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Primary branchial cleft carcinoma – a case report

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Abstract
Primary branchial cleft carcinoma is a diagnostic challenge. The majority of cases have been shown, following the application of rigid criteria, to have developed from an occult metastasis elsewhere.

We describe what we believe to be only the second reported case in a patient who fulfils all of the established criteria.

Key words: Branchial apparatus; Carcinoma; Neoplasm metastasis, occult

Introduction

Primary branchial cleft carcinoma is an enigma. A critical analysis of previously reported cases has shown that the diagnosis of a primary focus of disease has been inappropriate. Long-term follow-up has revealed that the majority of cases have arisen from an occult primary somewhere else in the nasopharynx. Martin and Morfit (1950) reviewed the literature and applied stringent criteria for establishing the diagnosis of a primary branchial cleft carcinoma. To our knowledge there has

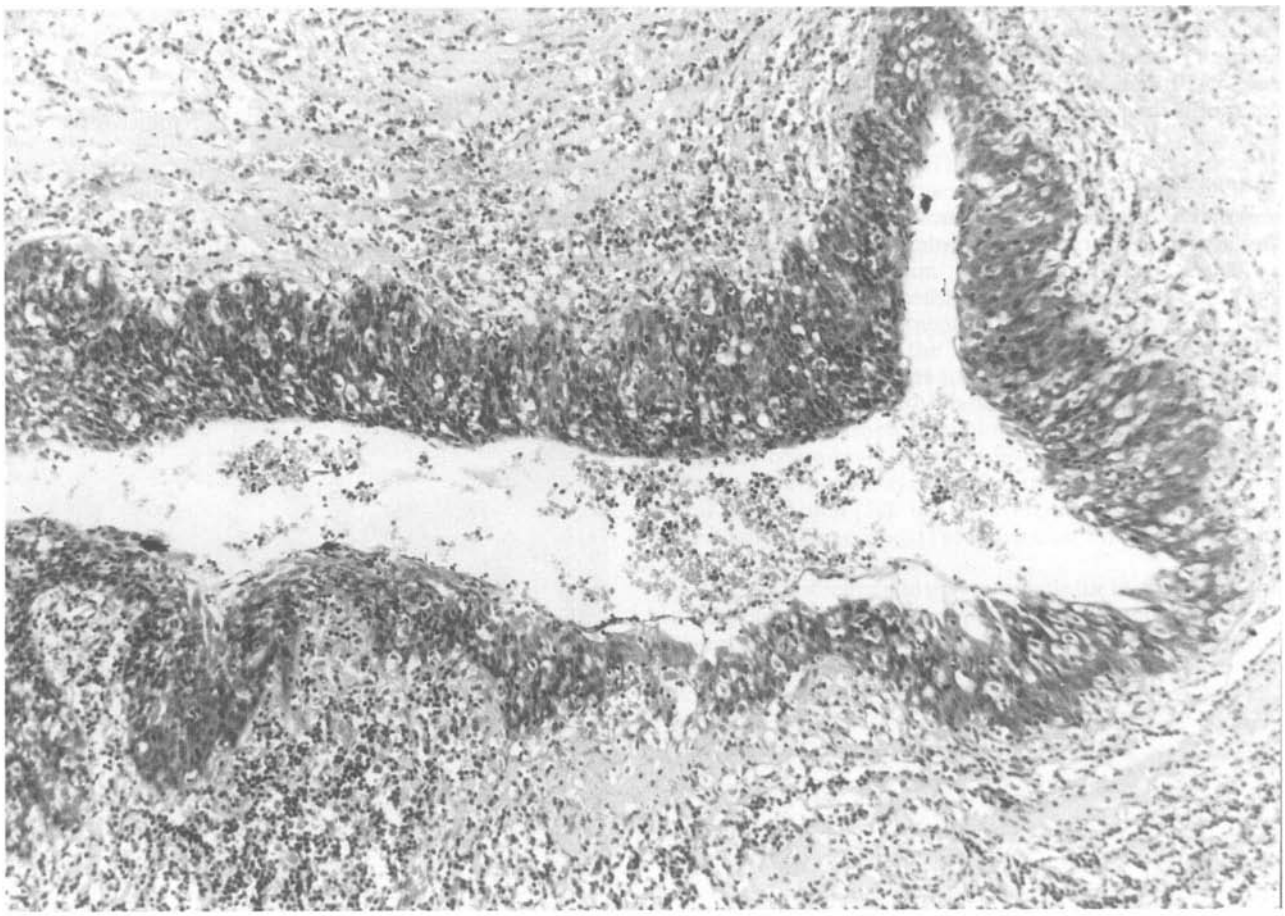

Fig. 1
Dysplasia and carcinoma *in situ* arising in the cyst epithelium (H & E; × 40).

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FIG. 2
Squamous cell carcinoma in the cyst wall (H & E; × 10).

been only one previously reported case in the English literature of a squamous cell carcinoma arising de novo from a first branchial cleft remnant.

Case report

A 55-year-old male presented in 1979 with a two-month history of a lump in the neck. It had been increasing in size since it was first noticed giving rise to occasional pain. The patient had not experienced any dysphagia nor discovered any other lumps elsewhere. He did report, however that his voice was becoming more hoarse. His past medical history included a hernia repair and the removal of a thyroid goitre eight years earlier. The latter was subsequently found to be a thyroid adenoma. The patient was otherwise well, smoked 30 cigarettes per day and drank four units of alcohol per week. Examination revealed a 2.5 × 2.5 cm round, smooth lump arising from the anterior border of the right sternocleidomastoid, which was fluctuant and slightly tender. It failed to transilluminate and was not fixed to any superficial or deep structure. There was no lymphadenopathy and the remainder of the examination was unremarkable. Following excision of the lump, histology showed a poorly differentiated squamous carcinoma producing some cystic areas and arising from normal cyst epithelium lining a branchial cyst (Figure 1). A photomicrograph of the squamous cell carcinoma is shown in Figure 2. No primary site was discovered on subsequent examination. Fifteen years later the patient has remained well with no evidence of any recurrence nor any evidence of any other primary focus of disease.

Discussion

Towards the end of the 18th century, Von Volkman (1882) described what he believed to be a carcinoma arising from a vestigium of the branchial apparatus. No other primary site has been located, and so he concluded that these tumours originated from the branchial cleft. As a result more than 200 cases were subsequently reported as being primary branchial cleft carcinomas. Some time later in 1950, Martin and Morfit reviewed the literature to date and suggested that such a diagnosis was in fact inaccurate. They went on to suggest four criteria that should be fulfilled in order to establish the diagnosis of a primary branchial cleft carcinoma:

(1) The neoplasm should occur in recognised sites of branchial clefts, namely on a line passing from the tragus, along the anterior border of sternocleidomastoid, to the clavicle.
(2) The appearance of the tissue histologically must be consistent with the native tissue at that site.
(3) The patient should be alive at least five years later, with no evidence of a primary site elsewhere.
(4) There must be histological proof that the carcinoma arises from the normal epithelium of the branchial cyst.

These criteria have ensured that the number of true cases of primary branchial cleft carcinoma has been limited (Beamish and Som, 1973; Shreedhar and Tooley, 1984; Sandiford *et al.*, 1987; Park and Karmody, 1992).

A controversy arises as to the phylogenetic origin of these carcinomas for several reasons. (1) Metastatic

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squamous cell carcinoma is expected to occur with a far greater frequency than a primary branchiogenic carcinoma; (2) a cervical metastasis may be the only presenting feature of an occult lesion that only later presents itself; (3) the jugular lymphatic system and the majority of branchial cysts are found in anatomically similar regions; (4) it is difficult to histologically distinguish a branchial cleft carcinoma from a cystic degeneration of a metastatic cervical node. It is clinically important to establish an early correct diagnosis in order to plan an appropriate therapeutic regime.

This case is one of the few that actually satisfies all four criteria. Confirmation that the squamous cell carcinoma was de novo in origin was seen with areas of dysplasia arising from the normal epithelial lining of the branchial cyst (Figure 1). Furthermore the lesion was located in a swelling at the anterior border of sternocleidomastoid and consisted of tissue which was appropriate for the location.

Fifteen years later the patient is alive and well and no other primary source has been isolated.

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