CASE REPORT

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Primary Cutaneous Adenosquamous Carcinoma of Scalp: A Rare Variant

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ABSTRACT Adenosquamous carcinoma is an aggressive and rare variant of squamous cell carcinoma that has both squamous and glandular differentiation. We present a case of occipital scalp adenosquamous carcinoma in a 43-year-old male who presented with occipital mass that was initially marble in size, which progressively enlarges over years. There were no constitutional symptoms. No neurological deficit and locoregional lymph nodes were not palpable. Contrast enhanced computerized tomography brain revealed a non-enhancing midline occipital scalp mass with no intracranial extension or bony involvement. Tissue biopsy of the occipital mass was performed, and preliminary microscopic and immunohistochemistry studies suggested adenosquamous carcinoma. The tumor was completely excised with wide local excision, followed by skull burring and primary closure.

Keywords: Adenosquamous carcinoma; squamous cell carcinoma; scalp; margins of lesion

Adenosquamous carcinoma (ASC) is a rare variant of squamous cell carcinoma (SCC) that shows both squamous and glandular differentiation with aggressive clinical behavior.[1] ASC is commonly seen over head and neck of patients, most frequently in male.[2-4] Clinically, it is usually indistinct from typical SCC which present as an indurated, keratotic plaque. To our knowledge, there are approximately 53 published cases in English literature describing ASC.[1-10] In the absence of Mohs micrographic surgery (MMS), this case shows an occipital scalp adenosquamous cell carcinoma treated with wide local excision and defect closed primarily.

CASE REPORT

A 43-year-old man presented with occipital mass associated with pain and itching. The mass was initially marble in size which progressively enlarges over years. There were no constitutional symptoms. Clinical examinations revealed a fungating mass (5x4 cm) with maggot infestation (Figure 1). There was no neurological deficit and locoregional lymph nodes were not palpable.

Contrast enhanced computerized tomography brain revealed a non-enhancing midline occipital scalp swelling (0.7x3.5x4.0 cm) with no intracranial extension or bony involvement.

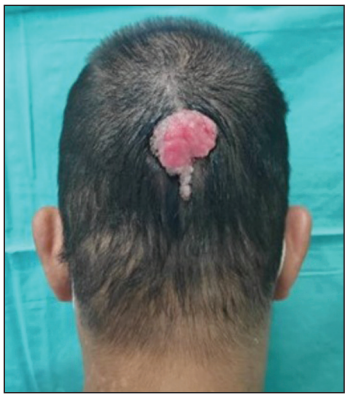

FIGURE 1: Adenosquamous cell carcinoma of the scalp at occipital region.

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Tissue biopsy of the occipital mass was performed. The histopathology report of the biopsy showed that the tumor tissue displayed squamous and glandular components. For immunohistochemistry staining, the malignant squamous cells were positive for p63, p40 and cytokeratin (CK) 7, whereas for malignant glandular cells, CK7 and carcinoembryonic antigen (CEA) were positive. Both malignant components were negative for CK20, S100 and SOX10. Based on the microscopy features and immunohistochemistry study, the occipital lesion was highly suggestive of ASC.

Wide local excision was performed, with 1 cm surgical margin from the mass and deep down until the pericranium. The exposed skull bone was burred until punctate bleeding was seen. The surrounding scalp was undermined and galeal scoring were done. The defect with size of 7.2x5.0 cm was closed primarily (Figure 2).

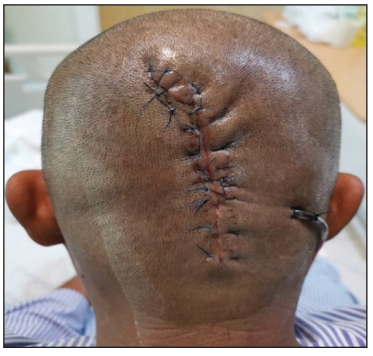

FIGURE 2: Post wide local excision and primary closure.

Pathological examination reported the scalp tissue was infiltrated by the malignant tumor arising from the epidermis and comprised of squamous and glandular components (Figure 3A, Figure 3B). The malignant squamous cells were arranged in solid nests intermixed with malignant glands invading into desmoplastic stroma (Figure 4). There were also vesicular nuclei with abundant eosinophilic cytoplasm. The nuclei of malignant glandular cells are hyperchromatic, while the cytoplasm is eosinophilic. Deep dermis invasion was found in the malignant cells, but no perineural invasion was seen. The occipital mass was excised completely with proper surgical margin microscopically (superior margin 14 mm; inferior margin >23 mm; lateral margin 17 mm; medial margin 17 mm and deep margin 10 mm). The overall pathological features were suggestive of ASC. The consent was obtained from patient.

DISCUSSION

Primary cutaneous ASC is a rare and aggressive carcinoma with distinct histological features that enables clinicians to distinguish it from other possible primary cutaneous SCCs. A survey of the literatures revealed 52 published cases, of these published cases, 32 cases were found in face and neck, Eight cases on the scalp, 7 on the extremities, 4 on the trunk and 1 on

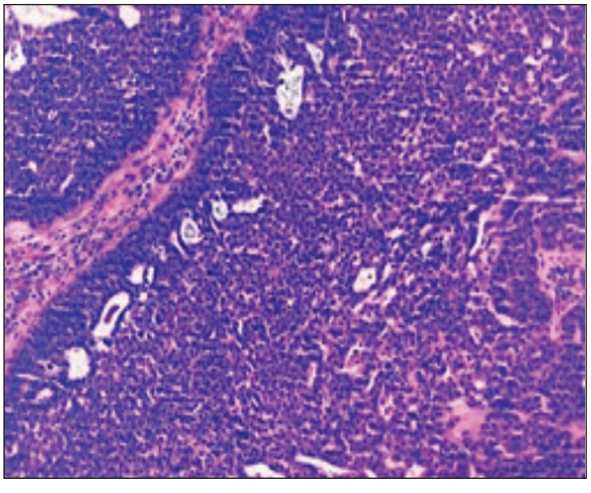

FIGURE 3A: Malignant squamous (H&E; x10).

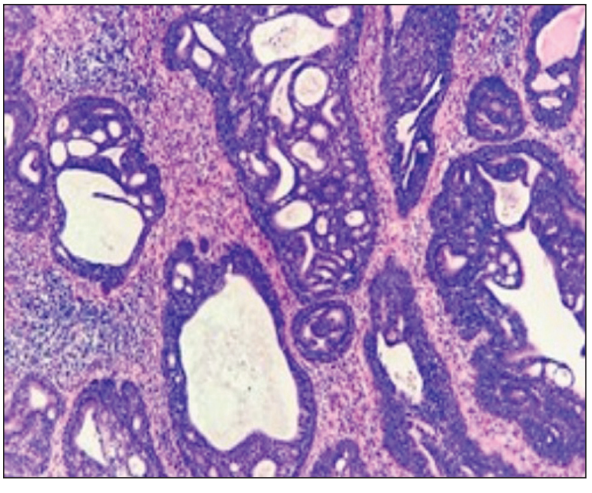

FIGURE 3B: Malignant glandular (H&E; x10).

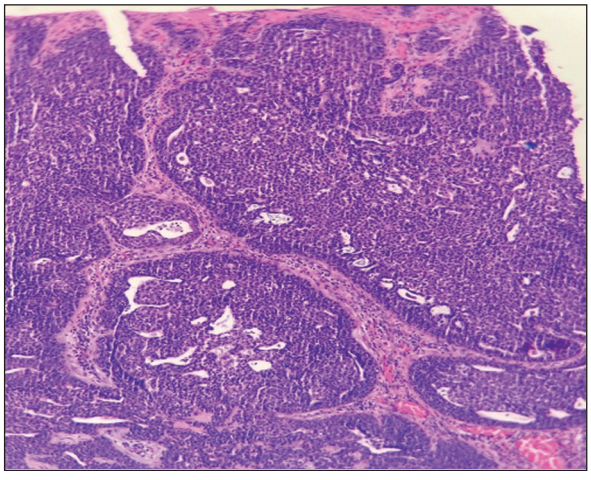

FIGURE 4: The malignant squamous cells are arranged in solid nests admixed with malignant glands surrounded by desmoplastic stroma (H&E; x10).

the inguinoscrotal region. The ASC seems to have a higher incidence in males and affects those between the ages of 48-95 with mean age of 72.[1-10]

The ASC typically presents as an indurated keratotic plaque or ulceration ranging from 1 to 6 cm in size which exhibits mixed squamous and glandular differentiation.[4] The histopathological findings for ASC include: 1) Malignant squamous cells arranged in solid nests with intercellular bridges in desmoplastic stroma; 2) Malignant glands with hyperchromatic nuclei and eosinophilic cytoplasm and 3) Significant mitoses in both squamous and glandular components.[11] The immunohistochemical evaluation of ASC confirms glandular differentiation with positive results of CEA and CK7 as described in the largest case series by Fu et al. in 2009.[3,8] A case series done by Banks et al. was reported all nine cases contained CEA within the tumor cells.[2] It enables the confirmation of true glandular differentiation to differentiate ASC from other cutaneous SCCs like acantholytic SCC which lack true gland formation with luminal and mucin production. The squamous differentiation produces a diffuse staining combination of P63, CK5/6/7 and P40.[3,8] This helps to support the diagnosis of a primary cutaneous malignancy rather than a metastatic adenocarcinoma.

The behavior of ASC is known to be more aggressive than conventional SCC. Banks et al. reported that 5 out of 10 cases died due to local recurrence.[2] Similarly, a study by Fu et al. reported recurrence in 3 out of 6 cases and important predictors that led to tumor recurrence and metastasis were identified i.e., lesion thickness/depth of invasion, microscopic perineural invasion and immunosuppression.[3] Therefore, it is highly recommended that patients are monitored closely after initial treatment of ASC.

Treatment of ASC includes surgery with or without chemoradiotherapy. Of all the cases described, 10 out of 53 cases underwent radiotherapy after surgical excision due to recurrence. Genois et al. recorded 1 case in which neoadjuvant chemotherapy was used but the patient did not respond satisfactorily and thus referred for palliative radiotherapy.[6] Otherwise, the rest of cases were treated by standard excision or MMS.[1-10] To date, there is limited data to recommend defined peripheral and deep margins for cutaneous ASC with standard excision. Therefore, strong caution is need when MMS is not available.[12]

Although MMS is recommended for cutaneous ASC, but it isn't broadly accessible. Thus, standard excision along with close follow up may be considered in center without the facility. More studies will be needed to define peripheral and deep margin when standard excision is performed. This brings up the lack of data to describe the treatment option in this region of the world to aid in further management of the disease.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Wee Yi lim, Hoe Vee Chuan, Yi Jun Lee;* ***Design:*** *Wee Yi lim, Zosimo Ken L Jimeno;* ***Control/Supervision:*** *Zosimo Ken L Jimeno;* ***Analysis and/or Interpretation:*** *Zosimo Ken L Jimeno;* ***Literature Review:*** *Wee Yi lim;* ***Writing the Article:*** *Wee Yi lim, Hoe Vee Chuan, Yi Jun Lee;* ***Critical Review:*** *Zosimo Ken L Jimeno.*

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