Recurrent neck mass after carotid body tumour excision: a case report and review of the literature

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We report a rare case of a malignant lymphomatous neck mass masquerading as a recurrent Carotid body tumour (CBT) and propose a classification for recurrences after CBT to aid further management.

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Case Report

A 53-year-old woman was referred to our vascular unit with a two-year history of a non-trauma related slow growing, painful left neck mass. The mass was pulsatile with an associated bruit and was easily mobile in the lateral plane but not cranio-caudally (positive Fontaine sign). She had no cranial nerve fall-out and no constitutional features to suggest tuberculosis. A computerised tomographic angiogram (CTA) was performed and revealed a 6 cm highly vascular tumour splaying the carotid bifurcation with near total encasement of the external carotid artery (ECA) and partial encasement of the internal carotid artery (ICA). A diagnosis of a Shamblin III CBT was made based on the above imaging findings.

We offered her a left CBT excision without prior embolisation or radiotherapy. The neck mass was approached through a longitudinal incision anterior to the sternocleidomastoid muscle and both proximal and distal control was attained. The CBT did not extend to the skull base but the ECA was totally encased by the tumour with near total encasement of the ICA. The vagus and hypoglossal nerves were identified and protected. The tumour extent necessitated ECA excision but the ICA was spared with no requirement for interposition grafting.

Her postoperative course was complicated by a transient hypoglossal nerve palsy which was fully rehabilitated by speech therapy at the time of discharge. Histological examination of the excised mass was in keeping with a CBT/chemodectoma with an associated high mitotic index and significant necrosis. However, in the absence of local invasion or metastases, it was reported as benign.

She was lost to follow-up and re-presented four years later with a rapidly growing recurrent neck mass of 3-months duration. During that period, she was diagnosed as HIV positive and placed on anti-retroviral therapy (current CD4 count 339 with an undetectable viral load). Clinically, she had a large, fixed, non-pulsatile left neck mass with associated hypoglossal and marginal mandibular facial nerve palsy. On intra-oral examination she had an associated left tonsillar fungating lesion. A duplex scan revealed a large heterogeneous non-vascular soft tissue mass which was confirmed on CT scan with significant paratonsillar and pharyngeal involvement.

A transoral tonsillar biopsy reported a high-grade, diffuse large B-cell lymphoma and she was referred to the oncology department for chemotherapy but unfortunately defaulted her vascular clinic reviews.

Figure 1. Recurrent Left Neck Tumour
Carotid body tumours (CBTs) are exceptionally rare extra-adrenal neuro-endocrine neoplasms derived from neural crest tissue. Together with jugular, vagal and tympanic nerve derived neoplasms, they are collectively referred to as head and neck paragangliomas. The occurrence of CBTs is generally sporadic but hyperplastic and familial/syndromic types are occasionally described. Hyperplastic CBTs occur in high altitude populations and patients with chronic pulmonary disease. These paragangliomas have a collective incidence of 1 in 30 000 to 1 in 100 000, with CBTs accounting for 60–80% of these tumours. Most case series report a higher female preponderance with a male: female ratio of 1:2. Approximately 60% of these tumours occur on the right and 17% are bilateral.

Familial/syndromic cases are seen in specific hereditary succinyl dehydrogenase mutations (SDH-B, C & D), Carneys complex and Neuro-fibromatosis. Open surgical excision remains the gold standard treatment of CBTs with or without preoperative embolisation depending on institutional practices. Carotid body tumour excision carries a very low “true” recurrence rate (0.5–3%) and very few have reported on same site recurrent neck masses not due to recurrent CBTs. O’Neill et al. reported their 22-year Northern Irish Experience with CBTs. They had only 1 recurrence in 29 patients. Similarly, Sajid et al. in a multicentre review of CBT management reported a recurrence rate of 4.1% with a median follow-up of 1 year. Whether these CBT recurrences were due to malignancy or incomplete resection is not entirely clear in these reports. There are only two cases described in the literature that reported on CBT recurrences associated with ipsilateral cervical lymph node metastases. Such recurrences are associated with familial and syndromic CBTs. Germline mutations of the genes for SDH subunits B, C and D have been extensively researched. The SDH-B subunit mutation is associated with malignant CBTs whereas, SDH-D subunit mutation is associated with multicentric head and neck paragangliomas.

Here we describe the development of a lymphoma after CBT excision. A similar case of a 56-year-old man has been described in the literature. The patient was diagnosed with a 2 cm CBT and a synchronous 5 cm vagal paraganglioma. He was noted to harbour an SDH-D mutation in keeping with his multi-centric disease. Five years after surgical excision he developed a diffuse large B-cell lymphoma with no additional alteration in SDH-D miss-sense mutation. They concluded that the SDH-D mutation could have played a limited role in his lymphoma genesis. Such conclusions cannot be drawn out of our case as it seemed to be a sporadic CBT, and SDH mutation testing is not routine in our practice. Lymphoma genesis in our patient could also be linked to the patient’s human immunodeficiency virus infection. The aetiopathogenesis of HIV associated lymphomas is complex. Factors purported to be involved are the HIV virus, immuno-compromise, chronic antigen stimulation and in some cases synergistic viral infections, most commonly Epstein-Barr virus (EBV). The remaining cause of a recurrent ipsilateral neck tumour is a vein graft false aneurysm which has been described as a cause of a neck mass seven years after a CBT excision.
This is also a very rare occurrence with only 2 such cases described in modern literature.
We propose that recurrent neck masses following CBT excision be classified as follows:

- **Type 1:** True CBT recurrences
- **Type 2:** Non-chemodectoma paragangliomas (multi-centric disease)
- **Type 3:** Metastatic lymphadenopathy
- **Type 4:** De novo neck lymphomas
- **Type 5:** Vascular neck masses (pseudoaneurysms)

**Conclusion**

The patterns and behaviour of CBTs are very difficult to establish due to their very low incidence. More robust data can be obtained if multicentre databases are set up with standardised reporting guidelines and follow-up schedules. Where clinically relevant, SDH testing should be utilised to better predict patterns of presentation and probabilities of recurrences, and imaging should guide need for biopsy.

**REFERENCES**