Giant Adenoid Cystic Carcinoma With intracranial invasion: Unique Tumor At Posterior Cranial Fossa

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Abstract

Giant ACC with intracranial extension and lower cranial nerve palsy is rarest. 60 year female presented with unilateral hearing loss, facial deviation and difficulty in swallowing. Imaging showed highly vascular infiltrating and osteolytic mass S/O glomus jugulare. Patient underwent percutaneous embolization followed by External carotid ligation and resection of mass.

Introduction

Adenoid cystic carcinoma is one of the most common salivary gland tumors. It accounts for 6-10% of all salivary gland tumors. Majority of them affects minor salivary glands followed by parotid [4, 9]. These tumors affect mainly women in their fifth decades. Rarely does it cause bone destruction and most of the time mandible is involved more than maxilla [5, 6, 7]. This is the first case in literature where adenoid cystic carcinoma presented as posterior and middle fossa tumor like large glomus jugulare or tympanicum with clinical involvement of all lower cranial nerves along with seventh and eighth cranial nerve. It caused destruction of Jugular foramen, lateral suboccipital bone along with foramen magnum. It was treated with multidisciplinary approach involving percutaneous embolization, radical resection and post-operative radiotherapy.

Case Report

A 60 years old female patient presented with post auricular swelling on left side for past 6 month. It was extending from upper part of neck to sub occipital region displacing the ear

laterally. Patient was complaining of unilateral hearing loss with tinnitus with blood discharge from left ear. There was difficulty in swallowing of solid food with hoarseness of voice for past 4 months. She was also having unilateral deviation of face with incomplete closure of left eye (H & B grade 4). On examination, she was having left sided 7 th to 12 th nerve palsy. Her tongue was deviating to left side with impaired gag and cough. Taste sensation was affected on left side

of tongue. Local examination revealed large lobulated irregular mass over left post auricular

region and occluding the left external auditory canal with lateral displacement of the pinna. It

was warm on palpation without any bruit or pulsation. MRI brain was done. It was Hypointense on T1, hyperintense on T2 with multiple flow voids just abutting the internal carotid artery. On contrast injection it was showing homogenous enhancement. Tumor was extending anteriorly from middle ear cavity to posteriorly till antero lateral aspect of cerebellum. Antero-medially it was abutting internal carotid artery (ICA). It was destroying jugular foramen, occipital condyle, completely engulfing jugular foramen and lower cranial nerves. The whole mastoid was filled with tumor only. Tumor plane was undisturbed with brain stem. CT brain was showing complete disappearance of bony landmarks from left suboccipital bone, mastoid

and foramen magnum. Chest and abdominal evaluation suggested no evidence of metastasis. (Fig.1.a-f) On Digital Substraction angiogram, Tumor was hypervascular. It was getting majority of blood supply from Occipital branch of External carotid artery and vertebral artery. Left side of internal jugular vein was completely blocked. Patient was subjected for percutaneous embolization with N-butyl cyanoacrylate glue. Posterior and inferior part of tumor was embolized after complete blockage of occipital artery. Only anterior aspect of tumor was showing mild blush as it was getting blood supply directly from ECA. (Fig.2.a-f) After 24 hours of embolization patient got operated. First neck exploration was done and external carotid artery (ECA) was ligated as majority of blood supply of tumor was

from ECA. Than internal carotid artery (ICA) control was taken and tumor attacked. Whole tumor debulking was done from cerebellum to ICA, medially up to occipital condyle and internal jugular vein. Tumor was infiltrating lower cranial nerves which got resected along it. Tumor was invading middle ear cavity which was excised and Eustachian tube was packed with complete obliteration of left ear. Tumor was also invading petrous aspect of cerebellum with involvement of dura. Whole affected dura was excised and augmented duraplasty done. Operative cavity was packed with fat and glue. There was 600 ml blood loss. Post-operative course was uneventful. Post-operative NCCT head was showing near total resection of tumor. (Fig.3.a) overlying skin and incision site was healthy after 2 month. (Fig.3.b) Patient received radiotherapy after 2 month. On follow up after one year, patient was examined. There was no improvement in lower cranial nerve paresis. However radiation induced cutaneous reaction occurred over incision site and surrounding area (both pigmentation and ulceration). (fig.4.a) Topical steroids were given to ameliorate further progress of skin damage. Histopathology was suggestive of high grade tumor in irregular branching sheets and nodules. Tumor cells are present in solid and cribriform pattern. Two types of cells are present, epithelial and myoepithelial cells. The stroma is hyalinised with myxoid features. On IHC it was positive for CK5/6, CK7. CK117 and EMA. S-100 and SMA was positive in myoepithelial cells. Histopathology was more likely suggestive of mixed ACCs. (Fig.5.a-d)

Discussion

ACC is most common tumor affecting minor salivary glands. Giant ACC with osseous invasion has been rarely reported. This is the first case report of giant ACC with jugular foramen destruction mimicking large glomus jugulare in location and clinical presentation. Intra-osseous occurrence of ACC is rarely reported. [4, 7, 9] ACCs occur mainly in middle aged females however, intra-osseous lesions can occur at any age (24-82 years) without any gender predilection. [1, 2] Most commonly these originate from posterior part of mandible followed by maxilla. However in present case both mandible and maxilla was intact. But there was complete destruction of sub occipital bone, foramen magnum, lateral aspect of occipital condyle and mastoid bone. ACCs are usually having late onset, slow growth, and insidious destruction of surrounding tissues, perineural invasion, and distant metastasis. [10] Clinical feature include pain swelling and numbness with paresthesia. In present case, patient presented with slow growing mass of left mastoid and sub occipital region with facial deviation and difficulty in swallowing. Thorough metastatic work up was done which was suggestive destructive lesion without any regional lymph node involvement and distant spread to chest, abdomen, breast and bone. Surgical management includes radical resection with wide macroscopically free margins even including the non-eloquent normal areas of brain parenchyma. Resection should be followed by

radiotherapy even if tumor borders are free of malignancy and especially when sinonasal areas are involved. Tumors involving nose, paranasal sinuses and maxilla usually have a poor prognosis. [8] Present case was not having any involvement of above areas. However Histopathologically it was high grade tumor. Therefore radical resection should be done after partial or near total embolization.

Spiro *et al.* found the following factors to be important in prognosis: Site , size a nd

extension into

adjacent structures . [13] Blanck *et al* .reported an increased mortality with perineural invasion

and numerous mitoses. [3] Perineural invasion and bone destruction was present in our case,

favoring poor prognosis. In ACC, tumor cells extend well beyond the clinical or radiographic margins and this tumor undergoes not only perineural invasion but also perineural spread.

Therefore, it generally requires radical resection with the widest margins possible and

postoperative radiation therapy of 6000-7500 cgy. [12]The best treatment considered for ACC is

unanimously radical surgical resection followed by radiotherapy. However, in cases of

nasopharyngeal extension and posterior fossa extension, the frequent perineural and perivascular

infiltrations along internal carotid artery, vertebral artery and lower cranial nerves make the

surgical approach risky. Due to this proximity it is difficult to create surgically negative margins.

Surgical treatment, therefore, is often characterized by incomplete excision, consequently

leading to increased frequency of local recurrence. Neurosurgeons should keep patients disability

free, if complete resection of tumor is not possible. However in patients with established

disability, target should be complete resection without leaving any metastatic lymph nodes and

without adding any new deficits. ACCs have a well-known prognostic profile. The 5-year survival rate is 75% but 10-year survival rate is only 20% and survival rate at 15 years is about 10%. Overall prognosis relates to several factors; histologically solid patterns are worse than cribriform pattern . In our case, lesion was mixed with majority having solid portions, which has poor prognosis. Clinical size >4 cm is indicative of more subclinical spread and is associated with a worse prognosis. Delayed diagnosis and treatment also worsens the prognosis. If surgical margins are not clear; the tumor is associated with a poor prognosis despite postoperative radiation therapy. It is one of the most important factors associated with local recurrence. [11] As per literature review, this is first case of ACCs in terms of involvement of posterior cranial fossa with breeching of posterior petrous dura and invading left cerebellar hemisphere. ACCs involving 7^{th} and eight nerves along with 9 to 12th cranial nerves have not been reported till now in literature. Also successful management utilizing percutaneous embolization followed by radical resection lays down treatment solution for these types of patients.

Conclusion

This case is a brilliant example of a giant ACC creating a diagnostic dilemma for posterior fossa

tumor. Clinical features and radiological findings were more in favor of large invasive glomus

jugulare versus sarcoma. However histopathological features established a more perfect

diagnosis of ACCs. Giant ACCs should be managed meticulously but aggressively with

multidisciplinary approach after proper planning and investigations.

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SHORT FORMS – MRI – Magnetic Resonance Imaging, ACC – Adenoid Cystic carcinoma, ECA- external carotid artery, ICA – internal carotid artery, DSA – digital substraction angiogram.

LEGEND - 1

Figure A – Patient showing large retroauricular mass with (arrow mark) black dot suggestive of puncture site for glue.

Figure B – CT head Axial view showing large herniating mass with destruction of, jugular foramen, mastoid and lateral suboccipital region

Figure C – T1 Sequence MRI (Axial) showing large hypointense mass occupying both middle and posterior cranial fossa abutting internal carotid artery.

Figure D – T 2 sequence (Axial) showing large hyperintense mass with multiple flow voids (Arrow mark showing internal carotid artery)

Figure E – T1 contrast sequence (Axial) showing large homogenously enhancing mass at retromastoid region extending anteromedially upto ventral brain stem.(arrow mark).

Figure F – T1 contrast sequence (coronal) showing large homogenously enhancing mass at retromastoid region (arrow mark) with preserved plane from brain stem.

LEGEND - 2

Figure A - Left ECA injection oblique view, showing hypervascular tumor with large intense tumor blush and multiple enlarged sinusoidal spaces within the tumor core.

Figure B - LVA injection, AP view showing the core of the lesion which is made up of large communicating sinusoidal spaces.

Figure C - LAT view, fluorospot image, direct needle puncture done under roadmapping with attempt to access the deep seated sinusoidal spaces first.

Figure D and 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)

Figure F - Final check angiogram ECA injection AP view, showing near total Devascularization

with small area of fine blush in the anteromedial portion of the tumor.

LEGEND 3

Figure A - Post op NCCT head showing near total resection of mass both at intracranial and extracranial region. (Arrow mark)

Figure B – Post op image of patient after 2 month showing healthy scar mark (arrow mark) and overlying skin.

LEGEND - 4

Figure A – Image of patient's postauricular region showing radiation induced cutaneous reaction

(Pigmentation [yellow arrow] and ulceration[green arrow]).

LEGEND – 5

Figure A to D – Showing tumour cells in solid and cribriform pattern with hyalinised stroma.