

### Primary branchiogenic carcinoma – is it a valid entity?

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#### Introduction

Branchial cyst is due to a congenital defect that occurs early in embryonic development. It is the most common congenital cause of a neck mass which results from a failure of obliteration of one of the branchial clefts during embryonic development. No ethnic or sexual predilection has been reported. Approximately 2-3% of cases are bilateral. A tendency for familial clustering has been documented [1].

Branchial cysts may not present clinically until early adulthood. They may become enlarged and tender especially following an upper respiratory tract infection. Sudden rupture of an abscess which has developed inside a branchial cyst may lead to the formation of a discharging sinus.

Depending on the size of the branchial cyst local symptoms such as dysphagia, dysphonia or dyspnoea may occur [2].

We present a case report wherein the patient underwent excision of a branchial cyst on the right side of the neck which revealed squamous cell carcinoma in its lining.

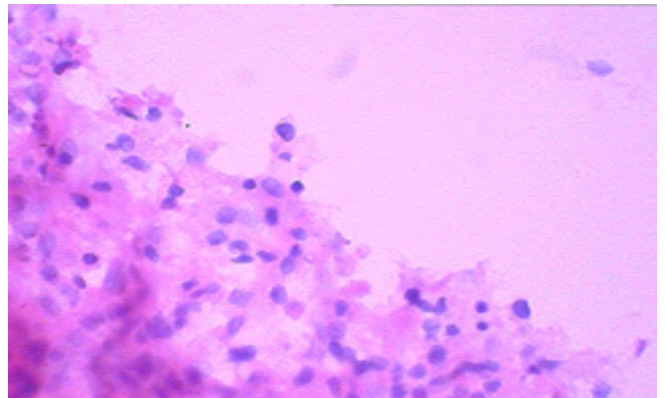
#### Case Report

A 35-year old male, non-smoker, presented to us with a fluctuant, non-tender lump. The lump was on the right side of the upper third of the neck in the anterior triangle and had been present for 4 months duration. Computerized tomography (CT) showed a well-defined cystic mass of 46 x 24 x 22 mm, which was anterolateral to the carotid sheath. There were no tracts connected to the cyst and no evidence of lymphadenopathy or any other malignancies of the head and neck region either clinically or radiologically. The preliminary diagnosis was a branchial cyst and the patient was referred for fine needle aspiration cytology.

#### Cytological findings:

Approximately 1.5 ml of a brown coloured thin fluid was

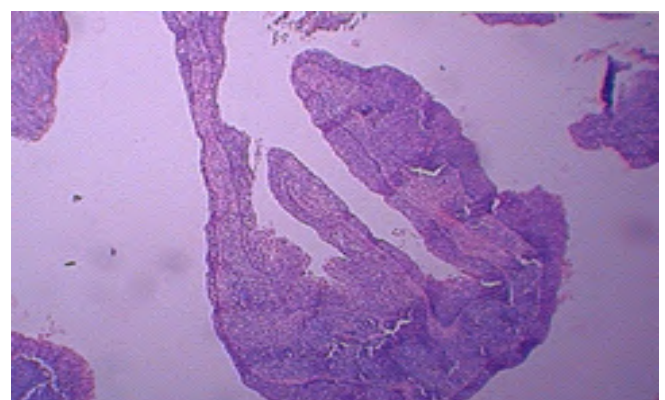
obtained by fine needle aspiration. Smears revealed sheets of squamous epithelial cells mixed with a large number of mature lymphocytes, neutrophils and macrophages. The epithelial cell component exhibited nuclear atypia and the smears were categorized as “C3/atypical cells” according to the WHO guidelines [Figure 1].



**Figure 1.** FNAC smear (H & E stain x 10) shows atypical squamous epithelial cells with large hyperchromatic nuclei.

#### Morphological findings:

In view of the atypical cytological findings the lesion was excised in toto. Gross examination showed a thin walled collapsed cyst measuring 3.5 x 2.5 x 2 cm [Figure 2] with a solid whitish (12 x 10mm) mural nodule.



**Figure 2.** H & E stain (x 4) - Cystic lesion with an epithelial lining and stroma rich in lymphoid tissue.

Representative haematoxylin & eosin stained sections showed a cystic lesion lined by the stratified squamous epithelium. Cyst wall consisted of lymphoid tissue with

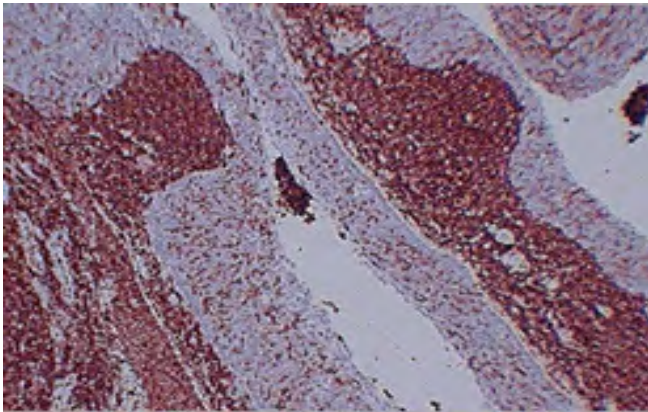
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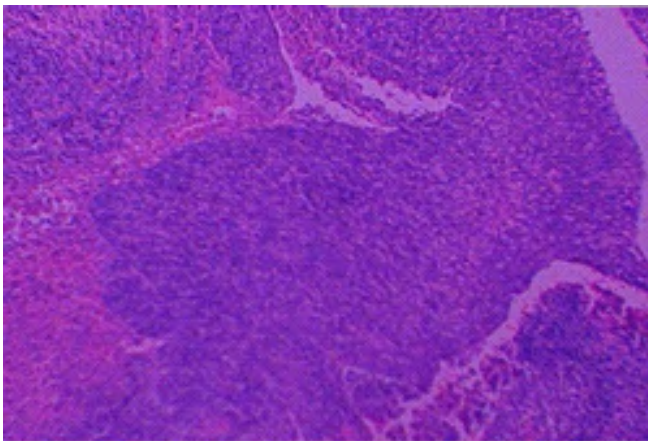
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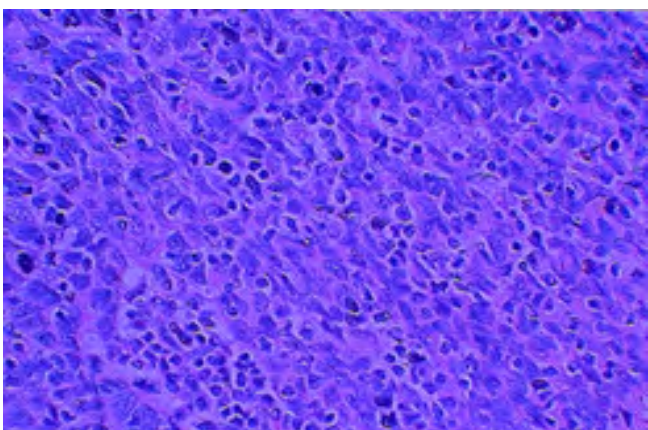
germinal centres [Figure 3]. There was an area of transition from a benign epithelium to full thickness dysplasia [Figure 4 and 5] and invasive carcinoma. Pancytokeratin stain highlighted the invasive area of the cyst wall [Figure 6].



**Figure 3.** Immunohistochemical stain LCA (x 10) - Positive staining of the lymphoid stroma (surface epithelium- negative).



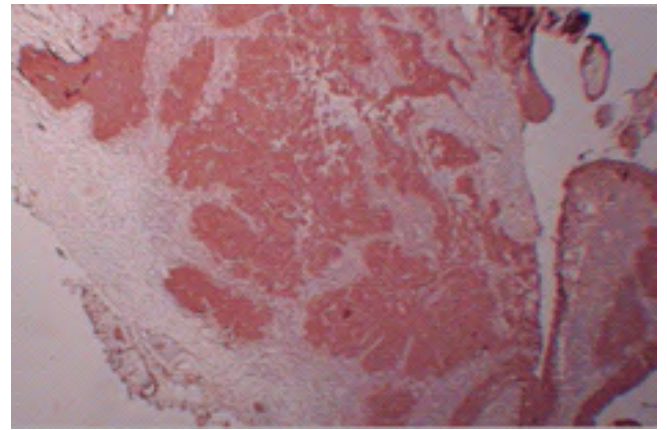
**Figure 4.** H&E stain (x 10) – Marked epithelial stratification (solid area).



**Figure 5.** H&E stain (x 40) - Severe epithelial dysplasia corresponding to carcinoma in situ.

### Discussion

Occurrence of primary carcinoma in a branchial cyst has been considered controversial. Khafif et al [1] described strict



**Figure 6.** Pancytokeratin stain (x 4) – Downward extension of the epithelial component; suggestive of early invasive carcinoma.

diagnostic criteria in 1989 following the earlier descriptions of the entity by Volkmann (1882) and Martin et al [2].

Criteria proposed by Khafif et al. [1] include:

1. The location of the tumour in the anatomic region of the branchial cleft cyst or sinus.
2. Histological appearance of the tumour consistent with its origin from branchial vestiges, i.e. squamous cell carcinoma.
3. Presence of carcinoma within the lining of an identifiable epithelial cyst.
4. Identification of transition from the normal squamous epithelium of the cyst to carcinoma.
5. Absence of any identifiable primary malignant tumour.

Bhanote [3] identified 14 cases between 1982 and 2008 that passed the Khafif criteria [1]. There were 6 reports totalling 9 cases of primary branchial cyst carcinoma between 2008 and 2014. These reports were by Roche (2010), Banikas (2011), Veivers (2012), Chauhan (2013), Tegeltija (2013) and Anantharajan (2014) [4 – 7]. Banikas [5] and others believe that true primary carcinomas occur, citing the simultaneous presence of in situ carcinoma, early invasion and invasive carcinoma. In spite of the above reports scepticism about this entity persists. Veivers and Dent have shown that current investigative methods are unsatisfactory in the diagnosis of hidden primaries. Therefore, item 5 of the Khafif criteria is difficult to be satisfied. Surgeons would be encouraged by the findings of Veivers and Dent to complete the diagnostic work up to minimize errors.

### Conclusion

Primary branchiogenic carcinoma is a controversial clinicopathological entity with conflicting results in literature. However, we believe our case satisfies all the diagnostic criteria described by Khafif et al [1]. The most important criterion emphasized by the author of demonstrating the zone of transition from benign to full thickness dysplasia and frankly invasive carcinoma, was

clearly identified in our case. Hence we report this case as a primary squamous cell carcinoma (early invasive) arising in a branchial cyst. Since the lesion was completely excised no further surgery was attempted. Patient has been regularly followed-up for the last 3 years. There is no evidence of recurrence or metastasis or the emergence of an occult primary to date. All above findings lend credence to the fact that it is indeed a primary branchiogenic carcinoma.

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#### Key Points:

- Primary branchiogenic carcinoma, which is a squamous cell carcinoma arising in a branchial cyst, is extremely rare and a highly contentious clinicopathological entity.
- The case report highlights the controversy surrounding this entity and establishes the diagnosis based on a set of histopathologic criteria proposed by Martin et al, and later modified by Khafif et al.
- In this case the identification of the normal cyst lining, zone of transition and the frankly invasive component, within the same lesion supported a diagnosis of primary branchiogenic carcinoma over a metastasis.