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A case report

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Cerebello Pontine Angle Hemangiopericytoma - an aggressive tumour needs aggressive management. A case report

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ABSTRACT

Background: CP angle Hemangiopericytoma are rare tumors. Pre-operative suspicion and gross total excision are key for better management due to their aggressive nature.

Case presentation: 18-year-old male presented with signs of progressive brainstem compression. Contrast MRI showed a polycystic enhancing SOL in the right CP angle region compressing the brain stem and cerebellar lobe. Operated with histopathology and IHC indicating it to be a Hemangiopericytoma.

Conclusions: Hemangiopericytoma being an aggressive tumour with a high rate of recurrence compared to other common tumours at CP angle, complete resection with definitive pathological diagnosis and radiotherapy are needed for a better outcome.

BACKGROUND

Hemangiopericytomas (HPCs) constitutes <1% of primary brain tumors. Mostly supra tentorial and dural based. Rare in Cerebello pontine angle (CPA), mimics meningioma radiologically, but because most are grade 2 or grade 3, this aggressive nature makes their pathological differentiation important. [2,3,4,5,6,7,8,9]

We describe here an unusual case of CPA hemangiopericytoma presenting with brainstem and cerebellar signs

CASE PRESENTATION

An 18-year-old male presented with chief complaints of disequilibrium, Progressive weakness all 4 limbs (left>right), slurring of speech, and dysphagia for liquid for 2 months. On admission, his neurological examination revealed a left facial paresis(Hunt & Hess grade 3) with weak gag reflex and uvula deviated to right and asymmetrical palatal arches(left 9th and 10th cranial nerve paresis), and left side weakness

Keywords

hemangiopericytoma,
cerebello pontine angle,
cystic enhancing tumor,
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more than right with increased tone with left cerebellar signs present, suggesting the presence of the Millard-Gubler syndrome.

Head CT scan and an MRI of the brain demonstrated a solid cystic enhancing 2.8×3.5 cm sized right CPA lesion compressing brain stem with no internal acoustic meatus extension (Fig. 1).



Figure 1. (A) Post-contrast magnetic resonance imaging of the brain in coronal views showing a cystic right cerebellopontine angle tumor with dural enhancement (dural tail sign); (B) T2 weighted image showing peritumoral CSF cleft; (C) T1 weighted image showing hyperintense nodule with septations in tumor.

As tumor was solid cystic with enhancement a diagnosis of cystic schwannoma was made with differential being pilocytic astrocytoma and hemangioblastoma, cystic meningioma.

A suboccipital retrosigmoid approach was attempted. During surgery, the tumor was solid and cystic, soft to firm, greyish pink, was suckable highly vascular, capsule was attached to cranial nerves passing in left cp angle with dural attachment superiorly. Tumor was not extending to or coming out from internal auditory canal. It was removed in parts by Cavitron Ultrasonic Surgical Aspirator (CUSA). Somatosensory evoked potentials, brainstem auditory evoked responses, and intraoperative facial nerve monitoring were used to minimise the damage intraoperatively.



Figure 2. Post operative CT of Patient showing decompression of cystic part and a small residual attached to the brain stem.

The patient had CSF discharge from tissue drain more than 100ml on 3rd post op day for which lumbar drain was inserted and tissue drain removed on following day. Surgical wound healed and lumbar drain removed on day 5. No leak or pseudomenigocele formation. Patient discharged on 8th post op day without fresh neurological deficit and improved cerebellar and brain stem signs.

The histopathology sections revealed moderately cellular neoplasm with closely opposed cells with round nuclei arranged in a haphazard pattern with limited intervening stroma. Nuclei are oval to spindle with dense chromatin and scanty cytoplasm. There was mild nuclear atypia with mitotic activity (<5/HPF). Cyst formation also seen. There was numerous slit like vascular spaces. Vimentin CD34 and CD99 are positive with STAT-6 strongly nuclear positive. Ki-67 is less than 5%. Immunomarkers favoured a grade 2 hemangiopericytoma. While negative for EMATLE, SOX10. (Fig. 3, Fig. 4, Fig.5, Fig.6).

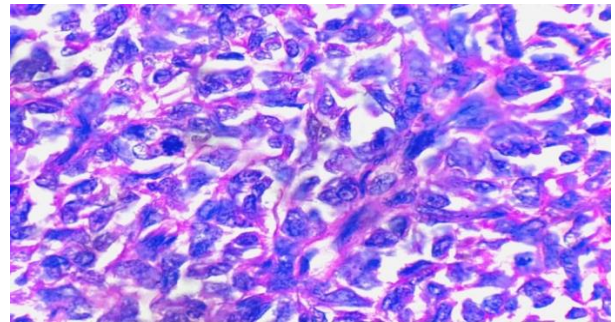


Figure 3. Photomicrographs of the tumor specimens showing. A: Diffuse sheets of relatively uniform population of cells interspersed by staghorn vascular channels (H&E, original magnification, ×10). B: Round to oval cells with finely dispersed nuclear chromatin and moderate cytoplasm and no signs of anaplasia (H&E, original magnification, ×20). H&E, haematoxylin and eosin.

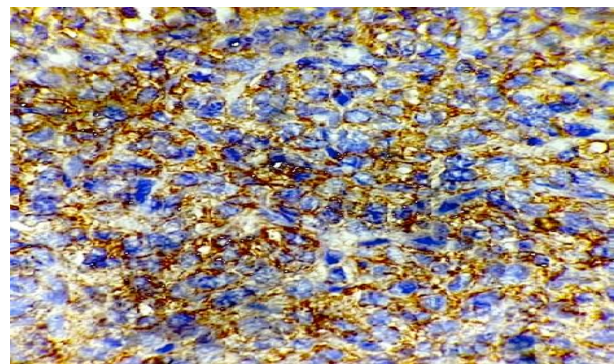


Figure 4. Immunohistochemical staining showing diffuse positivity with CD34 (original magnification, ×100).

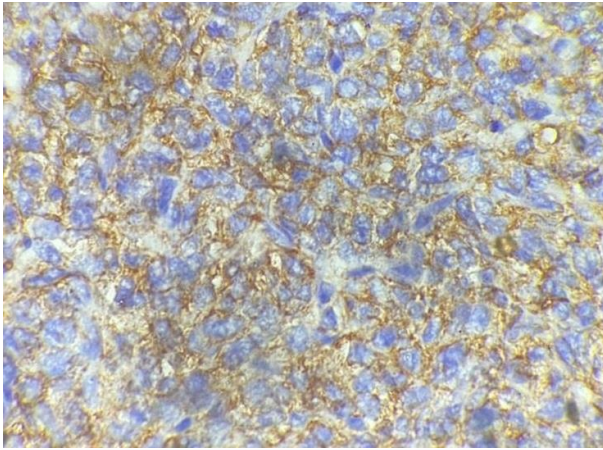


Figure 5. CD 99 STAINING (oil immersion; original magnification $\times 1000$).

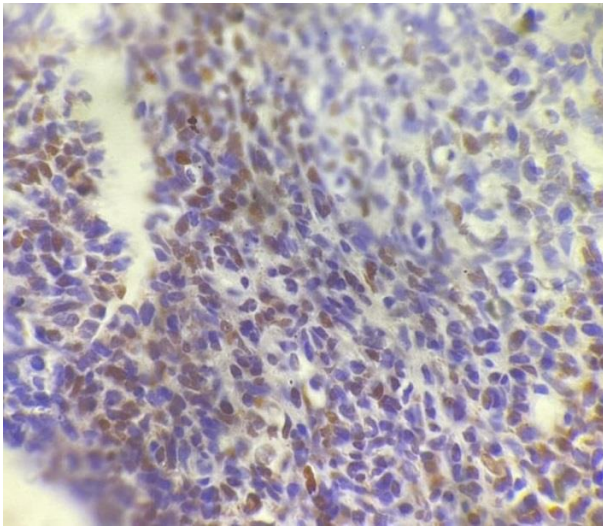


Figure 6. STAT 6 STAINING (original magnification $\times 100$)

Metastatic workup including CT of the chest/abdomen/pelvis did not reveal any evidence of extracranial HPC. Adjunctive radiation therapy was advised but he was lost to follow up since a month post-surgery.

DISCUSSION

HPCs are rare, and are aggressive neoplasms that arise from the pericytes of Zimmerman, which are contractile spindle cells surrounding capillaries and post capillary venules [4]; and most often involve the musculoskeletal system and skin [1]. Intracranial HPCs constitute just 0.4% of all intracranial tumors [10] and approximately 2% to 4% of all meningeal tumors [5].

Immunohistochemistry has major role in diagnosis of HPC. It also helps in differentiation

between meningioma and HPC, where HPC is positive with CD34, CD99 and STAT-6 [5].

In the present case, the morphological features were distinctive enough to place the lesion in the category of an HPC. Fairly uniform cellularity of the tumor and the high mitotic activity was more supportive of this lesion being labelled as grade II HPC instead of a cellular SFT

The similarity between meningioma and HPC is limited to radiology and gross morphology. In fact, with a mean survival of 84 months from the time of initial diagnosis [5], a local recurrence rate as high as 91% and a 15-year risk of distant metastasis approaching 70%; intracranial HPCs harbour one of the most aggressive biological/clinical behaviours [16].

In present scenario gross total resection followed by radiotherapy is standard of care.

Radiation therapy has in fact extended the mean time of local recurrence from 34 to 75 months, and the survival from 62 to 92 months [1].

CONCLUSIONS

In conclusion, HPC being aggressive tumor with high rate of recurrence and metastasis, it should be included as a differential diagnosis in dural Based extra axial CPA tumors in age-appropriate cases.

A high index of suspicion on radiology imaging is essential to plan for complete excision, and role accurate histopathological diagnosis can't be overemphasised. As postoperative recurrence seems unavoidable, long-term follow-up with serial imaging should be considered in all cases.

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