Case Report

rare tumors

Rare Tumors
Volume 15: 1–7

DOI: 10.1177/20363613221150218
journals.sagepub.com/home/rtu
SAGE

Posterior fossa giant adenoid cystic carcinoma with skull base invasion mimicking glomus jugulare: A case report and review of literature

Anand Kumar Das, MCh Neurosurgery[1], Saraj Kumar Singh, MCh Neurosurgery[1], Kranti Bhavana, MS Otolaryngology[2] and Subhash Kumar, MD Radiodiagnosis[3]

Abstract

The author describes a rare case of giant adenoid cystic carcinoma (ACC) mimicking large paraganglioma with lower cranial nerve palsy. A 60-year-old female presented with a progressive increase in postauricular swelling with unilateral hearing loss, facial deviation, difficulty in swallowing, and hoarseness of voice. MRI brain showed highly vascular infiltrating and osteolytic mass suggestive of large glomus jugulare versus sarcoma. It was completely engulfing the jugular foramen and lower cranial nerves with bony erosion of the jugular foramen and occipital condyle. The whole mastoid was filled with the tumor. On digital subtraction angiography the majority of blood supply was from the occipital branch of the external carotid artery and vertebral artery. The patient underwent percutaneous embolization followed by external carotid ligation and resection of the mass. The postoperative course was uneventful. Histopathology was suggestive of mixed ACCs. The patient received radiotherapy. After 1 year of follow up no recurrence or distant metastasis was noted.

Keywords

Giant adenoid cystic carcinoma, glomus jugulare, lower cranial nerve palsy, embolization

Introduction

Adenoid cystic carcinoma (ACC) is one of the most common salivary gland tumors affecting both major and minor salivary glands. It accounts for 1% of all head and neck malignancies.[1] The majority of them affect minor salivary glands followed by parotid.[2,3] Other less frequent sites of occurrence are the orbit, sinonasal cavity, and oral cavity.[1,4] These tumors affect mainly women in their fifth decades. The typical characteristics of ACCs are moderate growth rates, high rates of distant metastasis, and a high propensity for perineural dissemination.[5] The majority of documented cases of ACC at the base of the skull originated from adjacent structures such as the salivary gland, lacrimal gland, paranasal sinus, or nasopharynx.[6-8] This is the first case in literature where ACC presented as a posterior and middle fossa tumor mimicking large glomus jugulare with clinical involvement of all lower cranial nerves along with the seventh and eighth cranial nerve. It was treated with a multidisciplinary approach involving percutaneous embolization, radical resection, and postoperative radiotherapy.

Case Report

A 60-year-old female presented with retro-auricular swelling on the left side, unilateral hearing loss with

[1]Neurosurgery, All India Institute of Medical Sciences, Patna, Bihar, India
[2]Otolaryngology, All India Institute of Medical Sciences, Patna, Bihar, India
[3]Radiodiagnosis, All India Institute of Medical Sciences, Patna, Bihar, India

Corresponding author:
Saraj Kumar Singh, Neurosurgery, All India Institute of Medical Sciences, Phulwari Sharif, Patna, Bihar 801507, India.
Email: dr.sarajkumarsingh@gmail.com

tinnitus, and blood discharge from the left ear for the past 6 months. She also had difficulty swallowing solid food with hoarseness of voice for the past 4 months. She had a deviation of the face towards the right side, with incomplete closure of the left eye (House-Brackmann grade 4). On examination, left-sided 7th to 12th nerve palsy was present. The tongue deviated to the left side on protrusion. The cough and gag reflex was impaired. Taste sensation was affected on the left side of the tongue. Local examination revealed a large lobulated irregular mass over the left postauricular region and occluded the external auditory canal with lateral displacement of the pinna. It was warm on palpation without any bruit or pulsation. MRI brain showed a mass in the left side of the posterior fossa which was hypointense on T1, hyperintense on T2 with multiple flow voids. On contrast injection, the lesion showed homogenous enhancement. The tumor was extending anteriorly from the middle ear cavity to posteriorly to the anterolateral aspect of the cerebellum. Antero-medially it was abutting the internal carotid artery (ICA). It was completely engulfing the jugular foramen and lower cranial nerves with bony erosion of the jugular foramen and occipital condyle. The whole mastoid was filled with the tumor. The tumor plane was undisturbed with the brainstem. CT brain revealed complete disappearance of bony landmarks from the left suboccipital bone, mastoid, and foramen magnum (Figure 1 a-f). Chest and abdominal evaluation suggested no evidence of distant metastasis. On digital subtraction angiogram, the tumor was hypervascular. The majority of blood supply was from the occipital branch of the external carotid artery (ECA) and vertebral artery. The left side of the internal jugular vein was completely blocked. The patient was subjected to percutaneous embolization with N-butyl cyanoacrylate glue. The posterior and inferior part of the tumor was embolized after complete blockage of the occipital artery. Only the anterior aspect of the tumor was showing mild blush as it had blood supply directly from ECA (Figure 2 a-f). After 24 h of embolization, the patient got operated. First neck exploration was done and the left ECA was ligated as the majority of the blood supply of the tumor was from ECA. The internal carotid artery (ICA) control was taken and the tumor was attacked. Lazy S skin incision was given over mass in the retroauricular region and whole tumor debulking was done from the cerebellum to ICA, medially up to the

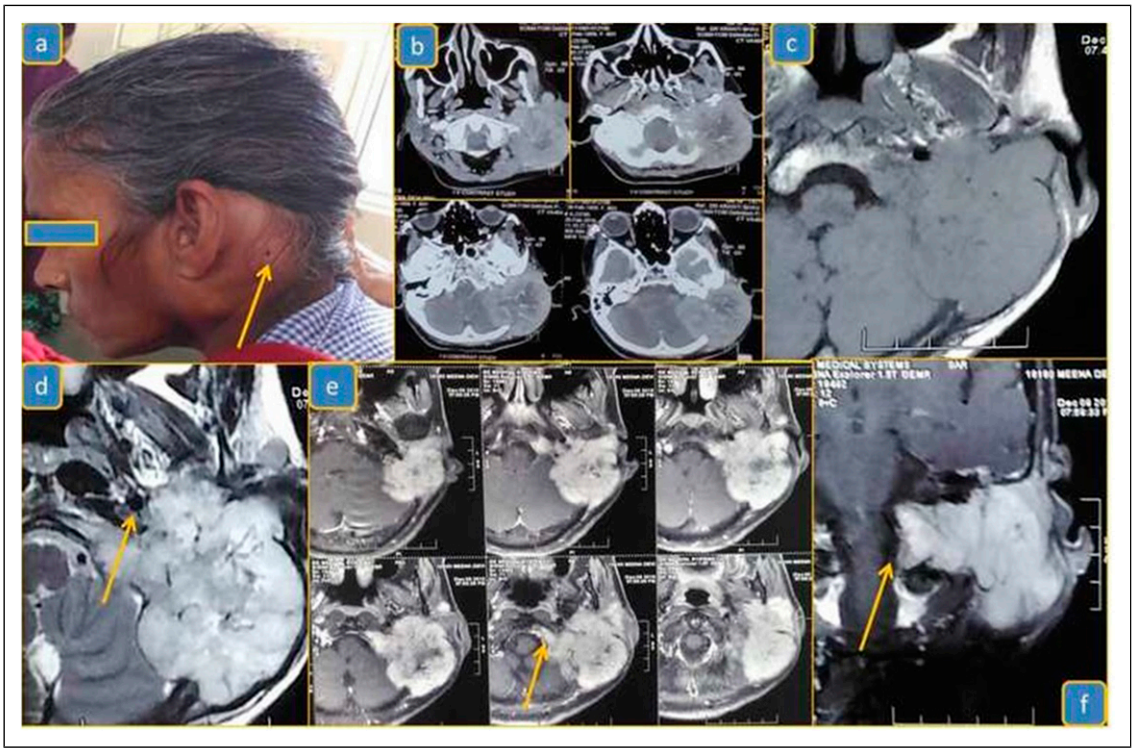

Figure 1. (a) Patient showing large retroauricular mass with (arrow mark) black dot suggestive of puncture site for glue, (b) CT head Axial view showing large herniating mass with the destruction of, jugular foramen, mastoid, and lateral suboccipital region, (c) T1 Sequence MRI (Axial) showing large hypointense mass occupying both middle and posterior cranial fossa abutting internal carotid artery, (d) T 2 sequence (Axial) showing large hyperintense mass with multiple flow voids (Arrow mark showing internal carotid artery), (e) T1 contrast sequence (Axial) showing large homogenously enhancing mass at retromastoid region extending anteromedially up to ventral brain stem (arrow mark), (f) T1 contrast sequence (coronal) showing large homogenously enhancing mass at retromastoid region (arrow mark) with preserved plane from the brain stem.

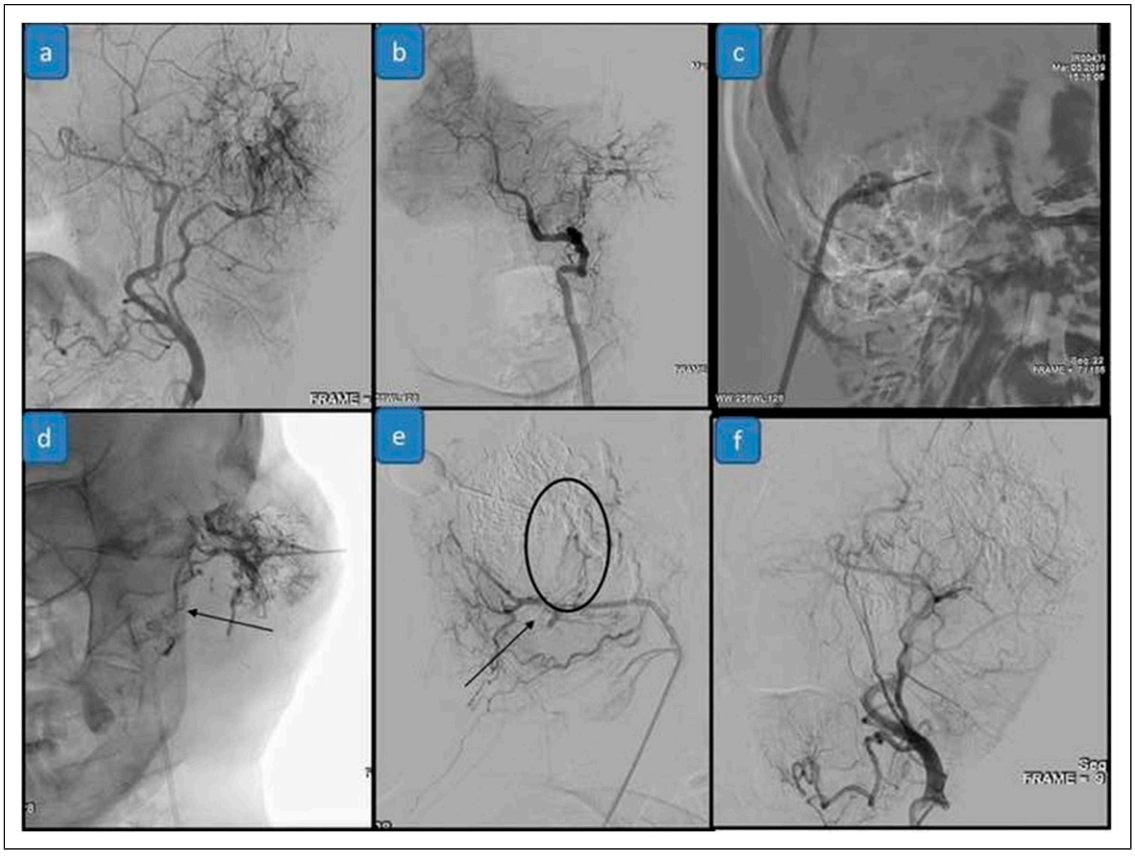

Figure 2. (a) Left ECA injection oblique view, showing hypervascular tumor with large intense tumor blush and multiple enlarged sinusoidal spaces within the tumor core, (b) LVA injection, AP view showing the core of the lesion which is made up of large communicating sinusoidal spaces, (c) LAT view, fluorospot image, direct needle puncture done under roadmap with an attempt to access the deep-seated sinusoidal spaces first, (d & e) Glue cast, Occipital artery angiogram, Lat view, showing no further filling and LVA angiogram, AP view, after Second percutaneous injection of nBCA, shows no further filling from LVA injection, and <10% filling from occipital artery injection (oval); the LVA is no more filling on the occipital artery injection as the muscular collaterals have been retrogradely filled with glue (thin arrow), (f) Final check angiogram ECA injection AP view, showing near total devascularization with a small area of fine blush in the anteromedial portion of the tumor.

occipital condyle and internal jugular vein. The tumor was infiltrating lower cranial nerves which got resected along it. The tumor was invading the middle ear cavity which was excised and the eustachian tube was packed with complete obliteration of the left ear. The tumor was also invading the petrous aspect of the cerebellum with the involvement of the dura. The affected dura was excised and augmented duraplasty was done. The operative cavity was packed with fat and glue. There were 600 mL of blood loss during the surgery. The postoperative course was uneventful. Postoperative NCCT head showed near total resection of the tumor. The overlying skin and incision site were healthy (Figure 3 a & b). Histopathology was suggestive of a high-grade (grade III) tumor with irregular branching sheets and nodules. Tumor cells were present in the solid and cribriform pattern. Two types of cells were present, epithelial and myoepithelial cells. The stroma was hyalinized with myxoid features. On immunohistochemistry, it was positive for CK5/6, CK7, CK117, and EMA. S-100 and SMA were positive in myoepithelial cells. Histopathology was suggestive of mixed ACCs (Figure 4 a-d). The patient received radiotherapy. After one year of follow-up, the patient was examined. There was no improvement in lower cranial nerve paresis and no evidence of distant metastasis. However, the radiation-induced cutaneous reaction occurred over the incision site and surrounding area (both pigmentation and ulceration) (Figure 5 a). Topical steroids were given to ameliorate further progress of skin damage.

Discussion

ACC can arise in any salivary gland tissue and within the head and neck - the major salivary glands represent the most common site, followed by tumors originating in the sinonasal tract.[9] Giant ACC with osseous invasion has been rarely reported. This is the first case report of giant ACC

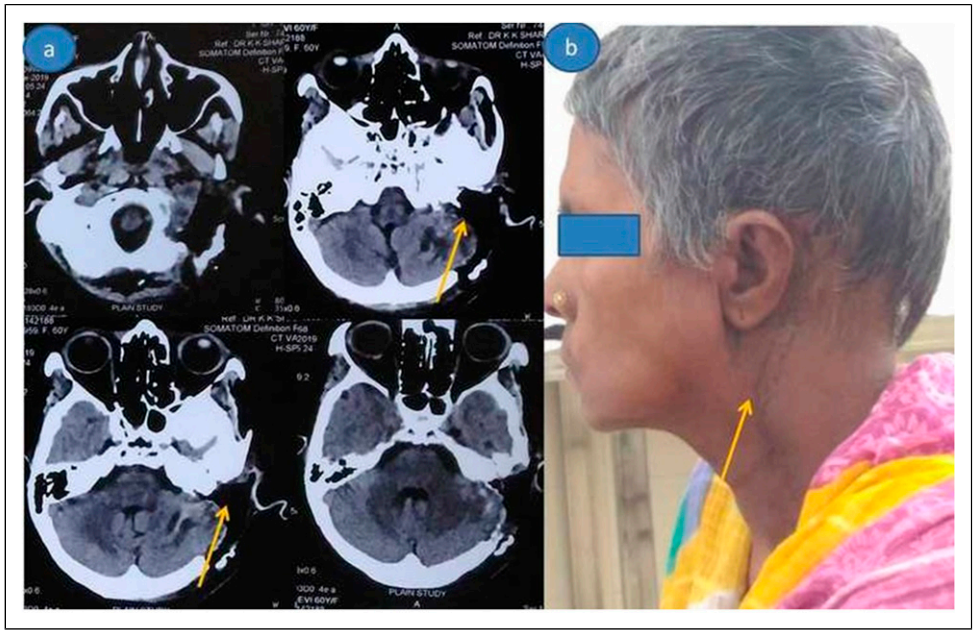

Figure 3. (a) Post-op NCCT head showing near total resection of mass both at intracranial and extracranial region (Arrow mark), (b) Post-op image of the patient after 2 months showing healthy scar mark (arrow mark) and overlying skin.

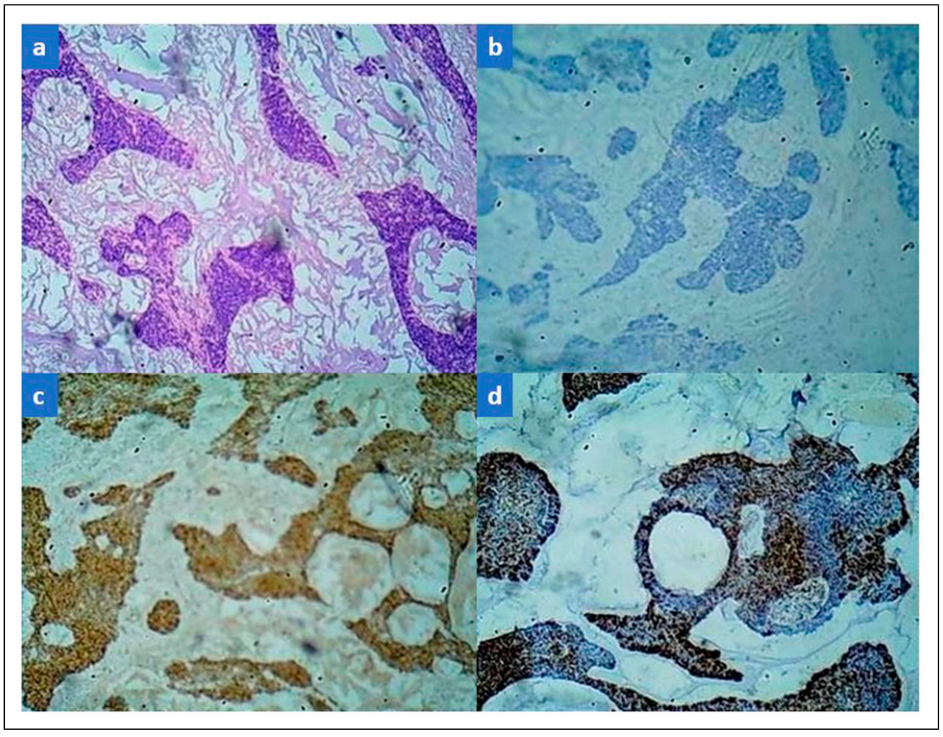

Figure 4. (a) Histopathology suggested mixed adenoid cystic carcinoma with both solid and cribriform components, but solid components are predominant (H&E, 100X), (b) IHC demonstrated GFAP negative tumor cells (100X), (c) S100 positive on IHC showing ductal component (100X), (d) p63 positive on IHC showing myoepithelial component (100X).

Figure 5. (a) Image of patient's postauricular region showing radiation-induced cutaneous reaction (Pigmentation [yellow arrow] and ulceration[green arrow]).

with jugular foramen destruction mimicking large glomus jugulare in location as well as clinical presentation. Intraosseous occurrence of ACC is rarely reported.[2,3,8] ACCs occur mainly in middle-aged females however, intraosseous lesions can occur at any age (24–82 years) without any gender predilection.[10,11] Most commonly these originate from the posterior part of the mandible followed by the maxilla. However, in the present case, both the mandible and maxilla were intact. But there was complete destruction of the suboccipital bone, foramen magnum, lateral aspect of the occipital condyle, and mastoid bone. ACCs usually present with a gradual onset, slow progression, sneaky destruction of the nearby structures, perineural invasion, and distant metastasis.[12] Local invasion is the most prevalent type of tumor growth.[13] The tumor does not become apparent until a significant amount of the soft tissues, bones, or cranial nerves have been affected. Clinical features include pain, swelling, and numbness with paresthesia.[14,15] In the present case, the patient presented with a slow-growing mass of the left mastoid and suboccipital region with facial deviation and difficulty in swallowing. A metastatic workup was performed because distant metastasis usually occurs and most commonly affects the lung, bones, and liver.[14,16] It showed local bony destruction without any regional lymph node involvement or distant metastasis. Surgical management includes radical resection with wide macroscopically free margins even including the non-eloquent normal areas of brain parenchyma. Surgery is exceedingly difficult and risky because of the anatomy of the skull base and the presence of numerous vital structures, especially in patients with locally advanced recurring tumors.[17-19] Resection should be followed by radiotherapy even if the tumor borders are free of malignancy. Overall, the role of chemotherapy (carboplatin and paclitaxel) is uncertain because studies yield contradictory results regarding overall survival.[13,14]

Typically, tumors with extension from the nasopharynx, paranasal sinuses, and maxilla carry a poor prognosis.[10] The present case was not having any involvement in the above areas. However, histopathology suggested it to be a high-grade tumor. Therefore, radical resection was planned after embolization. Embolization of hypervascular tumors helps to reduce blood loss, and operative time and aid in maximal tumor resection.[20,21] The following elements are deemed crucial to prognosis: location, size, extension into neighboring structures, perineural invasion, and a histological grade.[12,13]

Bone destruction with perineural invasion was present in this case, favoring a poor prognosis. The radiographic margins of the ACC are significantly overrun by tumor cells, and this tumor not only exhibits perineural invasion but also perineural dissemination. As a result, a radical resection with wide margins and postoperative radiation therapy of 60–75 Gy is often required.[22] Considered by many authors to be an optimal treatment for ACC is radical surgical resection followed by radiation. However, the frequent perineural and perivascular intrusions along the internal carotid artery and lower cranial nerves render the surgical approach unsafe in situations with nasopharyngeal and posterior fossa extension. Due to this proximity, it is difficult to create surgically negative margins. Because of this, surgical procedures frequently result in partial excision, which increases the likelihood of local recurrence.[9,23] Neurosurgeons should keep the patient disability-free if complete resection of the tumor is not possible. However, in patients with established disabilities, the target should be complete resection without leaving any metastatic lymph nodes and without adding any new deficits. A well-established prognostic profile exists for ACCs. A substantial risk of distant metastasis occurs even in ACC cases where the tumor is entirely excised. Although in this case follow-up period was short but no local recurrence or metastasis was noted in 1 year. The overall survival rate at 5 years is 75%, while the 10-years survival rate is 20% and the 15-years survival rate is approximately 10%.[5] The overall prognosis depends on a

number of variables. Tubular, cribriform, and solid ACC are the three conventional categories, with the solid subtype having the worst prognosis. It's not uncommon to see all three patterns coexisting. A tubular pattern with no solid part is categorized as grade I. The presence of a cribriform component with at least 30% solid form is classed as grade II and a cribriform component with more than 30% solid pattern is categorized as grade III.[24] In our case, the lesion was mixed with a majority having solid portions (grade III), which has a poor prognosis. In patients with tumors larger than 4 cm in size, the prognosis is poorer since it indicates a more extensive subclinical spread. The prognosis is also affected by a delay in diagnosis and treatment.[25] If surgical margins are not clear; the tumor is associated with a poor prognosis despite the postoperative radiation therapy. It is one of the most important factors associated with local recurrence.[26] As per the literature review, this is the first case of ACCs in terms of involvement of posterior cranial fossa mimicking glomus jugulare with bony erosion and breaching of posterior petrous dura. ACCs involving the 7th and 8th nerves along with lower cranial nerve involvement ($9–12^{th}$) have not been reported till now in the literature. Moreover, successful management utilizing percutaneous embolization followed by radical resection lays down a treatment solution for these types of patients.

Conclusion

This case is a brilliant example of a giant ACC creating a diagnostic dilemma for a posterior fossa tumor. Giant ACCs should be managed meticulously but aggressively with a multidisciplinary approach after proper planning and investigations.

Acknowledgements

We would like to thank Dr. Yasmin for her assistance in the study.

Authors' Contribution

Saraj Kumar Singh contributed to the study conception. Material preparation and data collection were performed by Anand Kumar Das, Saraj Kumar Singh, Kranti Bhavana and Subhash Kumar. The first draft of the manuscript was written by Anand Kumar Das and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Informed Consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

Ethical approval

AIIMS, Patna does not require ethical approval for reporting individual cases or case series.

ORCID iD

Saraj Kumar Singh https://orcid.org/0000-0003-3156-7096

References

1. Pahwa S, Sharma A, Kamboj M, et al. Nasopharyngeal adenoid cystic carcinoma with intracranial spread: A rare presentation. *Indian J Pathol Microbiol*. 2022;65(1):200–202.
2. Capodiferro S, Scully C, Macaita MG, et al. Bilateral intraosseous adenoid cystic carcinoma of the mandible: report of a case with lung metastases at first clinical presentation. *Oral Dis*. 2005;11(2):109–112.
3. Indu S and Roy ID. Intraosseous adenoid cystic carcinoma of the mandible: A rare presentation. *Journal of Oral and Maxillofacial Pathology: JOMFP* 2020;24(Suppl 1):S37.
4. Macri C, Juniat V, Davis G, et al. Intraorbital and intracranial extension of adenoid cystic carcinoma without clinical or radiological lacrimal gland involvement. *Orbit*. 2021:41, 1–5.
5. Gadkaree SK, Parikh AS, Rodarte AI, et al. Adenoid Cystic Carcinoma of the Skull Base: Response to Radiation Therapy and Outcomes in a Retrospective Case Series. *J Neurol Surg B Skull Base*. 2020;81(5):505–510.
6. Chen YK, Chen CH, Lin CC, et al. Central adenoid cystic carcinoma of the mandible manifesting as an endodontic lesion. *International Endodontic Journal*. 2004;37(10): 711–716.
7. Favia G, Maiorano E, Orsini G, et al. Central (intraosseous) adenoid cystic carcinoma of the mandible: report of a case with periapical involvement. *Journal of Endodontics*. 2000; 26(12):760–763.
8. Li Y, Li LJ, Huang J, et al. Central malignant salivary gland tumors of the jaw: retrospective clinical analysis of 22 cases. *Journal of Oral and Maxillofacial Surgery*. 2008;66(11): 2247–2253.
9. Guazzo E, Bowman J, Porceddu S, et al. Advanced adenoid cystic carcinoma of the skull base - The role of surgery. *Oral Oncol*. 2019;99:104466.
10. Mahajan A, Kulkarni M, Parekh M, et al. Adenoid cystic carcinoma of hard palate: A case report. *Oral Maxillofac Pathol J*. 2011;2(1):127–131.
11. Al-Sukhun J, Lindqvist C, Hietanen J, et al. Central adenoid cystic carcinoma of the mandible: case report and literature review of 16 cases. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*. 2006;101(3): 304–308.

12. Orhan K, Yuksel Y and Gorur D. Solid adenoid cystic carcinoma of maxilla: A Case report. *Clinical Dentistry and Research*. 2006;30:42–47.
13. Gadkaree SK, Parikh AS, Barbarite E, et al. Adenoid Cystic Carcinoma of the Nasopharynx with Skull Base Involvement: Retrospective National Cohort Analysis of Treatment Paradigms, Outcomes, and Provider Density. *J Neurol Surg B Skull Base*. 2020;83(1):99–104.
14. Ramakrishna R, Raza SM, Kupferman M, et al. Adenoid cystic carcinoma of the skull base: results with an aggressive multidisciplinary approach. *J Neurosurg*. 2016;124(1):115–121.
15. Singh SK and Agarwal H. Supratentorial extension of large cerebellar pilocytic astrocytoma in adult. *Br J Neurosurg*. 2020;34(5):595. DOI: 10.1080/02688697.2018.1438587.
16. Singh SK and Kumar N. Extraaxial Cerebellopontine Angle Medulloblastoma with Multiple Intracranial Metastases in Adult: A Rare Case Report with a Review of Literature. *Asian J Neurosurg*. 2020;15(3):695–698.
17. Xu N, Zheng L, Wu WJ, et al. Definitive ^{125}I Brachytherapy of Locally Advanced Adenoid Cystic Carcinoma Involving the Skull Base With Satisfying Efficacy and Safety. *J Oral Maxillofac Surg*. 2019;77(10):2143–2153.
18. Das AK, Mani SK, Singh SK, et al. High-definition two-dimension video telescope operating monitor-assisted brain and spinal surgery in pediatrics: is it an acceptable substitute for microscopic surgery? *Childs Nerv Syst*. 2022;38(11):2171–2177.
19. Satyarthee GD and Singh SK. Management of Giant Vestibular Schwannoma in Second Trimester Pregnancy–Review of Literature. *Am J Clin Neurol Neurosurg*. 2015;1(3):142–146.
20. Das AK, Mani SK, Singh SK, et al. Management and outcome of unusual pediatric brain tumors: challenges experienced at a tertiary care center of a developing country. *Childs Nerv Syst* 2022, DOI: 10.1007/s00381-022-05694-2.
21. Das AK, Mani SK, Singh SK, et al. Giant proliferative spindle cell neoplasm of scalp. *Acta Neurol Belg* 2022, DOI: 10.1007/s13760-022-02129-2.
22. Nangia S, Gaikwad U, Noufal MP, et al. Proton therapy for skull-base adenoid cystic carcinomas: A case series and review of literature. *J Cancer Res Ther*. 2022;18(3):629–637.
23. Mendenhall WM, Dagan R, Bryant CM, et al. Definitive Radiotherapy for Skin and Adenoid Cystic Carcinoma with Perineural Invasion. *J Neurol Surg B Skull Base*. 2016;77(2):169–172.
24. Volpi L, Bignami M, Lepera D, et al. Endoscopic endonasal resection of adenoid cystic carcinoma of the sinonasal tract and skull base. *Laryngoscope*. 2019;129(5):1071–1077.
25. Guazzo E and Panizza B. Management of advanced adenoid cystic carcinoma infiltrating the skull base: a contemporary review. *J Neurooncol*. 2020;150(3):419–427.
26. Pushpanjali M, Sujata DN, Subramanyam SB, et al. Adenoid cystic carcinoma: An unusual presentation. *Journal of Oral and Maxillofacial Pathology: JOMFP* 2014;18(2):286.

Appendix

Abbreviations

MRI	Magnetic Resonance Imaging
ACC	Adenoid Cystic carcinoma
ECA	external carotid artery
ICA	internal carotid artery
DSA	digital substraction angiogram