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An unusual cause of carotid sinus hypersensitivity/syndrome

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Syncope is extremely common, especially in the elderly population. Although neurocardiogenic (vasovagal) syncope accounts for ≥35% of cases, carotid sinus hypersensitivity (CSH) is a notable, albeit rarer, cause of reflex syncope.

We present an unusual case of CSH resulting from malignancy, representing a diagnostic and therapeutic challenge.

Case report

A 78-year-old gentleman presented acutely via ambulance following an unprovoked and unwitnessed cardinal episode of syncope whilst driving. He had no known medical conditions and was not on any regular medications. He was highly functioning and still worked as a freelance accountant. He had a 50-year history of daily pipe smoking. In retrospect, he reported a mild difficulty in swallowing and a 5-kg weight loss over the preceding 6 weeks.

Physical examination was unremarkable except for palpable cervical lymph nodes. Preliminary electrolytes, thyroid function, resting electrocardiography and echocardiography were all normal. He had no orthostatic hypotension. He experienced another unprovoked syncopal episode in hospital associated with bradycardia (and preceded by 6 second pause) and hypotension. He had a positive controlled carotid sinus massage (CSM) test.

He also had computed tomography imaging of his head and neck, which showed an invasive malignancy in the right tonsilllar fossa associated with significant necrotic lymphadenopathy affecting the neck bilaterally (Figures 1A and 1B). The nodes were directly anterior to, and encroaching on, the carotid sheath at multiple levels including the carotid sinus. There were also multiple pulmonary nodules but no other metastatic spread. Histology from a subsequent core biopsy confirmed squamous cell carcinoma.

A Medtronic dual chamber (VVI) pacemaker was implanted. Due to the extent of his malignancy (Stage T4a N2c M1), he was only amenable to palliative radiotherapy with 30 Gy administered in 10 fractions with small clinical improvement in his lymphadenopathy.

Despite significant reduction in syncope frequency, he still experiences rare events associated with hypotension but no bradycardia.

Figure 1A. Coronal view showing the relationship of necrotic cervical lymph nodes (white arrows) in relation to bilateral external and internal carotid arteries (black arrows)

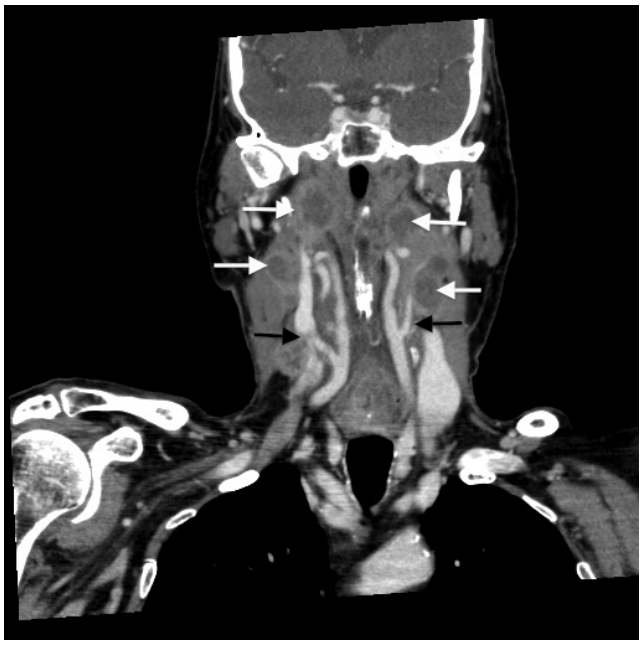
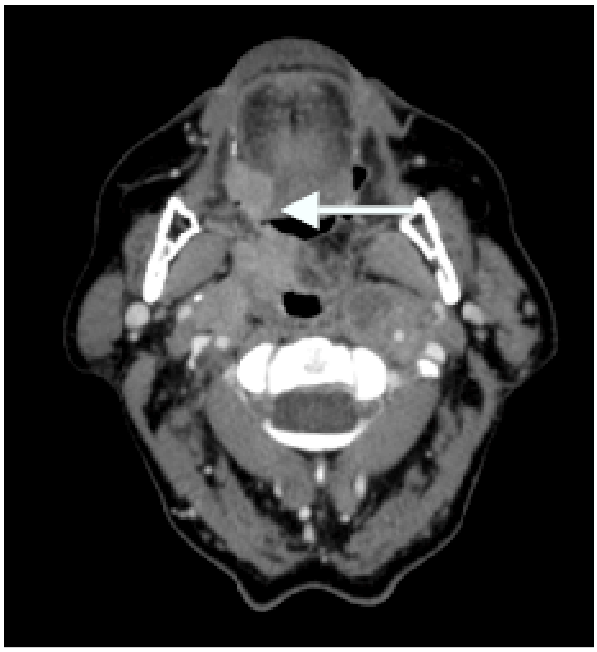


Figure 1B. Transverse view of the primary mass in the right tonsillar fossa (white arrow) and involvement of the palate



Discussion

CSH results from exaggerated responses to carotid sinus stimulation; cardioinhibitory resulting in bradycardia (70–75%), vasodepressor with reduction in vasomotor tone (5–10%), or a mixture of both (20–25%).[1,2] Regardless, they all culminate in reduction of cardiac output.[1,2]

CSH is more common in males and accounts for 1–5% of recurrent syncope (uncommon <50 years, incidence increases with age).[3] CSH from primary or metastatic malignancies (direct carotid sinus pressure) is uncommon.[4–6]

Diagnosis requires exclusion of all other causes of syncope in addition to a positive CSM test.[2-3] A pause (asystole) of >3 seconds (cardioinhibitory), systolic blood pressure drop of ≥50 mmHg (vasodepressor) or both (mixed) constitutes a positive test.[2,3] It is recommended that CSM is done in all patients >40 years presenting with recurrent syncope (and without absolute contraindications).[3]

Pacemaker implantation (DDD, DDI or VVI) is effective in reducing syncope frequency in cardioinhibitory and mixed CSH in most cases, but intuitively, not in vasodepressor mediated CSH.[7,8] The vasodepressor effect may well be difficult to manage though epinephrine has been trialled with limited success[4] Midodrine, fludrocortisone and sertraline were previously trialled by Sharma et al without any success.[6]

This case highlights the importance of high clinical suspicion and thorough clinical examination in the diagnostic workup of unexplained syncope, especially in the elderly, as not all cases are due to vasovagal or orthostatic causes. Additionally, this case highlights the fact that there is no curative therapy for CSH since syncopal episodes may still occur following pacemaker implantation. This is especially true in incurable malignancy-related CSH (especially with predominant vasodepressor response), representing an intractable and difficult therapeutic challenge.

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References:

1. Weiss S, Baker JP. The carotid sinus reflex in health and disease: its role in the causation of fainting and convulsions. Medicine 1933;12:297–354.
2. McIntosh SJ, Lawson J, Kenny RA. Clinical characteristics of vasodepressor, cardioinhibitory, and mixed carotid sinus syndrome in the elderly. Am J Med 1993;95:203–8.
3. Moya A, Sutton R et al. Guidelines for the diagnosis and management of syncope (version 2009). Eur Heart 2009: Nov; 30(21):2631–71.
4. Patel AK, Yap VU, Fields J, Thomsen JH. Carotid sinus syncope induced by malignant tumors in the neck. Emergence of vasodepressor manifestations following pacemaker therapy. Arch Intern Med. 1979 Nov;139(11):1281–4.
5. Campbell S, Walker D, Lanzon-Miller S, Gurr P. Nasal carcinoma--a rare indication for a permanent pacemaker. Clin Med. 2004 Mar-Apr;4(2):165–7.
6. Sharma J, Dougherty AH. Recurrent syncope in a cancer patient: a case report and review of the literature. Cardiol Res Pract. 2011 Apr 4; 2011:678237.
7. Claesson JE, Kristensson BE, Edvardsson N, Währborg P. Less syncope and milder symptoms in patients treated with pacing for induced cardioinhibitory carotid sinus syndrome: a randomized study. Europace. 2007 Oct;9(10):932-6. Epub 2007 Sep.
8. Sutton R, Brignole M, Menozzi C, et al. Dual-chamber pacing in the treatment of neurally mediated tilt-positive cardioinhibitory syncope: pacemaker versus no therapy: a multicenter randomized study. The Vasovagal Syncope International Study (VASIS) Investigators. Circulation. 2000 Jul 18;102(3):294–9.