fossa NECs display variable patterns of MRI signal and are commonly considered as part of a broad differential of cystic posterior fossa masses. We identified tumor insinuation between the ventral brainstem and vertebrobasilar system as a highly sensitive and specific radiographic sign for NECs. This finding was not observed among a large cohort of posterior fossa masses representative of other multiple pathologies.**

## INTRODUCTION

Since Puusepp (10) first described a cervical neurenteric cyst (NEC) as an "intestinome" in 1934, multiple nomenclatures have attempted to describe these lesions. Historically known also as gastrocytomas, enterogenous, bronchogenic, gastroenterogenous, enteric, respiratory, and archenteric cysts, NECs are recognized as benign, malformative lesions of endodermal origin (9, 11). During the third week of development, primitive endodermal rests may insinuate in proximity to the developing neuro-ectoderm through incomplete obliteration of the neurenteric canal, notochordal splitting and adhesion, failed notochordal escalation, or aberrant cellular migration (7). The precise embryological mechanisms accounting for the full spectrum of NECs (spinal, infratentorial, and supratentorial) remains controversial, and may ultimately be multifactorial or location-dependent.

In concordance with the embryological theories presented previously, neurenteric cysts typically arise in relation to the embryologic remnants of the notochord from posterior clinoid to sacrum. They are most commonly reported as intradural extramedullary lesions ventral to the spinal cord, and are followed in frequency by posterior fossa locations (5). Within the posterior fossa, NECs are typically located anteriorly along the clivus, at the cervicomedullary junction, and in the cerebellopontine angle (CPA). These lesions display heterogeneous MRI imaging features typically characterized as diffusion negative and T1/T2 variable with rare enhancement (2). As a result, NECs often are considered broadly among other cystic mass lesions. In this study, we present a surgical case series and identify vertebrobasilar artery displacement from the brainstem surface as a novel pathognomonic sign for NECs. This finding is reviewed in relation to current embryological mechanisms, and also informs our discussion of surgical technique.

## METHODS

Prior to the start of this study, approval by the Institutional Review Board at St. Joseph's Hospital and Medical Center was obtained per established standards. The medical records of all patients treated by the senior authors (P.N., R.F.S., S.W.C.) for posterior fossa neurenteric cysts at Barrow Neurological Institute (Phoenix, Arizona) between 1998 and 2011 were retrospectively reviewed. Cases without confirmed histopathological diagnosis of NEC, those located below the level of the foramen magnum, and those managed nonoperatively were excluded from analysis. To further compare and validate

**Table 1.** Clinical and Imaging Characteristics

| Case | Age/Gender | Presenting Symptoms | Location | Magnetic Resonance Imaging Appearance | | | | Vascular Displacement |
|---|---|---|---|---|---|---|---|---|
| | | | | T1 | T2 | DWI | Contrast | |
| 1 | 23/F | Headache, nausea/vomiting | Anterior pontomedullary junction | + | – | – | – | Basilar, bilateral vertebrals |
| 2 | 35/F | Headache | Anterior pontomedullary junction | + | – | – | Scattered | Basilar, bilateral vertebrals |
| 3 | 49/M | Headache | Anterior pontomedullary junction | – | + | – | – | Basilar, bilateral vertebrals |
| 4 | 19/F | Hemiparesis, headache | Anterior cervicomedullary junction | – | N/A | N/A | – | Right vertebral |
| 5 | 57/F | Headache | Anterior pontomedullary junction | – | + | – | – | Basilar, right vertebral |
| 6 | 40/F | Right V3 trigeminal neuralgia | Right paramedian pontomedullary junction | = | + | N/A | – | Basilar, right vertebral |
| 7 | 27/M | Headache, vision changes | Right paramedian pontomedullary junction | + | N/A | N/A | – | Basilar, right vertebral |

+, hyperintense; –, hypointense; =, equal intensity to brainstem tissue at that level; DWI, diffusion-weighted imaging; M, male; F, female; N/A, not applicable.

specific radiologic findings relevant to posterior fossa neurenteric cysts, we also reviewed imaging studies from a consecutive series of other newly diagnosed infratentorial masses by 2 of the senior authors (P.N., R.F.S.) from 2007 to 2011. Only nonvascular lesions occurring at the level of the basilar artery in the posterior fossa with an established diagnosis through tissue pathology were considered.

All patients harboring an NEC underwent detailed neurological examination at the time of preoperative assessment. Parameters collected included presenting signs and symptoms, cranial nerve dysfunction, sensory-motor impairment, headache, visual changes, and nausea/vomiting. Duration of symptoms also was recorded. Preoperative MRI was performed for all patients on a 1.5-T or 3-T platform (GE Discovery 750, Fairfield, Connecticut). In 2 cases, a limited stereotactic study without T2-weighted sequences or available reconstructions was performed. Diffusion-weighted sequences were performed in 4 of 7 cases.

## RESULTS

A total of 7 patients (2 men, 5 women, mean age 36 years, range 19 to 57 years) harboring NECs were identified for this study. Clinical presentations were variable, but headache was the most common presenting symptom (6 of 7 patients). Duration of symptoms ranged from 1 day to 12 years, and 2 patients presented with acute complaints. A 35-year-old woman (case 2) presented with 1 day of severe headache, and a 19-year-old woman (case 5) reported 10 days of progressive headache followed by 24 hours of progressive hemiparesis. This patient displayed evidence of frank myelopathy and 2 of 5 unilateral lower-extremity motor power. There were no associated MRI findings of hemorrhage, acute hydrocephalus, or cyst rupture in either case, with no prior comparison images available.

A summary of patient symptoms and imaging findings is presented in Table 1. MRI findings revealed varied T1- and T2-weighted signal characteristics across cases, although imaging intensity was relatively homogeneous within each lesion. There was no evidence of diffusion restriction in all 4 available cases, and scant central gadolinium enhancement was evident in 1 of 7 cases. Peripheral or rim enhancement was not seen. Mean lesion size was $1.72 \pm 0.56$ cm in greatest anterior-posterior dimension by $2.17 \pm 0.84$ cm in medial-lateral extent. Ventral vertebrobasilar displacement was seen in all patients, with the NECs seen insinuating between the vertebrobasilar vessels and the pial surface of the brainstem, as seen in Figure 1. Assessment of brainstem perforator, anterior inferior cerebellar, posterior inferior cerebellar, and anterior spinal artery courses could not be adequately evaluated at the imaging resolutions available.

To validate the radiographic findings of ventral vertebrobasilar displacement by NECs, we further reviewed the imaging studies of 266 consecutive patients with nonvascular posterior fossa masses who were operated on by the senior authors (P.N., R.F.S.) from 2007 to 2011. Many of these lesions exerted mass effect onto the basilar artery, but none resulted in ventral displacement of the basilar artery as seen in Figures 2A to 2C. A list of the 19 most common lesions comprising 95% (n = 252) of all pathologies encountered is listed in Table 2. These results correlate to a 100% sensitivity and specificity measure of this imaging finding.

Four patients underwent resection with a far lateral approach, and 3 underwent resection with a retrosigmoid approach. Surgical and clinical data are presented in Table 3. The authors' preferences for positioning, incision, and skull base bony resection are reviewed in detail elsewhere (1, 3). Intraoperatively, tumor was encountered early during cisternal dissection. Tumor consistency was heterogeneous, ranged from mucinous to thin, and was generally amenable to light suction aspiration. Specimens of cyst wall were taken for histopathological examination in all cases. The vertebral, basilar, and associated perforating arteries generally were encountered late in the resection, as cyst decompression approached the lesion's anteromedial borders. The cyst wall was variably adherent to the brainstem, cranial nerves, and/or vasculature in selected cases, and further microdissection risked neurovascular injury. In these scenarios, extensive cyst fenestration and decompression was preferred rather than aggressive gross total resection.

Postoperatively, all patients remained neurologically stable without any worsening of symptoms or neurological deficits. Four of 7 patients obtained a gross total resection by surgeon observation as well as postoperative imaging. At a