Syncope associated with pain as the presenting feature of neck malignancy: failure of cardiac pacemaker to prevent attacks in two cases

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Two patients are described in whom syncope was the presenting clinical feature of an undiagnosed neck malignancy. Both patients also had attacks associated with paroxysms of severe neck pain. Neither patient responded to cardiac pacing.

Recurrent syncope as a complication of recurrent neck malignancy is an uncommon but well documented association. However, syncope as the presenting feature of previously undiagnosed neck malignancy is exceedingly rare. The exact mechanism of the syncope is uncertain; however, a suitably placed neck mass is presumed to cause irritation of afferent fibres from the carotid sinus and body, travelling first in Hering’s nerve and then in the glossopharyngeal nerve. Abnormal excitation of these afferent fibres causes excessive vagal activity. Syncope occurs because of bradycardia or asystole and usually prominent vasodepressor hypotension. The mechanism of syncope is therefore similar to that of idiopathic carotid sinus hypersensitivity, which consists of both cardioinhibitory and vasodepressor components in varying proportions.

We describe two patients in whom syncope was the presenting clinical feature of an undiagnosed neck malignancy. Both patients also described attacks associated with paroxysms of severe neck pain.

CASE 1
A 59 year old man attended his local hospital emergency department having lost consciousness for two minutes after a brief episode of sharp left sided neck pain while driving. Physical examination, chest x ray, and brain computed tomography (CT) were normal. He was discharged home.

He continued to experience occasional left ear pain until two months later, when he had two further blackouts preceded by a similar pain. During the second of these, an ambulance crew reported an unrecordable blood pressure with a pulse rate of 20 beats/min which responded to intravenous atropine.

On admission to hospital, he described a three month history of intermittent left sided tongue numbness, a change in voice quality, dysphagia, and 4 kg weight loss.

In hospital, he suffered a further episode of sharp neck pain, beginning in the left ear spreading to the left side of the neck and throat, lasting for 30 minutes, when the patient began to feel faint. This was followed by sudden loss of consciousness and urinary incontinence with a blood pressure of 90/60 mm Hg and pulse 30 beats/min. He recovered consciousness spontaneously after 30 seconds.

A temporary pacing wire was placed but he had a third episode with blood pressure falling to 77/40 despite a satisfactorily paced heart rate. He was subsequently transferred to this hospital. There was no other relevant medical history.

Physical examination revealed a left posterior pharyngeal mass, a left 12th nerve palsy, and a left Horner’s syndrome. A dual chamber permanent pacemaker was implanted. However, he suffered two further attacks, with no clear precipitant, during which the pulse rate was normal but blood pressure fell to systolic value of 70 mm Hg, each time with spontaneous recovery. CT revealed a large mass in the left parapharyngeal region, the upper end of which involved the carotid sheath (fig 1, panels A and B). There was bilateral deep cervical and submental lymphadenopathy. Angiography showed that the internal carotid artery was displaced anteriorly and medially by the tumour and the left internal jugular vein was occluded. There was no abnormal tumour vascularity.

Autonomic function tests after pacemaker insertion revealed no postural hypotension, and neck/head movements did not precipitate hypotension or syncope. Blood pressure responses to isometric exercise, mental arithmetic, cutaneous cold, hyperventilation, and Valsalva manoeuvre were normal. Carotid sinus massage was felt to be unsafe and was not done.

The patient underwent fine needle aspiration followed by Trucut biopsy. The histology was consistent with a high grade B cell lymphoma. He was started on carbamazepine and subsequently received chemotherapy, after which the carbamazepine was discontinued with no further episodes of syncope. He remains in remission from his lymphoma two years later.

CASE 2
A 71 year old man described an overnight change in his voice, becoming hoarse and strained. He also developed difficulty in swallowing with occasional coughing after swallowing. Two days later he developed a headache affecting the left side of his head, predominantly posteriorly and radiating to his neck. The pain was constant and worse with sitting up but improved over several weeks.

Three months later, he experienced sudden onset of a similar but more severe headache after which he vomited and lost consciousness for approximately two minutes. There were no other prodromal symptoms. He recovered without focal weakness or confusion, and there were no abnormal movements, incontinence, or tongue biting. He was admitted to his local hospital.

On admission, he was in sinus rhythm with a heart rate of 68 beats/min and his blood pressure was 155/70 mm Hg lying and 102/58 standing. Neurological examination was normal.

Abbreviations: CSM, carotid sinus massage; CSS, carotid sinus syndrome
After admission, he had one further episode of feeling faint and he collapsed but without loss of consciousness. His blood pressure was recorded as 75/0 and a cardiac monitor showed sinus bradycardia with prolonged pauses up to eight seconds (fig 2) which lasted for seven minutes. A permanent demand pacemaker was fitted. A few hours after the procedure, he had a further collapse with blood pressure unrecordable and pulse of 80 beats/min. CT of the head and neck was normal. He was discharged but had five further collapses without loss of consciousness while sitting or standing, some of which were associated with headache and neck movements failed to induce hypotension or syncope. Carotid sinus massage was not done for safety considerations. Fine needle aspiration of the mass followed by Trucut biopsy revealed a poorly differentiated squamous cell carcinoma. He was referred to his local radiotherapy service.

Shortly after admission, he had one further episode of feeling faint and he collapsed but without loss of consciousness. His blood pressure was recorded as 75/0 and a cardiac monitor showed sinus bradycardia with prolonged pauses up to eight seconds (fig 2) which lasted for seven minutes. A permanent demand pacemaker was fitted. A few hours after the procedure, he had a further collapse with blood pressure unrecordable and pulse of 80 beats/min. CT of the head and neck was normal. He was discharged but had five further collapses without loss of consciousness while sitting or standing, some of which were associated with headache and neck movements failed to induce hypotension or syncope. Carotid sinus massage was not done for safety considerations.

DISCUSSION

Syncope as a result of recurrent head and neck cancer has been reported quite often. However, the possibility that syncope could be a presenting feature of an occult tumour is unlikely to be considered by most physicians, especially in the absence of other signs related to local tumour infiltration. Very few cases of this association have been reported. In our case 2, the patient had no focal neurological signs at first presentation and the initial CT of the neck was normal. Both cases had syncope with documented bradycardia/asystole with hypotension, which was not abolished by cardiac pacing. Both were subsequently found to have malignant tumours in the parapharyngeal region. Neither patient had a previous diagnosis of malignancy. Both cases had prominent unilateral head and neck pain.

The exact mechanism of syncope in head and neck malignancy is not well characterised, but is probably similar to that of the carotid sinus syndrome (CSS). In CSS, syncope is painless, often triggered by head movement, and carotid sinus massage (CSM) may be diagnostic. Weiss and Baker described three forms of CSS: the cardioinhibitory type, the vasodepressor type, and the rare cerebral type. By contrast, syncope from head and neck cancer usually differs from CSS in several respects. In the case of underlying neck malignancy, while clear triggers may be identifiable, the attacks are usually spontaneous and CSM does not characteristically provoke syncopal attacks. Although excessive vagal tone and consequent cardioinhibition are characteristic of both CSS and malignant syncope attacks, vasodepressor effects are thought to be much more prominent in the latter. Deafferentation of the carotid sinus by tumour invasion may account for the inability to provoke...
attacks by CSM in some cases of neck malignancy. It may be
that paroxysms of afferent neural activity occur as a result of
ephaptic transmission generated in the afferent nerves by the
invading tumour.

Both of our cases had cervical lymph node involvement but
no direct involvement of the region of the carotid sinus. Therefore, we hypothesise that the combination of pressure
on the left carotid sinus and direct injury to or irritation of
Hering’s nerve or the glossopharyngeal nerve in our cases
resulted in prominent vasodepressor effects in association
with vagal hypersensitivity with head and neck malignancy:

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