Solid hemangioblastoma in the cerebellopontine angle: Importance of external carotid blood supply with regard to the probable site of origin and preoperative embolization

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Received: 02 June 15   Accepted: 27 November 15   Published: 07 January 16

Abstract

Background: Hemangioblastoma (HBL) is rare in the cerebellopontine angle (CPA) with questionable origin and limited access for circumferential dissection and “en-bloc” excision. We report a case of surgical removal of large solid CPA-HBL and discuss the pattern of blood supply suggesting its origin and indicating preoperative embolization.

Case Description: The solid and highly vascular CPA-HBL had feeders mainly from neuromeningeal division of ascending pharyngeal branch of external carotid artery, suggesting true extra-axial origin. We could achieve “en-bloc” excision without significant blood loss or morbidity using preoperative embolization.

Conclusion: Large solid HBL is rare in CPA necessitating arduous “en-bloc” excision. The pattern of blood supply probably indicates the site of origin and safety of preoperative embolization.

Key Words: Ascending pharyngeal artery, cerebellopontine angle, external carotid artery, hemangioblastoma

INTRODUCTION

Hemangioblastomas (HBLs) of the central nervous system are benign vascular tumors that may occur sporadically or in Von Hippel–Lindau disease. They are composed of stromal and endothelial cell components and account for 1.5–2.5% of all intracranial tumors and 7–12% of posterior fossa tumors.[1,7,14,15] HBLs in the cerebellopontine angle (CPA) are rare with only 11 cases reported in English literature.[2,3,5,8,11-14]

We present a rare case of solid CPA-HBL and discuss the pattern of blood supply with regard to its probable site of origin and safety of preoperative embolization so that the tumor can be excised “en-bloc” following circumferential dissection with minimal blood loss and morbidity.

CASE REPORT

A 45-year-old female with profound left-sided sensorineural hearing loss, left facial palsy (House–Brackmann Grade 3), and left cerebellar signs of 3 months duration with magnetic resonance...
imaging (MRI) showing well enhancing mass in the
left CPA with adjacent nonenhancing cerebellar cyst
[Figure 1], underwent left retromastoid suboccipital
craniectomy and gross total removal of tumor at
local district hospital. Histopathology was that of
HBL. Immediate postoperative period was uneventful
without any added deficits, and she showed gradual
improvement in her neurological status (markedly
in cerebellar signs and mild subjective improvement
in hearing and facial palsy). However, she was lost to
follow-up and presented 10 years later to our institute
with recent onset cerebellar signs and headache,
with persistent left-sided hearing loss and facial
palsy (House–Brackmann Grade 3). MRI showed
recurrent solid densely enhancing tumor in left CPA
[Figure 2]. Digital subtraction angiography of cerebral
vessels showed majority of blood supply (anterior 2/3)
from neuromeningeal division of ascending pharyngeal
branch of external carotid artery (ECA). Posterior 1/3
was supplied by left posterior inferior cerebellar artery
(PICA) and few small dural branches from left vertebral
artery [Figure 3]. Near total (>90%) embolization of
both the feeders was done. Check angiogram showed
very little blush in the tumor region [Figure 4].
Embolization was followed on the next day by “en-bloc
excision of the tumor [Figure 5]. The tumor was dark
red, highly vascular, abutting the lower cranial nerves.
The blood loss was minimal, and no blood transfusion
was required. There was transient worsening of the facial
palsy (House–Brackmann Grade 4) and impairment of
the gag reflex (feed by Ryle’s tube), which improved
to preoperative state by the 9th postoperative day.
The histopathology was typical of HBL. The blood
vessels within the tumor showed presence of glue
material within their lumen. Some of the blood vessels
also contained fibrin thrombi. There was no area
of infarction within the tumor. However, secondary
degenerative changes were seen within the stroma
in form of stromal hyalinization and edema. There
were microcystic changes as well. These changes were
more prominent adjacent to the vessels [Figure 6].
Postoperative contrast MRI confirmed total excision of
the tumor [Figure 7].

**DISCUSSION**

HBLs of the posterior fossa typically arise within the
substance of the cerebellum or the brainstem. However,
unusual HBLs involving the cranial nerves or CPA are
extremely rare with only 11 cases reported so far. CPA HBL
may be either an exophytic component of the
intra-axial HBL or a true CPA mass arising from vessels
around seventh/eighth nerve complex. While the
exact site of origin is debatable, the blood supply may
give some indication. Usually, intra-axial HBL receives
their blood supply through pia mater, from branches
of PICA, anterior inferior cerebellar artery or superior
cerebellar artery, and minimally through branches of
ECA. Ascending pharyngeal artery (APA) is the first

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**Figure 1:** Contrast axial (a) and coronal T2 (b) magnetic
resonance images at initial presentation showing solid enhancing
cerebellopontine angle mass with adjacent nonenhancing cerebellar
cyst

**Figure 2:** T1-weighted (a) and T2-weighted (b) axial magnetic
resonance images at recent presentation. Contrast axial (c) and
coronal (d) magnetic resonance imaging showing solid densely
enhancing tumor in left cerebellopontine angle

**Figure 3:** Preoperative digital subtraction angiography showing
feeders from left posterior inferior cerebellar artery (a), and feeders
from ascending pharyngeal branch of external carotid artery (b)
branch of ECA, dividing further into pharyngeal and neuromeningeal trunks. The neuromeningeal trunk further has hypoglossal and jugular branches supplying meninges near the internal auditory meatus and vasa nervosa of cranial nerves in CPA. The predominant blood supply of CPA-HBL from branches of APA as in our patient has not been reported previously in literature. This may either be due to the true extra-axial origin of HBL from blood vessels of CPA or due to tumor invasion of the dura mater, effect of previous surgery or bleeding over the pia mater which causes adhesion of the tumor to the dura mater.

HBL may either be cystic with a mural nodule or solid. The solid subtypes of HBL have been regarded as difficult to treat surgically because of their arteriovenous malformation-like characteristics. Internal decompression and piecemeal resection may have devastating complications, necessitating circumferential dissection, and “en-bloc” excision. Also, during circumferential dissection, the feeders are carefully ligated to shrink the lesion. Moreover, large solid HBLs in CPA represent an additional challenge because of limited access for en-bloc excision via retromastoid approach and due to complex neurovascular anatomy. Various approaches including retrosigmoid, translabyrinthine, transcondylar fossa, and transcochlear respectively have been used. Conventionally, the threshold to perform embolization in these tumors is high as embolization is contraindicated when the lesion shares its blood supply with the adjacent normal neural tissue. Also, the consequences of postembolization peritumoral edema are significantly higher and potentially life-threatening because of risk of brainstem compression or herniation. As most of the tumor in our patient was supplied by the dural branches of APA originating from ECA, preoperative embolization of the enlarged APA could be performed safely, thereby minimizing intraoperative bleeding in our case. It is further relevant for dural feeders supplying the tumor from the anterior aspect which become accessible only in the later part of surgical exposure.

The main cause of recurrence of HBLs is incomplete tumor resection because of multicentricity or the presence of tumor cells in the cystic wall or the brainstem involvement. Probably for these reasons, regular
radiologic follow-up is necessary to detect the recurred and newly developed lesions, even if the mass was totally removed.

**CONCLUSION**

True solid CPA-HBL is very rare tumor, and the main blood supply is from dural branches of APA. Recognizing this pattern of blood supply preoperatively is helpful in deciding on preoperative embolization so that circumferential dissection and “en-bloc” excision is performed with good hemostasis.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**REFERENCES**


