Branchial Cleft Cyst Carcinoma: a case report

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Branchial cleft squamous cell carcinoma is extremely rare, compared to the far more frequent cystic metastastic cervical lymph nodes arising from primary malignancies. This was a 40-year-old man presenting with a well-defined, two-month history of a slowly-growing lateral neck mass. He underwent complete tumor excision. The pathological diagnosis was squamous cell carcinoma originating from a branchial cleft cyst, which was characterized histologically by an epithelial-lined cyst with dysplasia to in situ carcinoma transitional zone.

Key words: Branchial cleft cyst; Squamous cell carcinoma

Branchial anomalies exist in one of the three forms: sinuses, fistulas, or cysts. They are considered as persistence of vestigial remnants of a branchial cleft or pouch. A branchial cleft cyst results from an entrapped portion of cleft or pouch with or without an intervening tract. Most of them are located along the anterior border of the sternocleidomastoid muscle as are the cervical lymphatics.

The well-known Martin et al. criteria [1] for establishing the diagnosis of primary branchial cleft carcinoma proposed in 1950 was: (1) the cervical tumor must occur somewhere along the anterior border of the sternocleidomastoid muscle; (2) the histological appearance of the growth must be consistent with an origin from tissue known to be present in branchial vestigial; (3) the patient must have survived and have been followed by periodic examinations for at least five years without the development of any other lesion which possibly could have the primary tumor; (4) there should be histological demonstration of a cancer developing in the wall of an epithelial-lined cyst situated in the lateral aspect of the neck. The fourth criterion was emphasized by Martin el al. for the confirmations of a branchial cleft carcinoma.

Here presents a case report of a primary branchial cleft carcinoma.

CASE REPORT

A 40-year-old male patient with a history of 2-month slowly-growing, soft, movable nontender mass in left lateral neck measuring about 4x3x3 cm in size, located anterior to the sternocleidomastoid muscle. General physical examinations and plain chest radiography were done without presence of abnormalities.

Computed tomography (CT) of the neck revealed a 3.2 × 3 × 2.5 cm circumscribed cystic mass lesion lateral to the left carotid sheath, behind the left submandibular gland, and deep to the left sternocleidomastoid muscle (Fig. 1). Moderate enhancement of the slightly lobulated thick wall with internal nonenhancing homogeneous fluid density was shown on contrast-enhanced CT (Fig. 2).

No definite remarkable abnormal enlarged
regional or lymph nodes in the neck noted.

The routine workup for nasopharyngeal carcinoma was performed including nasopharyngeal examinations with random biopsies with negative result. Panendoscopy for upper gastrointestinal tract also revealed negative finding. Then, complete tumor resection with regional lymph nodes dissection was performed. The histological microscopic findings are an encapsulated lymphoid and squamous cell-lining epithelial cystic lesion with focal invasive squamous cell carcinoma with adjacent foci of dysplastic or in situ squamous carcinomatous epithelium in the cystic wall, suggestive of malignant transformation from branchial cleft cyst (Fig. 3). The post-operative follow up CT performed half year later showed no residual tumor, or recurrence.

**DISCUSSION**

The diagnostic criteria of branchial cleft carcinoma is quite complicated, and controversial. Most cases of suspicious branchial cleft carcinomas are probably cystic metastases from primary tumors in head and neck. The incidence of the primary branchial cleft carcinomas is extremely rare. Khafif et al. (1989) argued the third criterion of Martin et al. (1950) for confirmation of primary branchial cleft carcinoma: a five-year follow up without evidence of a primary tumor of upper aerodigestive tract. Due to (1). The patient may die of unrelated causes in five years; (2) post-operative irradiation may control an occult primary tumor. He proposed two other modification

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**Figure 1.** A 3.2 × 3 × 2.5 cm circumscribed cystic mass lateral to left carotid sheath behind the left submandibular gland, and deep to the anterior margin of left sternocleidomastoid muscle.

**Figure 2.** Postcontrast CT showed moderate enhancement of focal irregular thick wall, and internal fluid density.

**Figure 3.** Papillomatous growth of a keratinizing squamous cell carcinoma arising from a branchial cleft cyst, with adjacent foci of adjacent carcinoma in situ, the transition zone.
instead of the “five-year rule” as: (1) the absence of an identifiable primary cancer by a thorough evaluation, including endoscope, CT of the head and neck, and appropriate biopsies; and (2) histological identification of a cystic structure partially lined by normal squamous or pseudostratified columnar epithelium with gradual transition to invasive squamous cell carcinoma [2].

Although Singh (1998) suggested the necessity of a five-year disease-free period must be maintained toward establishing a diagnosis of branchial cleft carcinoma with certain minor modifications in that: (1) retained branchial cleft carcinoma diagnosis in cases dying of unrelated diseases in five years; (2) the diagnosis of branchial carcinoma should be discarded in patients receiving radiotherapy who are found to have squamous cell carcinoma in the upper aerodigestive tract within five years, because metastatic diseases to the upper jugular lymph nodes are more common [3]. The Khafif et al. modified criteria appeared to be more practical for diagnosis, and also treatment guidelines as well.

This presented case satisfied the Martin criteria except for the third criterion of a five-year disease-free survival. However, the modified criteria of Khafif et al. was fulfilled instead, which supports the diagnosis of a primary branchial cleft carcinoma.

The differentiation of a branchial cleft carcinoma from a cervical metastases was difficult, since the location and histological features are very similar. The three features that are exclusive to lymph nodes are: peripheral lobulation, internodular trabeculae, and perinodal sinuses. Cervical metastases may show some of these features, and thus discernible branchial cleft carcinomas. Normal epithelial-lined portion, or dysplastic epithelium may be presented next to the squamous cell carcinoma, which supports the branchial cleft origin. On the basis of histological findings, the transitional zone of normal to dysplastic epithelium to carcinoma is the most important feature for the confirmation of branchial cleft carcinoma [4].

Wide surgical resection and neck dissection are recommended for branchial cleft carcinoma. Radiation therapy may be reserved when all dissected lymph nodes are negative for tumor. However, radiation therapy is recommended when patients refuse further lymphatic dissection or positive lymph nodes are detected following a neck dissection [1]. This case report help reminding us the rare existence of branchial cleft carcinoma, and familiarizing with the two diagnostic criteria. Although only carcinoma in situ was confirmed on the basis of pathology, close follow-up examination should be suggested.

REFERENCES

鰓裂性囊腫之鱗狀細胞癌：病例報告

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鰓裂性囊腫之鱗狀細胞癌的診斷頗具爭議。自從1950年Martin et al首度提出四項診斷準則後，鰓裂性囊腫之鱗狀細胞癌存在之爭議才告一段落。自此，正式之病例報告僅偶見報導，但其發生率仍屬稀見。主要的鑑別診斷在於囊腫性頸部淋巴腺轉移病灶，需術前排除且於五年內未検出上呼吸道消化道及甲狀腺之惡性腫瘤。Khafif et al於1989年針對Martin準則作出修訂，認為針對上呼吸道消化道及甲狀腺之完整的術前檢查排除惡性腫瘤即可。如此鰓裂性囊腫之鱗狀細胞癌的診斷與治療變得更實際可行。

關鍵詞：鰓裂性囊腫：鱗狀細胞癌