STUDENTS' CORNER
CASE REPORT

External auditory canal carcinoma with an unusually extensive invasion

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Abstract

External auditory canal carcinoma, while starting out as a seemingly benign condition, if left untreated can have an aggressive course of disease and involve multiple lower cranial nerves. Squamous cell carcinoma remains the most frequent histological type of malignant neoplasm of the external auditory canal and temporal bone. Here we describe a patient with a history of chronic suppurative otitis media with an extensive spread, the tumour was reaching from the skull base to the oropharynx involving neurovasculature along with soft tissues and bones, as well as the cerebellum. The involvement of the recurrent laryngeal and hypoglossal nerves were the most unusual presentation here. The case differed from all previous reported cases as the spinal accessory nerve was to spared. Surgery and radiotherapy are the treatment options but for the inoperable cases presenting with an already poor prognosis, concomitant radiotherapy is the only choice.

Keywords: "Ear Canal/abnormalities" AND "Carcinoma, Squamous Cell, Temporal Bone abnormalities, , Otitis Media.

Introduction

External ear canal carcinoma is a potentially lethal condition, spreading aggressively into surrounding temporal bone. Squamous cell carcinoma has the worse prognosis than other tumours. Biopsy is the test for diagnosis, with CT and MRI being the best modalities to determine the extent of the disease. We observed nerve involvement in a widespread tumour involving the VII, IX, X and XII cranial nerves, whereas the XI nerve was spared as reported earlier.[1] Pyriform fossa invasion was also seen. Here we highlight the importance of detecting chronic suppurative otitis media (CSOM) lest it develops into carcinoma and presents at a late stage of malignancy because of misdiagnosis.

Case Report

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A 48-year-old female patient presented in April 2022 to the ENT department of Dr. Ruth K.M Pfau Civil Hospital, Karachi with a progressively increasing mass in the left external auditory canal, with otorrhoea and otalgia, radiating to the face and the head. She had a chronic history of aural discharge, white and thin in consistency, recently associated with blood. Two months ago, the patient developed left sided facial weakness (grade IV), paralytic lag ophthalmos, and facial droop (Figure-1) There was increased lacrimation, hoarseness and dysphagia for solids. Left sided tongue deviation was present. She also reported occasional vertigo, blurred vision, and headaches. There was decreased hearing in the affected ear, but the patient had no difficulty in understanding speech. Nystagmus was positive, directed towards the right and there were gait disturbances, with a positive Romberg's test. Otoscopic examination showed a grey mobile mass completely obstructing the left ear canal, bleeding upon touch. There was swelling over the left parotid area. The patient had repeatedly visited local clinics over the past few years and had taken antibiotics which would provide short term relief. She was a known case of hypertension and was on Beta blocker.

Fiberoptic laryngoscopy showed immobile left vocal cords, with swollen right aryepiglottic folds.

Figure-1: Patient presented with left sided facial weakness, drooping and paralytic lagophthalmos. Consent has been taken.

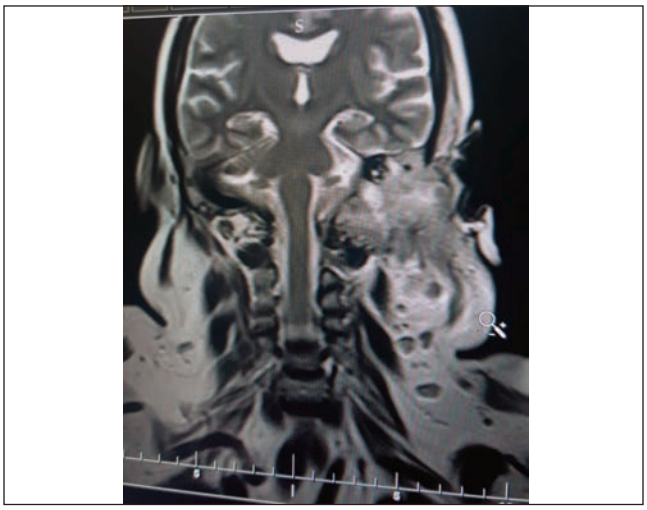

Figure-2: Abnormal signal intensity mass along left mastoid region extending to periauricular space with external, middle and inner ear cavity involvement, as well as the carotid space.

Haematological examination revealed reactive neutrophilic leukocytosis.

HRCT temporal bone showed complete ossification of left middle ear cavity, with erosion of ossicles and scutum, extending medially to involve the lateral semi-circular canal and the petrous bone, laterally, into the external auditory canal, and posteriorly eroding the mastoid air cells, resulting in the widening of the temporomandibular joint. Right sided soft tissue thickening of the middle ear cavity was also seen with erosion of scutum and ossicles and opacification of the right mastoid air cells.

MRI of the neck (Figure-2) revealed a large abnormal signal intensity mass involving the external auditory canal, middle ear, mastoid air cells and squamous temporal bone, infiltrating the left transverse sinus. Medially, it involved carotid space and surrounding muscles. Laterally it reached up to the skin causing overlying skin thickening and involvement of pinna. There was obliteration of the jugular fossa. It closely abutted the cerebellum on the left with adjacent dural enhancement. Soft tissue thickening is seen involving the left parapharyngeal wall, obliterating valleculae and pyriform sinus, as well as the supraglottic and glottic region, with vagus nerve invasion. The lesion measured 6.3 cm in anterior-posterior dimension, 5.3 cm in transverse section and 6.8 cm in cranio-caudal dimension.

Multiple enlarged cervical lymph nodes were seen at level V, the largest measuring 9.8 cm. There was a well-defined lymph node in the parotid gland.

Biopsy concluded the diagnosis of squamous cell carcinoma, grade IV. The case was declared inoperable and the patient was scheduled for combination chemoradiotherapy, but declined due to financial causes and is now seeking symptomatic treatment in Badin. Both the patient and attendant gave their informed consent for publication of the report.

Discussion

SCC happens to be the most frequent neoplasm (39%) of the external auditory canal, middle ear and mastoid.[2] The tumour invades surrounding structures such as the temporal bone, (primary temporal bone SCC), as well the parotid gland, auricular and peri-auricular skin, and temporomandibular joint. The carotid canal, jugular foramen, dura, middle and posterior cranial fossae are invaded in advanced stages.[3] Facial nerve, which runs close to the middle and inner ear, is commonly affected in SCC, particularly considering the neurotropic nature of the disease,[4] hence cranial VII nerve palsies are commonly seen in these cases, an indicator of extremely poor prognosis. Cervical node metastasis is common. Several anatomical routes and barriers for tumour spread have been identified. Cancer spread through temporal bone is highly dependent on pneumatization and presence of natural pathways. From EAC, the tumour approaches middle ear via tympanic membrane, intact or perforated, or by means of invasion of the mastoid air cell system through the posterior EAC wall. Intact TM was found to serve as a reliable tumour expansion barrier.[5] This case had an unusual presentation with dysphagia for solids, with the tumour reaching as far down as the pyriform sinus, as well as the glottic and supraglottic region, with vagus nerve invasion. Hypoglossal nerve was affected, with tongue deviation, as were the vocal cords and the recurrent laryngeal nerve. We found one case with a similar presentation[1] where jugular foramen involvement produced a constellation of symptoms due to related cranial nerves pareses (IX, X, XI), known as jugular foramen syndrome (aka Vernet syndrome) or, with involvement of the hypoglossal nerve (CN XII), Collet-Sicard syndrome, however our patient did not have accessory nerve palsy.

Otoscopic and TM examination can help in diagnosis. CT scan, MRI can judge the extent of tumour and help in staging. PET scans are used to identify metastasis in patients with advanced disease. Lack of research owing to the rarity of the disease means there is very little help for advanced cases. This is often encountered since external auditory canal/temporal bone carcinomas are misdiagnosed as benign otological conditions. This includes cholesteatomas, otitis media and otitis externa which is due to overlapping symptoms of otalgia, otorrhea and hearing loss.[6] Currently, the most widely used system for staging is University of Pittsburgh staging system,[7]

which uses tumour node metastasis system in conjunction with CT scan findings of soft tissue infiltration, EAC bony destruction and temporal bone involvement. Our patient was staged as T4. Among treatment options, surgery remains associated with the best prognosis. Surgery (mostly lateral temporal bone or subtotal temporal bone resection, with a neck dissection and a parotidectomy in case of extratemporal spread)[8] and adjuvant radiotherapy is the treatment of choice. While surgery remains important in advanced disease, for many patients, the carcinoma is declared to be inoperable if it encases the carotid or has direct brain invasion.[9] Chemoradiotherapy, then, is the treatment of choice. While there is conflicting data in literature, there is still plentiful evidence that concomitant chemoradiotherapy (CCRT with a combination of cisplatin, 5-fluorouracil, and docetaxel) for locally advanced temporal bone cancer is effective. One meta-analysis evaluated chemoradiotherapy as a therapeutic option in pre-and postoperative settings for T3 and T4 SCC.[10]

Conclusion

Advanced SCC of EAC can extend to the skull base and the pharynx. CSOM needs proper care, even a precautionary biopsy if the patient is elderly. A presumed benign otological disease not resolving with aural cleaning, otic drops and systemic antibiotics should raise suspicions. Advanced cases have significantly lower overall survival rates, although they seem to have improved recently.

Acknowledgement: Dr Sumera Tabassum, FCPS Radiology, Assistant Professor JPMC for providing help with imaging discussion.

Disclaimer: None.

Conflict of interest: Co-author Zeba Ahmed is also the head of department who signed the letter.

Funding disclosure: None.

DOI: https://doi.org/10.47391/JPMA.7247

Submission completion date: 26-06-2022

Acceptance date: 12-01-2023

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