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Clear cell variant squamous cell carcinoma of temporal bone: Case report and literature review

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ABSTRACT

INTRODUCTION: Squamous cell carcinoma (SCC) of the temporal bone is a rare malignancy. Clear cell SCC is a rare variant of SCC of temporal bone in which no clear risk factor has been suggested as possible etiology. Otalgia, otorrhea, and hearing loss are the usual presenting symptoms of SCC of the temporal bone.

PRESENTATION OF CASE: This is a case is of a 62-year-old female who presented with a 6 months' history of experiencing intermittent left hearing disturbance, loss of balance, persistent left tinnitus, left otalgia radiating to the lateral neck and post auricular swelling. Histology showed clear cell variant, well differentiated SCC. The patient's tumor was found to be non-resectable due to the extensive invasion. The patient was referred for palliative therapy by medical and radiation oncology, however, the patient couldn't tolerate it. One month later she passed away.

DISCUSSION: SCC of the temporal bone is a challenging clinical entity. It is diagnosed mainly by clinical and radiological assessment, and deep biopsies are used to confirm the diagnosis. Temporal bone SCC is usually diagnosed late due to delayed presentation. This case of temporal bone SCC was aggressive in nature and presentation. The presentation was different from the known triad of symptoms of temporal bone SCC which is offensive otorrhea, pain, and bleeding.

CONCLUSION: This case showed a very destructive and bizarre clinical presentation but more report of cases is needed to have a better characterization of the clinical presentation and prognosis of this variant of SCC of temporal bone.

1. Introduction

Squamous cell carcinomas (SCC) of the temporal bone are rare lesions, with annual diagnosis of one to five per 1 million people in United States [1,2]. First description of temporal bone SCC was provided by Schwartze and Wilde in the 18th century [3]. SCC affecting the temporal bone region is an aggressive malignancy with a poor prognosis. The reported incidence is less than 6 cases per million per year, which represents 0.3% of all head and neck cancers with 5-year disease-specific survival reported as ranging from 19% to 48% [1,4,5]. The reason for the aggressiveness of this malignancy might be found in the biological behavior of the disease, but also in the various potential routes of diffusion to the surrounding structures. Moreover, the role of human papilloma virus has been hypothesized as well [6]. Risk factors for SCC of the temporal bone include chronic suppurative otitis media, previous treatment with radiotherapy, and sun exposure [5]. Masterson et al reported that the most common presenting symptoms were offensive otorrhea, pain, and bleeding in which occurred in 33 (55%) of 60 patients, 53.3%, 28.3%, respectively [5].

Facial nerve paralysis is considered as a sign of advanced disease [1,5]. Furthermore, metastasis to cervical lymph nodes is considered relatively common (less than 20%) [5]. The current literature on temporal bone malignancies is limited by the rarity of these tumors, with only nine studies reporting more than 35 cases of any single histology [1,3]. There are multiple subtypes of SCC including clear cell variant [7]. Clinically, clear cell SCC appears as an ulcerated nodule or mass [7]. Clear-cell SCC is a rare entity with seven cases were described in the skin [8]. Moreover, four cases have been reported in the oral cavity and one case in the maxilla [9]. In the literature review, nothing was found regarding clear cell variant of SCC of the temporal bone. In this case report, we will report a rare and destructive case of temporal bone clear cell SCC, with an unusual presentation of such carcinomas. To the best of our knowledge, the patient we reported is the first case of clear variant cell

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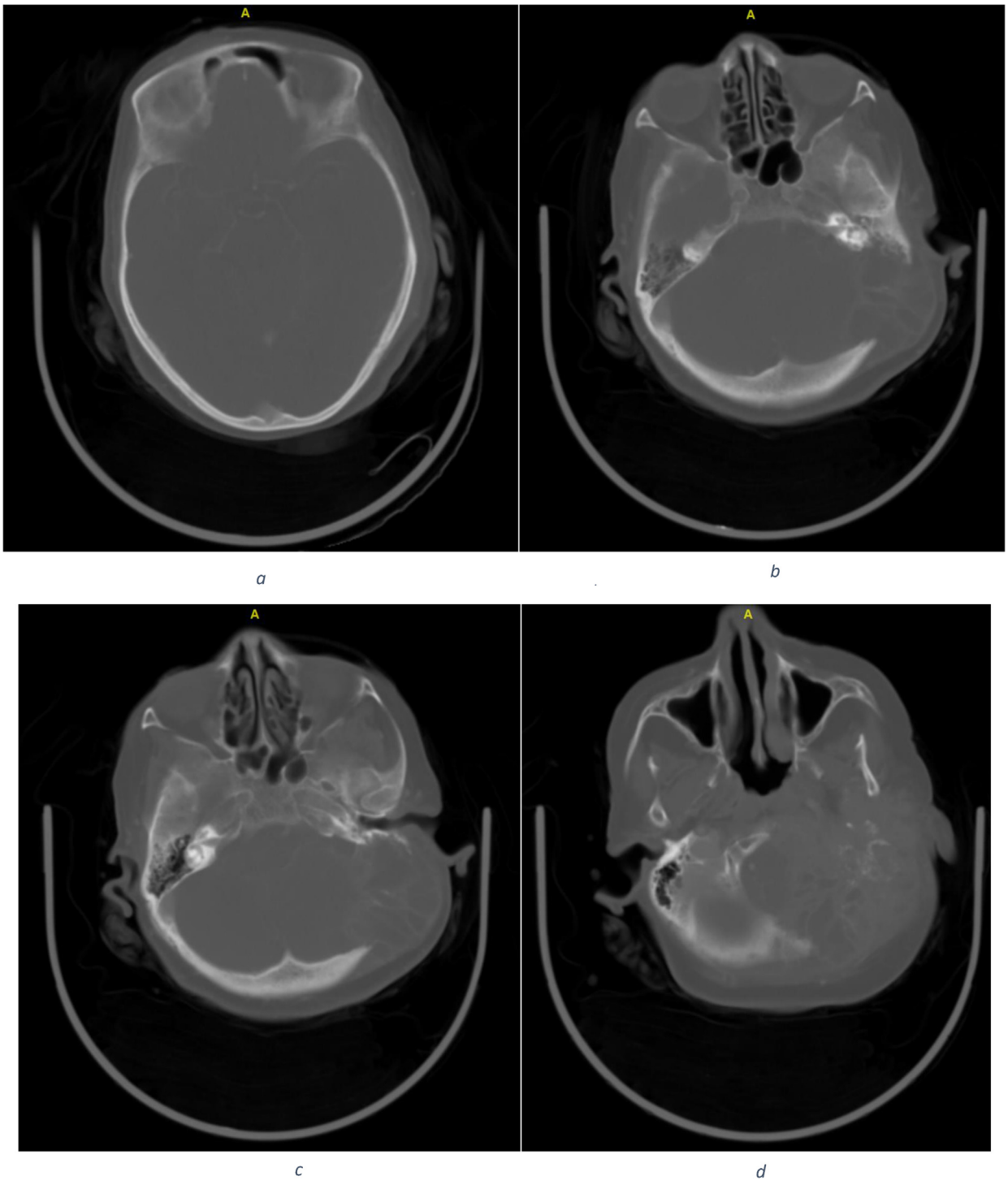

Fig. 1. a–d: Computed tomography (CT) scan of the lesion.
a: CT scan shows intact left parietal bone and skin, b: left mass destructing the temporal bone. c: the mass is larger and more destructive, with intact external auditory canal, d: the mass destructing temporal and occipital bones.

carcinoma of temporal bone. This paper has been reported in line with SCARE criteria [10].

2. Presentation of case

A case of 61-year-old female who presented to our institution in Saudi Arabia with 6 month of experiencing intermittent left hearing disturbance, loss of balance, left otalgia radiating to the lateral neck and post auricular swelling. The swelling was increasing in size for a few months, associated with persistent left tinnitus for a long time. This patient developed hoarseness, and liquid dysphagia for two months, however, there was no history of ear discharge or ulcer. The patient reported a history of upper respiratory tract infection, anorexia, and unintentional weight loss. There was negative history of loss of consciousness, vertigo, otorrhea, or trauma. One year back she had a history of sudden left facial weakness misdiagnosed and treated as idiopathic left facial nerve palsy. The patient was diabetic, and was not known to have hypertension or other systemic disease.

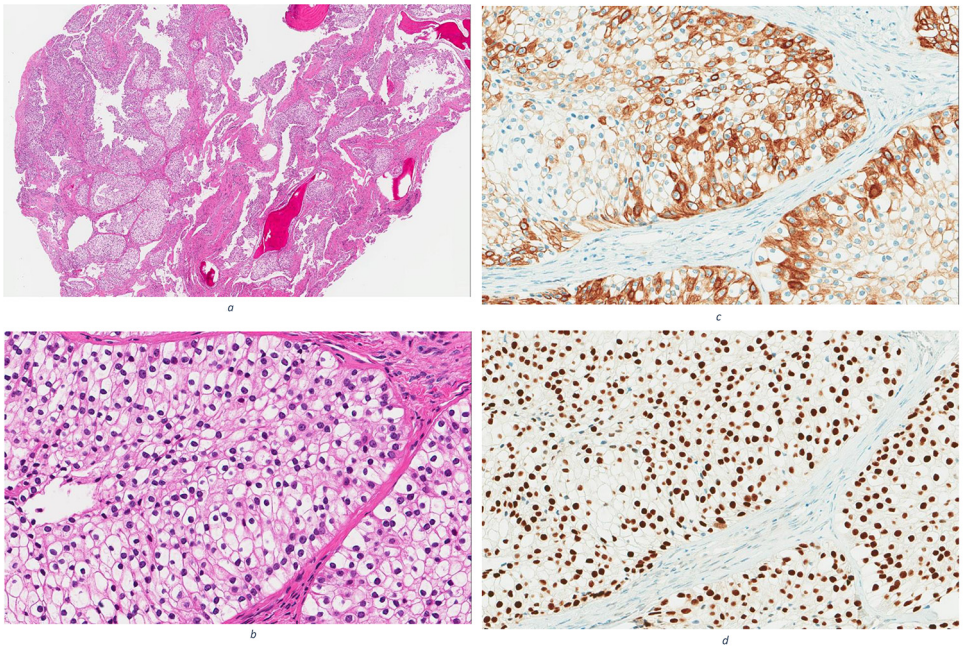

Fig. 2. a–d: Histopathological pictures.
a: Low Power, Panoramic view shows pink fibrous tissue with islands of the tumor. b: High power showing well-formed clear cell membranes with empty cytoplasm and dark nucleus. c: CK 5/6 stains the cytoplasm indicating squamous cell carcinoma. d: P63 immunohistochemistry stain. The nucleus is stained indicating squamous cell carcinoma.

The patient denied any history of smoking, alcohol use, chronic suppurative otitis media, previous treatment with radiotherapy, or excessive sun exposure. There was no family history of head and neck tumors.

On physical examination, there was left post auricular, soft, and tender swelling, with normal left auricle, and clear left external auditory canal and intact tympanic membrane on otoscopic examination. In the right ear, the otoscopic examination showed normal looking tympanic membrane without edema, no clear discharge or bulging of the tympanic membrane and normal appearance. The left facial paralysis was House-Brackmann grade 6. There were multiple enlarged bilateral lymph nodes, levels 1–3. Other systems were normal.

Computed tomography (CT) scan was done on 27 October 2017, and it revealed a large destructive mass lesion involving temporal and occipital bones, and extending to left side of the base of scalp (Fig. 1). There was complete destruction of the left mastoid air cells and extension to the petrous apex on the left side. The mass measured 7 cm in transverse dimension, 8 cm in anterior-posterior dimension, and 6 cm in cranio-caudal dimension. It extended intracranially to the posterior fossa, to the left lateral aspect of craniocervical junction, and to the posterior aspect of the left temporomandibular joint. The patient's skin was intact. There was effacement of fourth ventricle and mild dilatation of the lateral ventricles concerning of early hydrocephalus. There was invasion of left jugular vein. Brain MRI, MRA, and MRV were done and showed similar findings. A provisional diagnosis of skull neoplasm was made.

Tissue biopsy was taken from left skull base and the pathology report showed papillary/cystic architecture extending into the underlying stroma as broad sheets and protrusions showing a cohesive/pushing margin. In some areas, the carcinoma was more infiltrative/non-cohesive. It was clear cell variant, well differentiated (low-grade), no lymphovascular invasion was seen, and negative perineural invasion.

On immunohistochemistry, the carcinoma was positive for CK5/6, EMA (E29), P63 (4A4) and CK7 (Fig. 2). The carcinoma was negative for CK20, Androgen receptors, Ber-EP4, and BCL-2. A diagnosis of well differentiated invasive clear cell variant of squamous cell carcinoma was made. No metastasis was found on chest or abdominal CT scans. The patient was evaluated by head and neck surgery, and her tumor was found to be non-resectable due to the extensive local invasion. The patient was referred for management by medical and radiation oncology. She had very poor general condition and planned to start palliative radiation therapy, however, she was deteriorating clinically and had poor performance status. She didn't receive radiotherapy because she couldn't tolerate it. One month later she passed away.

3. Discussion

SCC of the temporal bone is a challenging clinical entity. The diagnosis is mainly by clinic-radiological assessment, and to be confirmed by local deep biopsies. Temporal bone malignancies are extremely rare, and SCC is the most common type of tumors to occur in the temporal bone. Temporal bone SCC is usually diagnosed late due to delayed presentation [4,11]. Our SCC case was aggressive in nature and presentation. The commonest triad of symptoms of temporal bone SCC is offensive otorrhea, pain, and bleeding which has been reported in other study [5]. However, in our case left otalgia radiating to the lateral neck and post auricular swelling were more prominent. There was no specific known risk factor for the very aggressive and large destructive mass lesion involving temporal and occipital bones. The diagnosis was based on histological examination of tissue from the tumor site which showed well differentiated clear cell variant SCC, which is very rare histopathological type.

Gidley et al. reported that 89% of SCC tumors typically occurred in the bony ear canal, extending laterally to the external auditory meatus, and medially to the middle ear [1]. Radiographic findings showed specific tumor extension to the jugular foramen, the carotid artery, the infratemporal fossa, and the temporomandibular joint [1]. However, in our case tumor was destructive. Factors such as facial nerve paralysis, the disease extension at presentation, positive margin, involvement of dural and cranial nerves are considered

for poor prognosis. As mentioned earlier, facial nerve in this case was grade 6 at presentation.

The temporal bone may be eroded by gross extension or microscopic undetectable intra-osseous infiltration. Adjoining sites such as jugular foramen, dura mater, internal carotid artery, facial nerve, parotid, and condyle may be involved by local growth of the tumor. As in our case, typical invasion of left jugular vein and facial nerve involvement were seen.

Our patient had multiple enlarged bilateral lymph nodes, levels 1–3, as it is reported in the literature that cervical lymph nodes metastases are relatively common (less than 20%) [5]. Our case showed advance stage of SCC with facial nerve involvement. Pittsburgh tumor staging system is most used for squamous cell carcinoma of the temporal bone [12].

Recently, Moody et al. proposed that facial nerve involvement is included in the classification of T4 tumors. The modified Pittsburgh staging of squamous cell cancer of the temporal bone classifies patients who present with facial paralysis as T4 tumors [13]. Perineural invasion and angio-lymphatic diffusion are local features of tumor aggressiveness [12].

The primary lesson from this case this study is to draw attention to the possible early intervention and diagnosis and more report of cases is required to characterize the clinical behavior and prognosis of this rare variant of SCC.

4. Conclusion

In summary, SCC of temporal bone is very rare, aggressive, and the management depends on the stage of the tumor. Early detection of the tumor before extensive spread allows for better treatment and prognosis. This patient was diagnosed as clear cell variant SCC with no evidence of systemic metastasis. To the best of our knowledge, the patient we reported is the first case of clear cell variant SCC of temporal bone, which appears to be quite aggressive, fast growing lesion, and shares the general poor outcome of temporal bone malignancies.

Conflicts of interest

There are no conflicts of interest.

Sources of funding

No funding source.

Ethical approval

Case reports do not require ethical approval by our institution if no patient identification and case is anonymous.

Consent

No consent because the patient has passed away and no next of kin could be reached. The head of our department has taken responsibility that exhaustive attempts have been made to contact the family and that the paper has been sufficiently anonymised not to cause harm to the patient or their family.

Author contributions

Abdulaziz AlEnazi: Study concept, writing the paper.
Fahad Alwadi: Writing the paper.
Yazeed AlOqaili: Study concept and review the paper.

Registration of research studies

NA.

Guarantor

Abdulaziz AlEnazi.
Yazeed AlOqaili.

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