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CLINICAL ARTICLE

A taxonomy for brainstem cavernous malformations: subtypes of pontine lesions. Part 2: inferior peduncular, rhomboid, and supraolivary

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OBJECTIVE Part 2 of this 2-part series on pontine cavernomas presents the taxonomy for subtypes 4–6: inferior peduncular (IP) (subtype 4), rhomboid (5), and supraolivary (6). (Subtypes 1–3 are presented in Part 1.) The authors have proposed a novel taxonomy for pontine cavernous malformations based on clinical presentation (syndromes) and anatomical location (MRI findings).

METHODS The details of taxonomy development are described fully in Part 1 of this series. In brief, pontine lesions (323 of 601 [53.7%] total lesions) were subtyped on the basis of predominant surface presentation identified on preoperative MRI. Neurological outcomes were assessed according to the modified Rankin Scale, with score ≤ 2 defined as favorable.

RESULTS The 323 pontine brainstem cavernous malformations were classified into 6 distinct subtypes: basilar (6 [1.9%]), peritrigeminal (53 [16.4%]), middle peduncular (100 [31.0%]), IP (47 [14.6%]), rhomboid (80 [24.8%]), and supraolivary (37 [11.5%]). Subtypes 4–6 are the subject of the current report. IP lesions are located in the inferolateral pons and are associated with acute vestibular syndrome. Rhomboid lesions present to the fourth ventricle floor and are associated with disconjugate eye movements. Larger lesions may cause ipsilateral facial weakness. Supraolivary lesions present to the surface at the ventral pontine underbelly. Ipsilateral abducens palsy is a strong localizing sign for this subtype. A single surgical approach and strategy were preferred for subtypes 4–6: for IP cavernomas, the suboccipital craniotomy and telovelar approach predominated; for rhomboid lesions, the suboccipital craniotomy and transventricular approach were preferred; and for supraolivary malformations, the far lateral craniotomy and transpontomedullary sulcus approach were preferred. Favorable outcomes were observed in 132 of 150 (88%) patients with follow-up. There were no significant differences in outcomes between subtypes.

CONCLUSIONS The neurological symptoms and signs associated with a hemorrhagic pontine subtype can help define that subtype clinically with key localizing signs. The proposed taxonomy for pontine cavernous malformation subtypes 4–6 meaningfully guides surgical strategy and may improve patient outcomes.

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KEYWORDS cavernoma; inferior cerebellar peduncle; pons; rhomboid; subtype; supraolivary; surgical approach; taxonomy; vascular disorders

In Part 2 of this 2-part series on the taxonomy of pontine cavernous malformations (CMs), we describe subtypes 4–6 of these lesions. A complete overview of the 6 pontine subtypes appears in Part 1 of this series. In brief, the pons lies between the midbrain and medulla and is the largest of the 3 brainstem parts.[1,2] It is the most common site of brainstem cavernous malformations (BSCMs)[3] and accounts for about 60% of such lesions in our experience. Approaches to pontine CMs are based on the location where the lesion presents to a pial or ependymal surface.[4] We believe that BSCM anatomy directs the choice of surgical approach and that a taxonomy for such pontine cavernomas could improve clinical decision-making and patient outcomes.

This brief overview of the anatomical taxonomy we developed sets the stage for defining subtypes 4–6 of pontine cavernomas. This BSCM taxonomy is similar to the one for arteriovenous malformations.[5,6] We hypothesized that BSCM types and subtypes would guide the selection of surgical approach and resection strategy more expres-

ABBREVIATIONS BSCM = brainstem cavernous malformation; CM = cavernous malformation; ICP = inferior cerebellar peduncle; INO = internuclear ophthalmoplegia; IP = inferior peduncular; MCP = middle cerebellar peduncle; MLF = medial longitudinal fasciculus; mRS = modified Rankin Scale; PICA = posterior inferior cerebellar artery.

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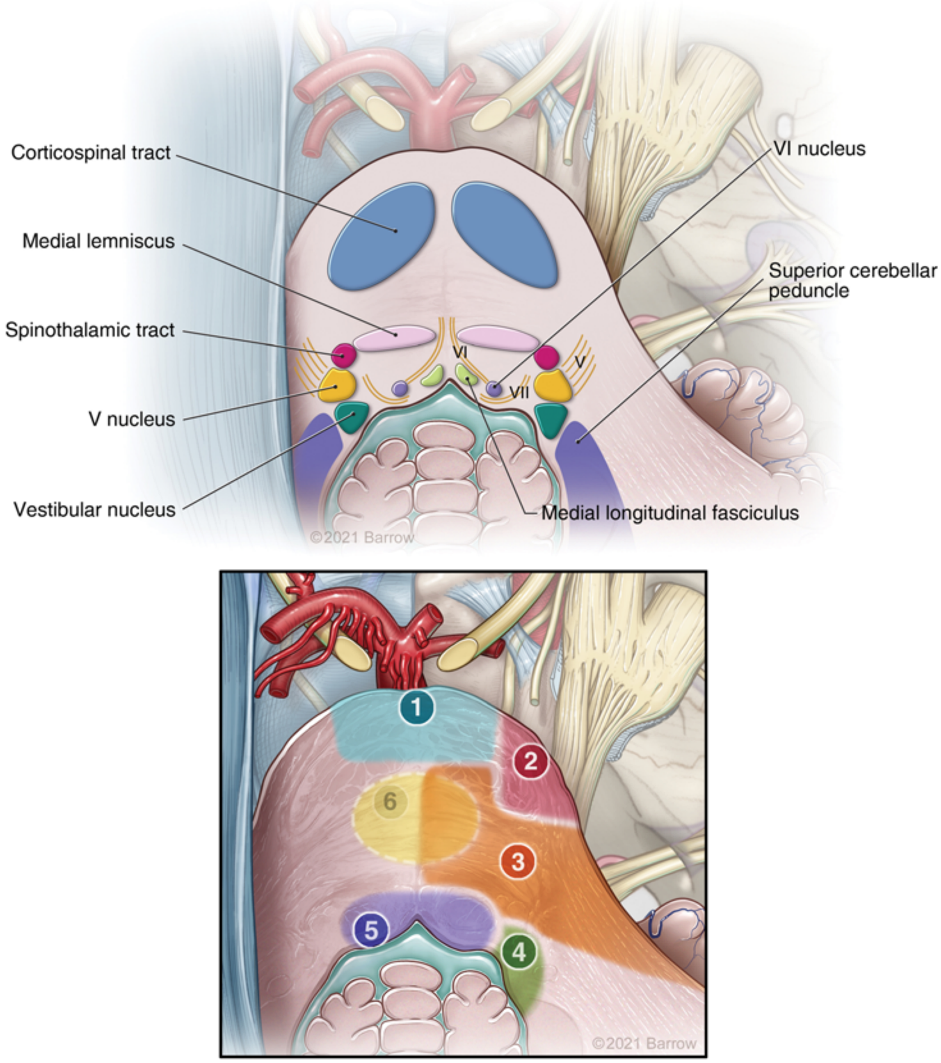

FIG. 1. Overview of pontine anatomy and pontine CM subtypes. This axial cross-section shows the anatomy of the pontine nuclei and their tracts. The locations of the 6 subtypes are shown in the **inset**: 1) basilar (*teal*), 2) peritrigeminal (*red*), 3) middle peduncular (*orange*), 4) IP (*green*), 5) rhomboid (*purple*), and 6) the more inferiorly located supraolivary subtype ghosted through the pons (*yellow*). The IP, rhomboid, and supraolivary subtypes are featured in this article (Part 2 of this 2-part series), whereas the basilar, peritrigeminal, and middle peduncular subtypes (1–3) are featured in Part 1. *Roman numerals* indicate the CNs. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

sively than the present practice of intuitive judgment. Additionally, the constellation of neurological symptoms and signs associated with a particular hemorrhagic BSCM suggests a unique clinical definition when considering the taxonomic subtype of the lesion. Therefore, any BSCM would be associated with a recognizable neurological syndrome and characteristic radiographic anatomy. Such a taxonomy would provide standardized descriptions of the range of BSCM lesions and enhance clinical communications and publications.

Methods

Patients

The overall BSCM taxonomy was developed for pontine lesions and applied to a large 2-surgeon experience to determine the unique clinical presentations and preferred surgical approaches for each of the 6 pontine subtypes (Fig. 1). The study protocol was approved by the St. Joseph's Hospital and Medical Center Institutional Review Board, Phoenix, Arizona. Informed consent was waived due to the retrospective nature of the study and the minimal risk of patient identification. A data-sharing and collaboration agreement with the University of California, San Francisco, was established. Data were collected from the medical records of patients who underwent microsurgical treatment for BSCMs from January 1990 through December 2019. The data were entered into a combined database. Neurological outcomes were measured using the modified Rankin Scale (mRS), with good outcome defined as mRS score ≤ 2 and poor outcome defined as mRS score > 2. The relative outcome was measured as the difference between mRS score at preoperative baseline and that at the last available postoperative examination, with good outcome defined as unchanged or improved mRS score and poor outcome defined as worsened mRS score. Statistical significance was defined as $p \leq 0.05$.

Of the 601 patients with BSCMs identified, 551 had the clinical and radiological information needed for inclusion in the study. The overall results showed that 151 of 551 (27%) BSCMs were found in the midbrain, 323 (59%) in the pons, and 77 (14%) in the medulla. BSCMs were classified into subtypes on the basis of lesion presentation to a pial or ependymal surface (e.g., anterior, anterolateral, lateral, posterolateral, posterior). Six subtypes (Fig. 1) were defined for pontine CMs,[2] of which subtypes 4, 5, and 6 are described in this report: inferior peduncular (IP) (47 of 323 lesions [14.6%]), rhomboid (80 of 323 [24.8%]), and supraolivary (37 of 323 [11.5%]).

Anatomy of Pontine CM Subtypes 4–6

Inferior Peduncular CMs

IP pontine CMs are located in the posterolateral pons at the level of the inferior cerebellar peduncle (ICP), just anterior to the lateral recess of the fourth ventricle. The stria medullaris, which delineates the pons above and the medulla below, forms the lower limit of IP lesions.[7,8] These lesions reside anteriorly to the stria medullaris and above the foramen of Luschka, distinct from rhomboid malformations on the floor of the fourth ventricle. IP CMs are lateral to the internal genu of the facial nerve and its intrapontine course and can involve the vestibular nuclei (medial, lateral), cochlear nucleus, spinal nucleus and tract of the trigeminal nerve, superior salivatory nucleus, nucleus solitarius, and restiform and juxtarestiform bodies.

Rhomboid CMs

Rhomboid pontine CMs are located in the posterior pons and present to the surface of the floor of the fourth ventricle. The ventricular floor, also known as the rhomboid fossa, has 3 parts: a pontine part extending from the aqueduct downward to form a triangle with an imaginary line connecting the lower margin of the cerebellar peduncles; a junctional or intermediate part forming a thin strip between the lateral recesses; and a medullary part extending downward to form a triangle with the obex.[1] The surface landmarks in the pontine part of the rhomboid fossa include 1) the median sulcus, which divides the floor longitudinally along the midline; 2) the median eminence, a raised strip parallel to the median sulcus; 3) the facial colliculus, which is centered on the median eminence; 4) the sulcus limitans, another longitudinal sulcus on the lateral border of the median eminence; 5) the locus ceruleus, a bluish-gray area at the rostral end of the sulcus limitans in the lateral margin of the floor; and 6) the superior foveae, which are dimples in the sulcus limitans just lateral to the facial colliculus.[1,9] The facial colliculus overlies the abducens nucleus and the ascending root of cranial nerve (CN) VII.[1] The stria medullaris consists of white matter strands traversing the junctional part of the floor, from the lateral recess to the sulcus limitans, and overlying the vestibular nuclei.[1,7] Rhomboid pontine CMs lie above the stria medullaris, whereas lesions at or below the level of the stria medullaris are considered trigonal medullary lesions. Critical structures below the floor include the medial longitudinal fasciculus (MLF), dorsal longitudinal fasciculus, reticular formation, tectospinal tract, central tegmental tract, ventral trigeminothalamic tract, medial lemniscus, lateral lemniscus, and trapezoid body.[1,4]

Supraolivary CMs

Supraolivary pontine CMs are located in the inferior pons and project downward to the pial surface at the pontomedullary junction. The intersection of the pons and the medulla creates a pontine underbelly or an overhanging of the basis pontis that enables access to lesions in the central pons from below. This anatomy allows the surgeon entry to a region that is otherwise surrounded by eloquent tracts and nuclei.[1,4,10,11] This point of presentation lies above the inferior olivary nucleus, lateral to the exit of the abducens nerve, medial to the exit of the facial nerve, and above the distal a1 (anterior pontine) and proximal a2 (lateral pontine) segments of the anterior inferior cerebellar artery. Here, CMs lie lateral to the corticospinal tract and transpontine course of the abducens nerve within the lower transverse pontine fibers. The supraolivary trajectory leads to the center of the pons near the medial lemniscus, lateral lemniscus, spinothalamic tract, superior olivary nucleus, rubrospinal tract, and trapezoid body.[1,4,10,11]

Results

Overall Clinical Presentation

We describe the nuances of the constellation of signs and symptoms of pontine subtypes 4–6 (the IP, rhomboid, and supraolivary subtypes) that the surgeon can use to determine the subtype of the lesion and the best approach. We summarize the overall characteristics of the patients with pontine lesions to provide the study context and continuity for reader convenience. Descriptions of subtypes 1–3 are provided in greater detail in Part 1 of this 2-part series.[2]

In brief, of 323 patients with pontine BSCMs (mean ± SD age 41.2 ± 14.2 years; 197 [61%] female patients; 126 [39%] male patients), most (310 of 323 patients [96%]) presented with symptomatic hemorrhage.[2] Of the 178 patients with pontine lesions who had Lawton BSCM grades available, 66 (37%) had low-grade BSCMs (grades 0–II), 100 (56%) had intermediate-grade BSCMs (grades III–V), and 12 (7%) had high-grade BSCMs (grades VI and VII) (Table 1). There was no significant relationship between BSCM grade and pontine CM subtype ($p = 0.20$).

Pontine Syndromes in Subtypes 4–6

Patients with IP pontine CMs were the most likely to present with nausea/vomiting, vertigo, or both (14 of 47 patients [30%]) caused by involvement of the vestibular nuclei. Common symptoms included limb or gait ataxia (22 of 47 patients [47%]), contralateral hemisensory loss (21 [45%]), and ipsilateral facial numbness (14 [30%]) (Fig. 2).

Patients with rhomboid pontine CMs frequently had diplopia (41 of 80 patients [51%]), primarily as a result of inter-

TABLE 1. Patient outcomes after resection of pontine CMs according to subtype

Variable	Pontine CM Subtype							p Value
	All Subtypes	Basilar	Peritrigeminal	Middle Peduncular	Inferior Peduncular	Rhomboid	Supraolivary	
BSCM grade available	178	3	40	51	25	35	24	0.20
Low (0–II)	66 (37)	1 (2)	13 (20)	19 (29)	7 (11)	10 (15)	16 (24)	
Intermediate (III–V)	100 (56)	1 (1)	25 (25)	30 (30)	16 (16)	21 (21)	7 (7)	
High (VI–VII)	12 (7)	1 (8)	2 (17)	2 (17)	2 (17)	4 (33)	1 (8)	
Follow-up data available	323	6	53	100	47	80	37	
Follow-up, mos	29 ± 39	17 ± 9	26 ± 38	31 ± 39	28 ± 36	32 ± 49	19 ± 22	0.58
Follow-up >30 days	279 (86)	6 (2)	46 (16)	84 (30)	42 (15)	68 (24)	33 (12)	0.91
Preop mRS score	2 (1–2)	2 (1.8–3.1)	1 (1–2)	1 (1–2)	1 (1–2)	2 (1–2)	2 (1–2)	0.73
Preop mRS score >2	54 (17)	2 (4)	6 (11)	20 (37)	7 (13)	15 (28)	4 (7)	0.73
Postop mRS score	2 (1–2)	2 (1.8–2.3)	1 (1–2)	2 (1–3)	2 (1–3)	2 (2–3)	2 (1–2)	0.31
Postop mRS >2	75 (23)	1 (1)	6 (8)	25 (33)	13 (17)	22 (29)	8 (11)	0.31
Final mRS data available	293	6	49	88	41	74	35	0.39
Final mRS score	1 (1–2)	1 (1–1)	1 (1–2)	1 (1–2)	1 (1–2)	1 (1–2)	1 (1–2)	0.30
Final mRS score >2	38 (13)	0 (0)	5 (13)	15 (39)	6 (16)	11 (29)	1 (3)	0.35
Final mRS score ≤2	255 (87)	6 (2)	44 (17)	73 (29)	35 (14)	63 (25)	34 (13)	0.35
Worse mRS score	41 (14)	0 (0)	8 (20)	11 (27)	7 (17)	10 (24)	5 (12)	0.98

Values are shown as number, number (%), mean ± SD, or median (interquartile range) unless indicated otherwise.

nuclear ophthalmoplegia (INO) and injury to the abducens nerve nucleus (37 [46%]). More than one-third of patients (29 of 80 [36%]) presented with facial palsy caused by the involvement of the facial nucleus. Hemisensory loss (41 patients [51%]) and ataxia (24 [30%]) were also common components of the rhomboid symptom constellation (Fig. 2).

Supraolivary CMs were often associated with an isolated ipsilateral abducens nerve palsy (26 of 37 patients [70%]) caused by the proximity of the lesion to the transpontine fibers of the nerve. A more anterior location of this lesion and its proximity to the corticospinal tract resulted in hemiparesis in 17 patients (46%). Proximity to the facial nerve laterally and lemniscal systems posteriorly accounted for facial palsy in 11 patients (30%) and contralateral hemisensory deficits in 18 patients (49%) (Fig. 2).

Overall, the most common symptoms of the patients with pontine CMs were hemisensory deficit (141 of 323 patients [43.6%]), diplopia (116 [35.9%]), ataxia (103 [31.9%]), and abducens nerve palsy (100 [31.0%]) (Fig. 2). Abducens palsy had a greater association with supraolivary (26 of 37 patients [70%]) and rhomboid (37 of 80 [46%]) lesions than with other subtypes (37 of 206 [18%], $p < 0.001$). Similarly, facial palsy had a greater association with rhomboid (29 of 80 patients [36%]) and supraolivary (11 of 37 [30%]) lesions than with other subtypes (32 of 206 [16%], $p = 0.001$). Nausea and vomiting were more strongly associated with ICP lesions than other lesions (30% vs 16%, $p = 0.04$). There was no significant association between hemisensory loss and pontine subtype ($p = 0.31$).

Symptom	Subtype					
	Basilar (n=6)	Peritrigeminal (n=53)	MP (n=100)	IP (n=47)	Rhomboid (n=80)	Supraolivary (n=37)
Hemisensory deficit	1 (17%)	18 (34%)	42 (42%)	21 (45%)	41 (51%)	18 (49%)
Hemiparesis	4 (67%)	22 (42%)	28 (28%)	9 (19%)	15 (19%)	17 (46%)
Ataxia	1 (17%)	15 (28%)	34 (34%)	22 (47%)	24 (30%)	7 (19%)
N/V, vertigo	0 (0%)	6 (11%)	20 (20%)	14 (30%)	16 (20%)	2 (5%)
Abducens nerve palsy	1 (17%)	10 (19%)	15 (15%)	11 (23%)	37 (46%)	26 (70%)
Facial weakness	2 (33%)	2 (4%)	17 (17%)	11 (23%)	29 (36%)	11 (30%)

FIG. 2. Heat map showing the symptoms of patients with pontine CMs according to subtype. Percentages were calculated as the frequency of a particular symptom or sign within a given population with a midbrain subtype. The sensitivity and specificity of neurological findings vary significantly. For example, hemiparesis is more likely to be accurately documented in medical records than trochlear nerve palsy. Thus, for each symptom or sign, the rates were sorted according to their relative frequency across the 6 subtypes. Conditional formatting was applied on a scale from least common (*red*) to most common (*green*). The syndromes most closely associated with each subtype are shown with green formatting. MP = middle peduncular; N/V = nausea/vomiting.

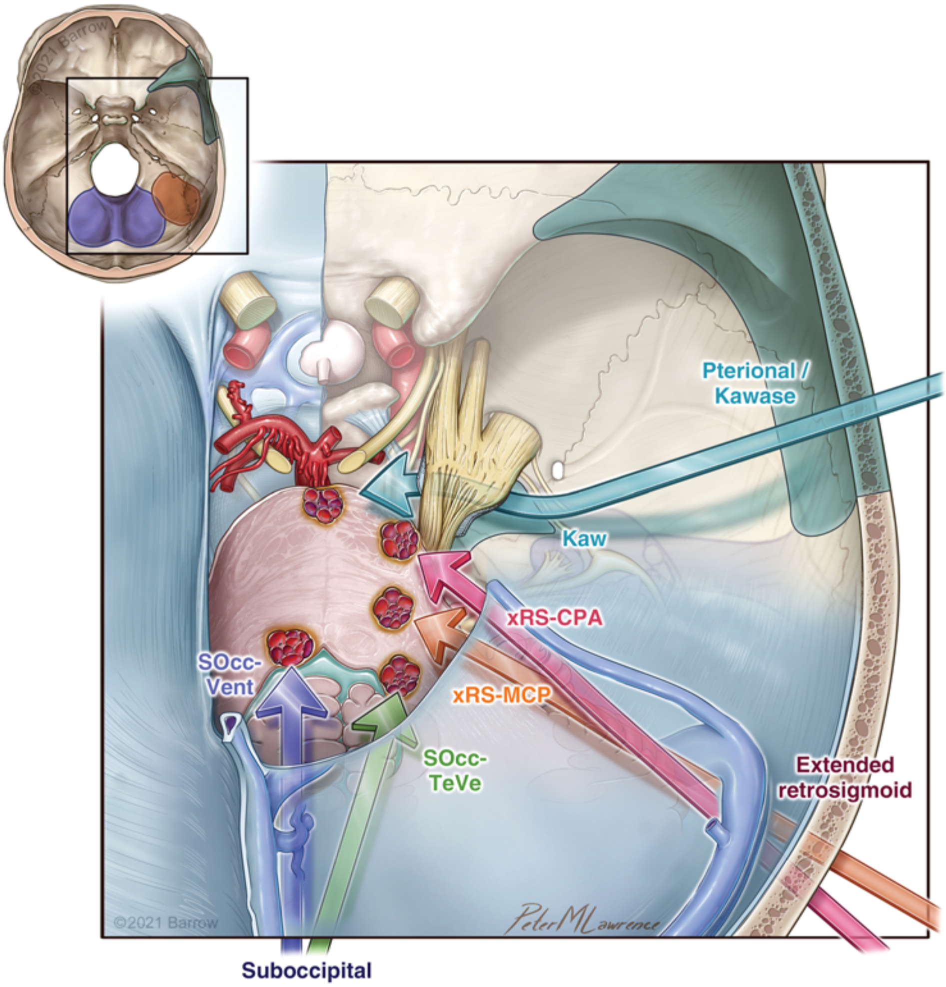

FIG. 3. Overview of surgical approaches to pontine CMs shown in an axial cross-section viewed from above. Craniotomies associated with these approaches are shown in relation to the skull base (**inset**). The pterional craniotomy and anterior transpetrous (Kawase) approach are used for basilar BSCMs (*Kaw*, *teal arrow*). The extended retrosigmoid craniotomy and transcerebellopontine angle approach (*xRS-CPA*, *red arrow*) is used for peritrigeminal BSCMs, and the extended retrosigmoid craniotomy and trans-MCP approach (*xRS-MCP*, *orange arrow*) is used for middle peduncular BSCMs. The suboccipital craniotomy and transventricular approach (*SOcc-Vent*, *purple arrow*) is used for rhomboid lesions, and the suboccipital craniotomy and telovelar approach (*SOcc-TeVe*, *green arrow*) is used for inferior peduncular lesions. Not shown is the far lateral craniotomy–transpontomedullary sulcus approach used for supraolivary lesions located more inferiorly in the pons. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

Surgical Approaches to Subtypes 4–6

Knowledge of the pontine CM subtype was useful for determining the optimal surgical approach (Fig. 3). All 47 IP lesions were resected through a suboccipital craniotomy and telovelar approach. The approach to rhomboid lesions was transventricular in most cases (65 of 80 patients [81%]) and transvermian for lesions located high in the superior portion of the floor (13 [16%]). Supraolivary lesions were resected with a far lateral craniotomy and transpontomedullary sulcus approach (22 of 37 patients [59.5%]), with the extended retrosigmoid craniotomy and transcerebellopontine angle approach used earlier in our experience (15 [40.5%]).

Outcomes of Subtypes 4–6

Favorable outcomes were observed after final microsurgical resection in 132 of 150 (88%) patients with pontine CMs (mean ± SD follow-up 28 ± 39 months) (Table 1). Postoperative mRS scores were unchanged or improved for most patients (128 patients [85.3%]). Neither the proportions of patients with favorable outcomes nor those with worsening mRS scores differed significantly between patients with different pontine subtypes.

Discussion

This study reports the nuances of pontine subtypes 4–6 (IP, rhomboid, and supraolivary), building on our effort to establish an anatomical BSCM taxonomy as described in detail in Part 1 of this series.[2] The taxonomy is intended to guide the selection of surgical approaches and resection strategies. Our study also confirms that the constellations of neurological symptoms and signs of the hemorrhagic BSCM subtypes help to define these clinical subtypes even before pathological anatomical findings are apparent on MRI. Pontine CMs have key localizing signs, unlike midbrain lesions that have classic syndromic presentations. Nonetheless, most eponymous pontine vascular syndromes[12] are unsuitable for describing the clinical presentations of pontine BSCMs. Specific signs are more useful than syndromes for localizing a pontine CM. For example, nausea and vomiting are commonly associated with IP lesions, facial weakness and INO are more common with rhomboid lesions, and ipsilateral abducens palsy is associated with supraolivary lesions. Thus, our proposed anatomical and syndrome-driven taxonomy can improve bedside diagnostic acumen.

Pontine CMs Subtypes 4–6

Inferior Peduncular CMs

The ICP carries afferent (posterior spinocerebellar, vestibulocerebellar, olivocerebellar, and reticulocerebellar tracts) and efferent fibers that integrate proprioceptive sensory input with motor vestibular functions such as balance and posture, unlike the MCP, which contains only afferent fibers.[13,14] The ICP occupies the inferolateral corner of the pons, ventral to the lateral recess of the fourth ventricle, as it courses upward before turning posteriorly under the MCP and superior cerebellar peduncle.[1,14] Patients with IP pontine CMs present with an acute vestibular syndrome of nausea, vomiting, vertigo, ataxia, imbalance, and nystagmus related to the involvement of adjacent vestibular nuclei (Fig. 4). No other pontine CM is associated with nausea, vomiting, and vestibular symptoms, thereby making this syndrome a localizing sign. Larger lesions that extend beyond the vestibular nuclei and ICP cause diplopia and ipsilateral facial hemisensory loss related to the paramedian pontine reticular formation and spinal trigeminal nucleus and tract.

The suboccipital craniotomy and telovelar approach were the only methods selected to treat IP lesions (Fig. 5). A generous midline suboccipital craniotomy is used along with C1 laminectomy to increase the exposure of the cisterna magna and fourth ventricle. The cerebellar tonsils are liberated to open the vallecula and mobilize the ipsilateral tonsil laterally. The p3 (tonsillomedullary) segment of the posterior inferior cerebellar artery (PICA) and its caudal loop are mobilized from the posterior medulla and swept laterally to access the inferior medullary velum. The incision of the velum from the foramen of Magendie to the foramen of Luschka superolaterally along the tela choroidea unroofs the fourth ventricle and leads to the lateral recess. A fixed retractor holds the tonsil laterally, thus forming a natural working triangle between the incised attachment of the velum inferiorly, the retracted tonsil superiorly, and the midline (i.e., the subtonsillar triangle). The stria medullaris provides an obvious surface landmark in the fourth ventricular floor, leading to the foramen of Luschka and revealing the inferior limit of the pons. The area acustica, defined as the floor of the lateral recess overlying the cochlear and vestibular nuclei and inferior to the stria medullaris, is one of the pontine safe entry zones available for accessing subependymal lesions.

Rhomboid CMs

Surgical access through the floor of the fourth ventricle requires caution and care because of the dense concentration of CNs and nuclei nearby.[1] The rhomboid fossa is so named because of its shape. Its upper border is created by the outlet of the sylvian aqueduct, and the lower border, by the stria medullaris (Fig. 6). The rhomboid fossa forms the posterior surface of the pons.[1] CMs presenting to the fourth ventricular floor constitute rhomboid lesions when they reside in the posterior pons. Patients with rhomboid pontine lesions present with disconjugate eye movement caused by injuries to the abducens nerve nucleus, MLF, and paramedian pontine reticular formation (Fig. 6). Syndromes vary depending on lesion size and midline or eccentric location. Syndromes associated with midline lesions include classic INO with MLF damage and failure of the medial rectus muscles to move synchronously with the lateral rectus muscles during attempted lateral gaze.[15] Reverse INO with intact MLF also occurs, with bilateral damage affecting the relay to the abducens nuclei and failure of the lateral rectus muscles to move synchronously with the medial rectus muscles during attempted lateral gaze. Two syndromes are associated with eccentric lesions.[16] The first syndrome is unilateral INO with impaired adduction during attempted gaze away from the lesion and normal abduction in the contralateral eye due to a unilateral MLF lesion. The second syndrome is one-and-a-half syndrome, with no movement of the eye ipsilateral to the lesion and only outward movement of the other eye (conjugate gaze palsy toward the lesion and INO away from the lesion) due to unilateral paramedian pontine reticular formation and MLF damage.[16] Larger eccentric lesions cause facial colliculus syndrome with ipsilateral horizontal gaze palsy and ipsilateral facial weakness (lower motor neuron pattern) caused by damage affecting the abducens nerve nuclei and the internal genu of the facial nerve (“eight-and-a-half” syndrome).[17–19]

A suboccipital craniotomy with C1 laminectomy and transventricular approach exposes the rhomboid fossa (Fig. 7). The natural trajectory of this approach is typically too low for most pontine lesions, but head flexion with the chin tucked in the prone position steepens the trajectory upward and allows the surgeon to reach the upper rhomboid fossa. Dissection along the p3 segment of the PICA opens the tonsillomedullary fissures, liberates the tonsils from the medulla, and widens the vallecula (vallecular triangle). The inferior vermis (uvula and nodule deep to it) can still obscure visualization. Thus, limited resection of the vermis could be required for lesions located high in the upper pons. Eccentric lesions lying off the midline may require release of the inferior medullary velum, as performed with a telovelar approach. The median sulcus, facial colliculus, superior fovea, and stria medullaris are landmarks on the upper floor. The facial colliculus and MLF are forbidden zones.

The safe entry zones include the median sulcus and suprafacial and infrafacial colliculi. The median sulcus runs along the midline. Careful entry for small lesions preserves the MLF, which is immediately lateral. The suprafacial and infrafacial collicular safe entry zones are off the midline within the median eminence.[10,11] The superior foveal safe entry zone is a fourth site lateral to the facial colliculus in the sulcus limitans.[20] The superior fovea is just a dimple in the sulcus limitans, giving the superior

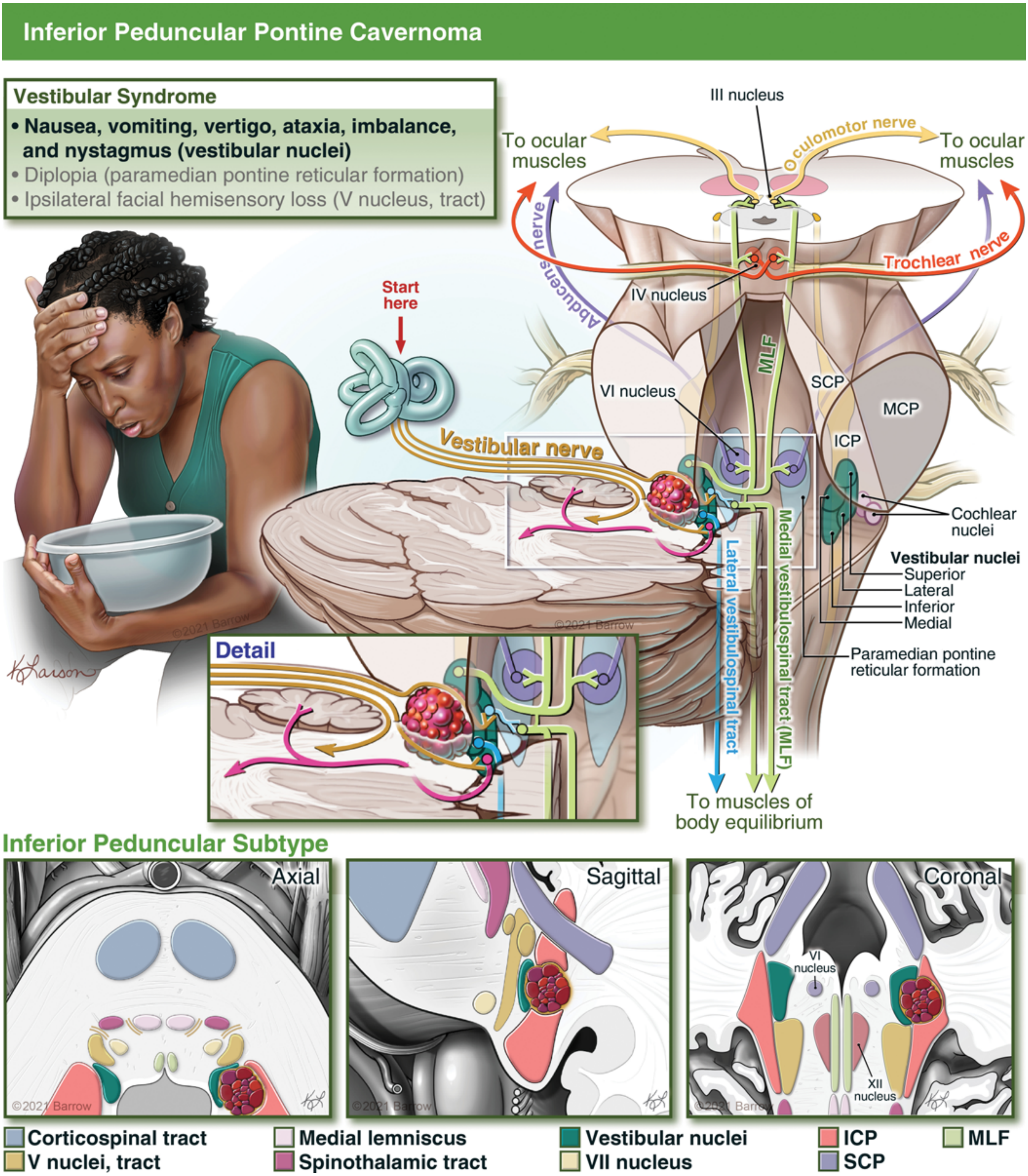

FIG. 4. Patients with IP pontine CMs present with an acute vestibular syndrome of nausea, vomiting, vertigo, ataxia, imbalance, and nystagmus related to the involvement of adjacent vestibular nuclei. Larger lesions cause diplopia and ipsilateral facial numbness (not shown) related to the paramedian pontine reticular formation and spinal trigeminal nucleus and tract (see detail in **inset**). This left-sided IP CM resides posterolaterally in the pons, above the lateral recess of the fourth ventricle, and involves the ICP and vestibular nuclei, as shown in the axial, sagittal, and coronal views (**lower insets**). *Roman numerals* indicate the CNs. SCP = superior cerebellar peduncle. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

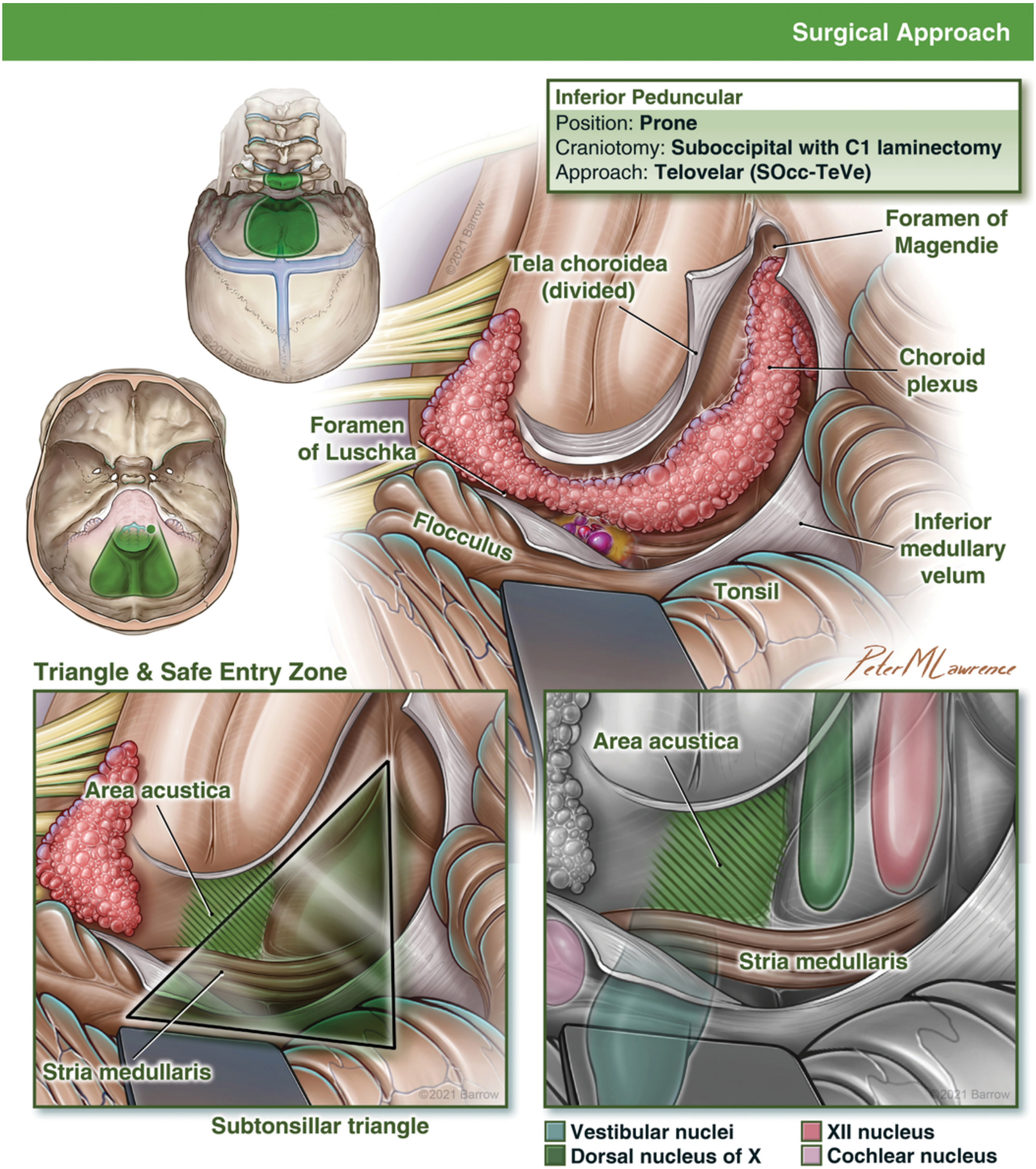

FIG. 5. IP pontine CMs are exposed surgically through a suboccipital craniotomy, C1 laminectomy (**upper left inset**), and telovelar approach (**middle left inset**). Retraction of the cerebellar tonsil superiorly and incision of the inferior medullary velum along the tela choroidea opens the subtonsillar triangle, through which the fourth ventricle is entered (**center panel**). Removal of the choroid plexus gives an overview of the ventricular floor, stria medullaris, lateral recess, foramen of Luschka, area acustica, and IP CM (**lower left inset**). The area acustica overlying the cochlear and vestibular nuclei creates a safe entry zone for accessing IP lesions located deep to the ependyma. Careful dissection protects the cochlear nuclei laterally and the vestibular nuclei, paramedian pontine reticular formation, and spinal trigeminal nucleus and tract medially (**lower right inset**). *Roman numerals* indicate the CNs. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

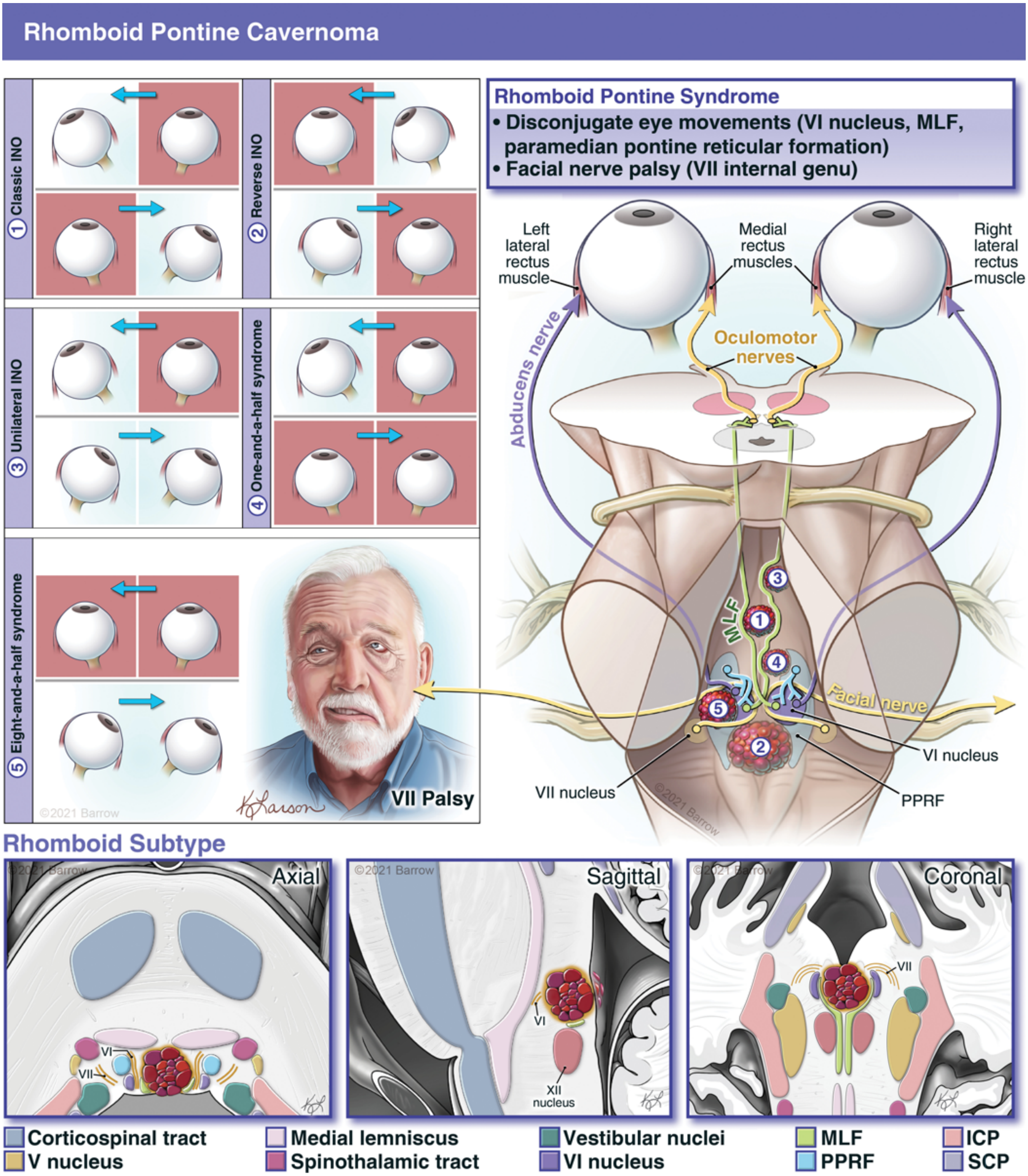

FIG. 6. Patients with rhomboid pontine CMs present with disconjugate eye movement and INOs caused by injuries to the MLF, abducens nerve nucleus, and paramedian pontine reticular formation. Syndromes associated with midline lesions include classic INO (*1*) (MLF damage and failure of the medial rectus muscle to move synchronously with the lateral rectus muscle during attempted lateral gaze) and reverse INO (*2*) (bilateral abducens nuclei injury with intact MLF and failure of the lateral rectus muscle to move synchronously with the medial rectus muscle during attempted lateral gaze). Eccentric rhomboid CMs can cause unilateral INO (*3*) (unilateral MLF lesion with impaired adduction during attempted gaze away from the lesion and normal abduction in the contralateral eye) and one-and-a-half syndrome (*4*) (unilateral paramedian pontine reticular formation and MLF lesion, with no movement of the eye ipsilateral to the lesion and only outward movement of the other eye [conjugate gaze palsy toward the lesion and INO away from the lesion]).
FIG. 6. (*continued*) →

FIG. 6. Larger eccentric lesions affecting the abducens nerve nucleus and internal genu of the facial nerve cause facial colliculus syndrome or eight-and-a-half syndrome (*5*) (ipsilateral horizontal gaze palsy and ipsilateral facial weakness). *Blue arrows* in the **left insets** indicate the direction of the patient's attempted lateral gaze. The *red background* indicates abnormal eye findings, and the *white background* indicates normal eye movements. This midline rhomboid CM resides in the posterior pons and floor of the fourth ventricle adjacent to the MLF, CN VI nuclei, and paramedian pontine reticular formation (PPRF), as shown in the axial, sagittal, and coronal views (**lower insets**). *Roman numerals* indicate the CNs. SCP = superior cerebellar peduncle. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

foveal triangle its name. The working triangle is defined superolaterally by the superior cerebellar peduncle, inferolaterally by the vestibular area, and medially by the sulcus limitans.[20] The apex of the triangle maps at the transverse level to the upper edge of the facial colliculus. The motor and main sensory nuclei of the trigeminal nerve lie deep to the superolateral border. Therefore, this triangle provides landmarks for the motor nuclei of the trigeminal, abducens, and facial nerves. A vertical incision in the superior foveal triangle provides safe access to more laterally placed lesions.[20] Hemosiderin staining or lesion presentation on the ependymal floor guides the ultimate entry site to open the lesion corridor. Stimulation mapping is helpful when the safe entry zone is not obvious or when the lesion distorts anatomy.

Supraolivary CMs

The ventral pontine underbelly creates a supraolivary access portal for inferolateral pontine CMs that lie lateral to the pyramidal tract and abducens nerve and medial to the lower CNs and project upward in the central pons.[1,11] Ipsilateral abducens nerve palsy was a strong localizing sign for this subtype, with more than two-thirds of patients presenting with this finding. Patients with supraolivary lesions present with mild Millard-Gubler syndrome, one of the classic pontine crossed syndromes (Fig. 8). It consists of ipsilateral abducens and facial nerve palsies, as well as contralateral hemiparesis or hemiplegia.[21] Supraolivary CMs displace the pyramidal tract and thus are associated with milder motor deficits. The medial lemniscus and spinothalamic tracts were spared in half of these patients. Nonetheless, deficits in these tracts were present in those patients with larger lesions that extended dorsally to involve the pontine tegmentum and produced contralateral hemisensory deficits (e.g., Foville syndrome).[21]

A far lateral craniotomy and a transpontomedullary sulcus approach are ideal for reaching the pontine underbelly (Fig. 9).[22] The park bench position used for this approach orients the clivus and vertebral artery vertically to allow for viewing along the axis of the brainstem anterolaterally. An extensive condylectomy is used to open the cerebellomedullary cistern and expose the vagoaccessory triangle, revealing the olivary nucleus. The vagus nerve is separated from the medullary rootlets of the accessory nerve to widen the triangle. The V4 segment of the vertebral artery resides at the depths of this triangle, and the hypoglossal nerve rootlets drape the segment.[1,22] With the steep upward trajectory to the pontine underbelly, the transpontomedullary sulcus approach moves superiorly through the vagoaccessory triangle into its suprahypoglossal portion. Deepening dissection through these triangles moves the surgeon medially toward the lower CN complex (CNs IX–XI).[22] The p2 (lateral medullary) segment of the PICA often travels in this working window. The contours of the superior olive and inferior pons intersect at the pontomedullary sulcus, which is the safe entry zone for these lesions.[10,11,22] CN VI also exits from this sulcus, and its cisternal segment can be traced in the retrograde direction to identify the sulcus. Entry lateral to CN VI protects the corticospinal tract. The superior pole of the olive, rather than the pontomedullary sulcus, can be entered safely if a more posterior trajectory is needed.[22]

Limitations

Some lesions in our experience did not conform with the definitions and descriptions of our taxonomy. About one-quarter of pontine lesions spanned multiple regional subtypes, complicating our analysis. Instead of considering these lesions as blended subtypes, we categorized them according to their dominant subtype. We acknowledge some overlap in symptoms and signs between pontine subtypes. Overlapping symptoms and signs exist between adjacent subtypes, especially because larger lesions occupy more brainstem territory and between adjacent types that extend up into the midbrain or down into the medulla. Characterization of clinical syndromes was limited by the lack of detailed documentation about the findings of the neurological examinations in the medical records. Furthermore, these data were more qualitative than the anatomical MRI findings.

Additionally, determination of the features of the surgical approach depended on precise documentation in the operative reports, which sometimes required retrospective clarification. Also, surgical selection bias could have played a potential role in this study of patients who were treated by 2 experienced neurosurgeons. Large BSCMs spanning 2 or more brainstem territories are more challenging to manage surgically than small lesions. A subtype's prescribed approach may apply to the portion of the lesion inside that subtype's territory but not to the portion outside this territory, thereby making approach selection less clear. In most cases, the choice of surgical approach was based on the dominant subtype, and communication with other portions of the lesion was established from within the resection cavity on the basis of hematoma evacuation, circumferential dissection, and piecemeal resection of the lesion. To our knowledge, this is the largest cohort study of patients with pontine BSCMs.

Conclusions

The proposed taxonomy offers a useful scheme to classify pontine CMs into subtypes, decipher their associated clinical presentations, and guide surgical decision-making for these relatively rare lesions. To date, this is the largest patient series of surgically treated pontine CMs. The majority of patients with pontine BSCMs had favorable functional outcomes.

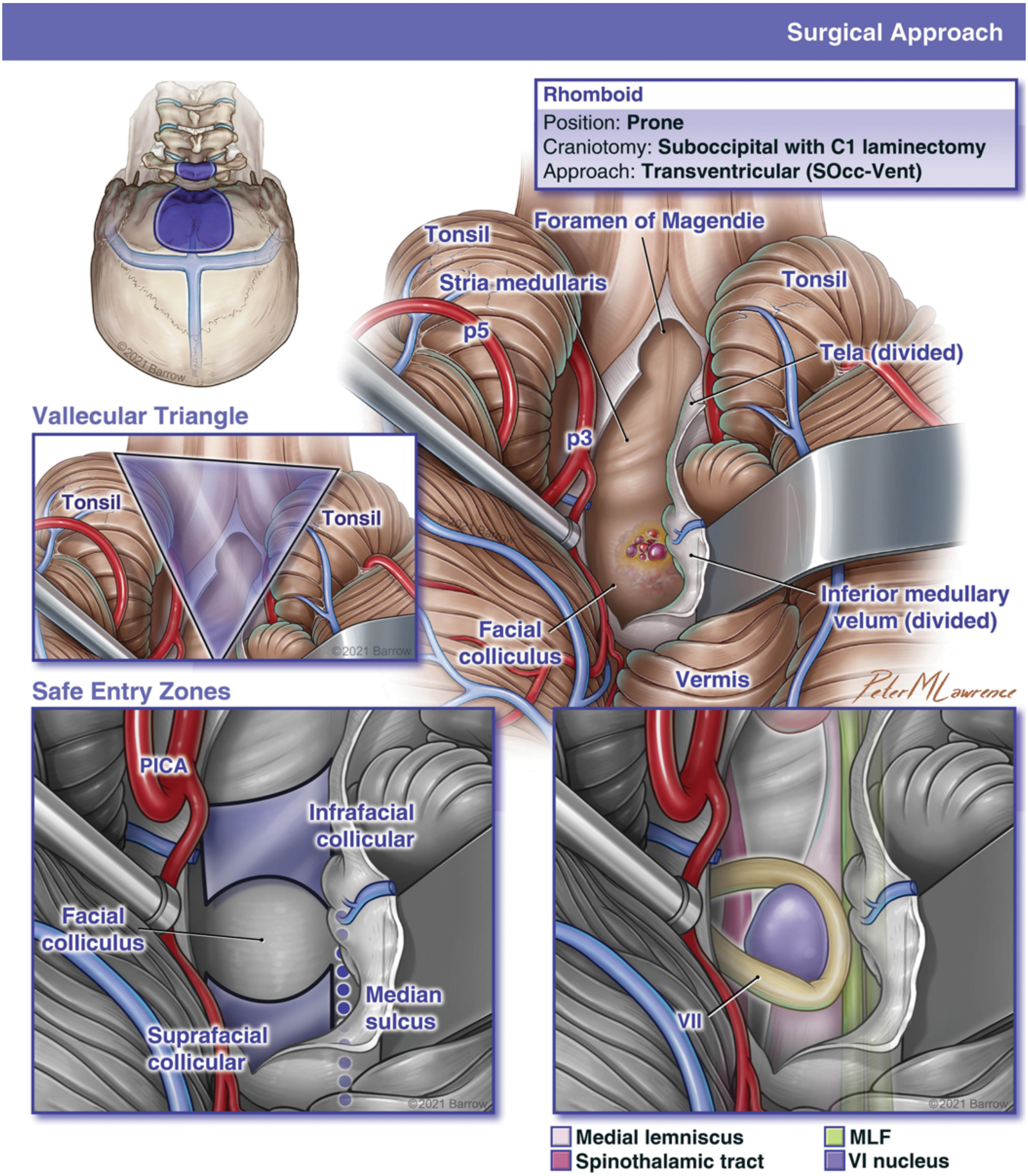

FIG. 7. Rhomboid pontine CMs are exposed with a suboccipital craniotomy, C1 laminectomy (**upper left inset**), and transventricular approach. Dissection into the tonsillomedullary fissures bilaterally and liberation of the tonsils opens the vallecular triangle (**middle left inset**). Incision and release of the inferior medullary velum, as with a telovelar approach, extends exposure superiorly and allows the surgeon to reach the pontine portion of the rhomboid fossa (**central panel**) where the stria medullaris, median sulcus, median eminence, sulcus limitans, facial colliculus, and superior foveae are visualized. The fourth ventricular floor can be entered safely at 4 locations (**lower left inset**): the median sulcus (*purple dotted line*) and the suprafacial collicular, infrafacial collicular, and superior foveal safe entry zones (not shown). Safe entry avoids the MLF (*green*, **lower right inset**), abducens nerve nuclei (*purple*), and internal genu of the facial nerve (*yellow*). *Roman numerals* indicate the CNs. p5 = cortical segment of the PICA. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

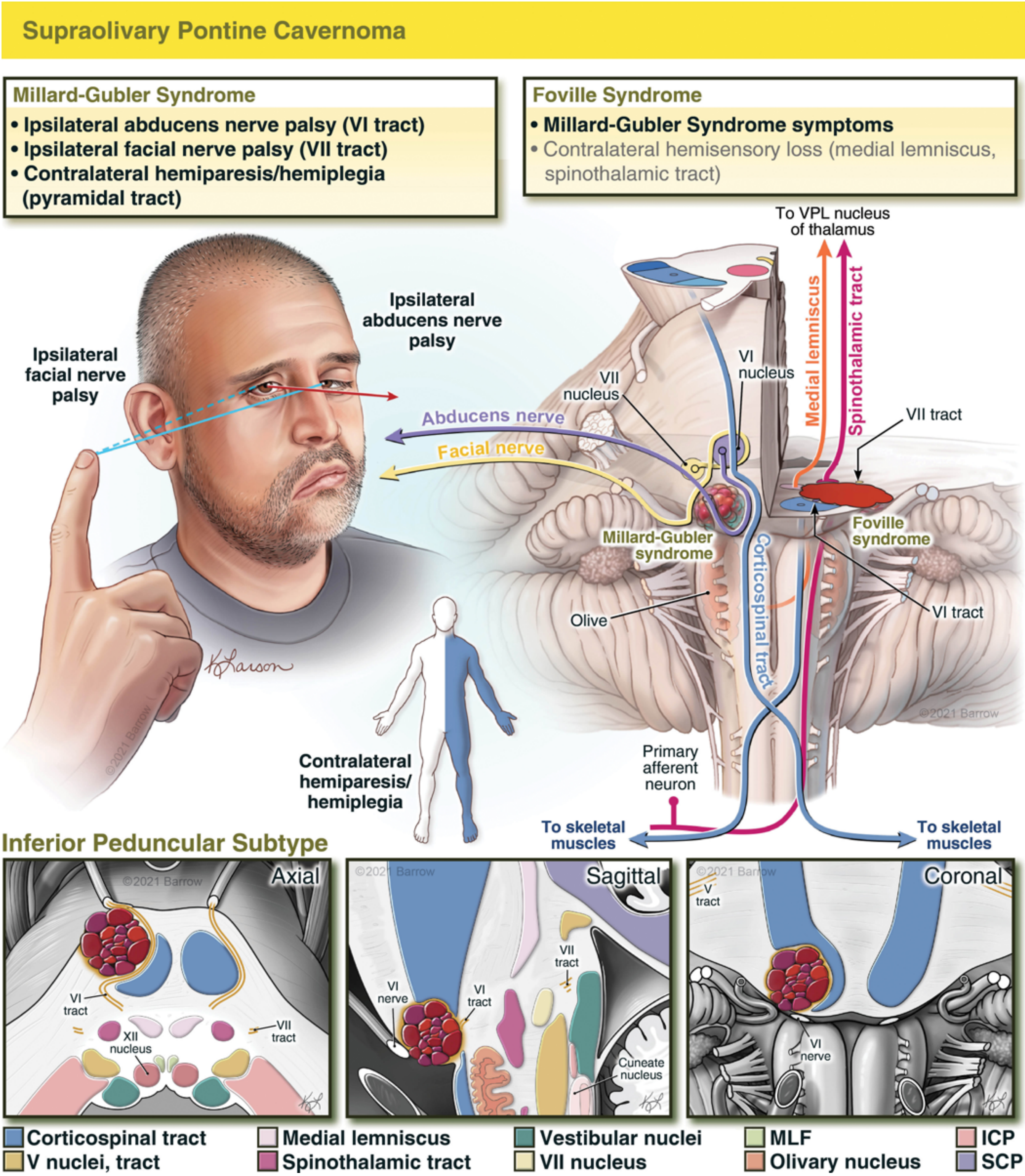

FIG. 8. Patients with supraolivary pontine CMs present with ipsilateral abducens nerve palsy (**upper left inset**). These patients may also have ipsilateral facial palsy and contralateral hemiparesis or hemiplegia due to involvement of the transpontine facial nerve fibers and pyramidal tract (mild Millard-Gubler syndrome) (**middle left inset** and **center panel**). Larger lesions may also cause contralateral hemisensory deficits due to involvement of the medial lemniscus and spinothalamic tracts (Foville syndrome) (**center panel**). This left-sided supraolivary CM resides in the underbelly of the pons adjacent to CN VI, CN VII, corticospinal tract, medial lemniscus, and spinothalamic tract, as shown in axial, sagittal, and coronal views (**lower insets**). *Roman numerals* indicate the CNs. SCP = superior cerebellar peduncle; VPL = ventral posterolateral. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

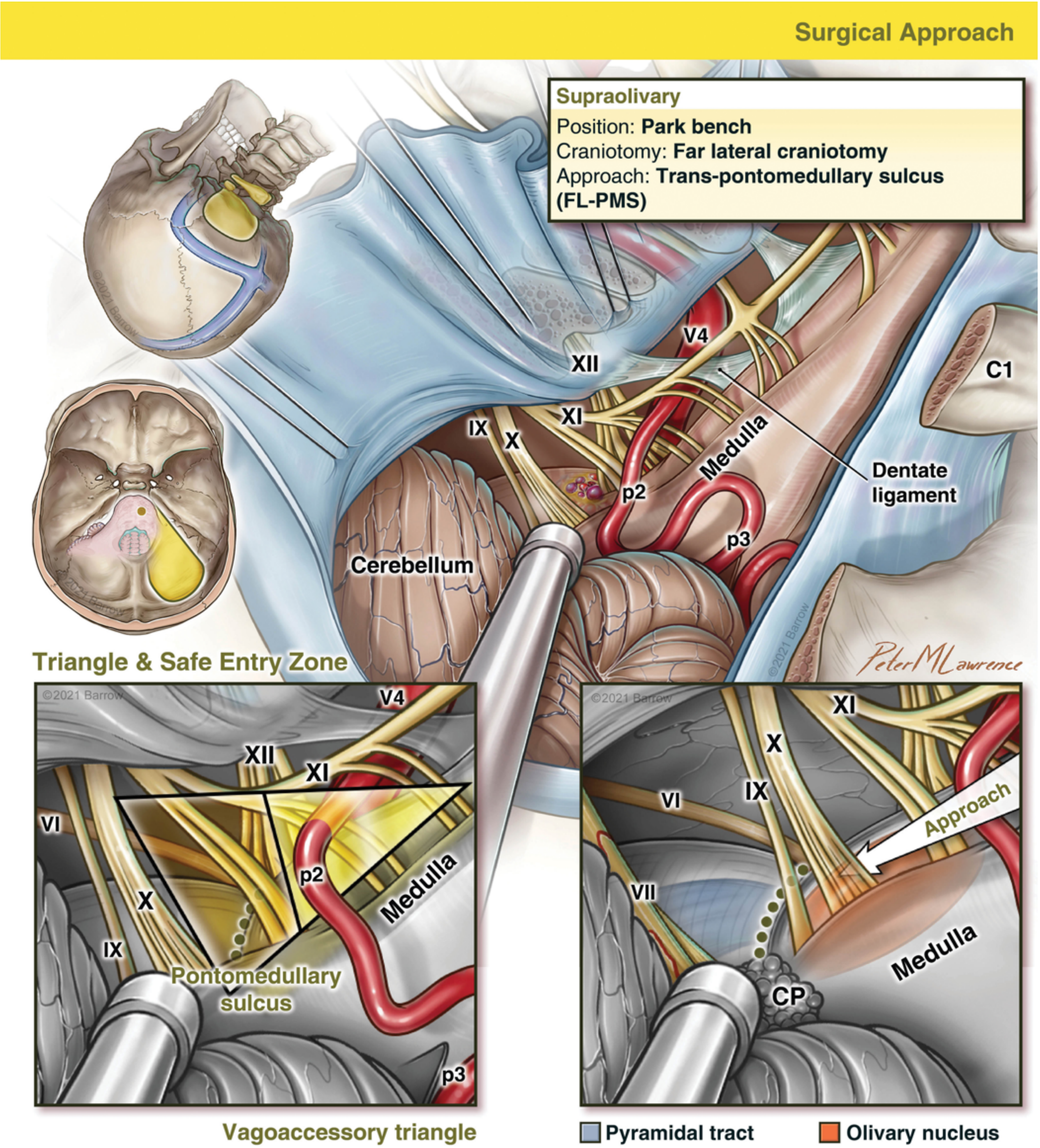

FIG. 9. Supraolivary pontine CMs are exposed with a far lateral craniotomy and transpontomedullary sulcus approach, with the patient in the park bench position to orient the clivus and vertebral artery vertically for viewing along the axis of the anterolateral brainstem (**upper left insets**). Condylectomy opens the cerebellomedullary cistern and exposes the tonsil, lower CNs, V4 of the vertebral artery, p1 (anterior medullary segment) and p2 (lateral medullary segment) of the PICA, and lateral brainstem (**center panel**). Dissection into the suprahypoglossal portion (superior to CN XII) of the vagoaccessory triangle (*yellow*) leads to the olivary nucleus, pontine underbelly, and pontomedullary sulcus (*gray dotted line*) at their intersection (**lower left inset**). Supraolivary lesions surface at the pontomedullary sulcus, lateral to the exit of the abducens nerve. Sulcal entry lateral to the abducens nerve protects the pyramidal tract of the corticospinal tract (*blue*, **lower right inset**). The superior pole of the olive (olivary nucleus ghosted in *orange*, **lower right inset**) meets the inferior pons and forms the pontomedullary sulcus (*gray dotted line*), which is the zone that can be entered safely when a more posterior trajectory is needed to reach a central pontine lesion. *Roman numerals* indicate the CNs. C1 = first cervical vertebra; CP = choroid plexus; V4 = intradural segment of the vertebral artery. Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Lawton, Catapano, Rumalla, Srinivasan. Acquisition of data: Lawton, Catapano, Rumalla, Srinivasan. Analysis and interpretation of data: Lawton, Catapano, Rumalla, Srinivasan. Drafting the article: Lawton, Catapano, Rumalla, Srinivasan. Critically revising the article: Lawton, Catapano, Rumalla, Srinivasan. Reviewed submitted version of manuscript: Catapano, Rumalla, Srinivasan. Statistical analysis: Lawton, Catapano, Rumalla. Administrative/technical/material support: Lawton. Study supervision: Lawton. Medical illustration: Lawrence, Larson Keil.

Supplemental Information

Companion Papers

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