An Unusual Cause of Reflex Cardiovascular Syncope: Vagal Paraganglioma

Ertan Okmen, M.D., Izzet Erdinler, M.D., Enis Oguz, M.D., Ahmet Akyol, M.D., and Nese Cam, M.D.

From the Department of Cardiology, Siyami Ersek Cardiovascular and Thoracic Surgery Center, Istanbul, Turkey

Vagal paraganglioma is one of the rare tumors of the neuroendocrine system. We are reporting a vagal paraganglioma case presented with uncommon features of the disease: asystole and syncope. Syncope episodes occurred 3 years before the major symptoms of the disease. Dual chamber pacemaker failed to prevent syncope attacks because of the vasodepressor component. The patient was treated successfully with en bloc removal of tumor and vagal nerve. Syncope episodes disappeared after operation.

A.N.E. 2003;8(2):173-176

Vasodepressor-cardioinhibitor syncope; asystole; vagal paraganglioma; dual chamber pacemaker

The extra-adrenal neuroendocrine system is a complex system dispersed throughout the body with a regulatory function. Vagal paraganglioma (VP) originates from the paraganglia cells within the vagal nerve and is the third most common paraganglioma of the head and neck after carotid body tumors and jugulotympanic paragangliomas.¹ They account for less than 5% of all head and neck paragangliomas.^{1,2} The tumor typically manifests as a painless, slowly growing mass in the neck.^{3,4} We are reporting a VP as an unusual cause of syncope and asystole. According to the authors' knowledge this is the unique VP case presenting with syncope episodes at the early stage of the disease.

CASE

A 41-year-old man, who was healthy until December 1996 when he began having episodes of loss of consciousness lasting less than 5 seconds, was admitted to our hospital. These episodes were nonexertional and almost always associated with a feeling of slight squeezing sensation in his epigastric region. There was no known precipitating factor. The frequency of the episodes was five to six times a month with less than 5-second duration in the beginning and increased approximately to 20 seconds when he was admitted to hospi-

tal in 1996. Examination of head, eyes, nose, and throat was within normal limits. ECG, echocardiography, carotid Doppler, and exercise stress tests were found to be within normal limits. During his first admission in 1996, electroencephalography and computerized tomography (CT) of brain and neck were found to be normal. Passive tilt table test revealed progressive orthostatic hypotension followed by bradycardia, 3-second asystole, and vasovagal syncope. Forty-eight-hour Holter recording revealed seven runs of paroxysmal bradycardia, and 17 pauses with the longest being 6.1 seconds (Fig. 1). Ischemic ST changes were not evident during symptoms, and activation of patient event button correlated with pauses and bradycardia. According to these findings a DDDR pacemaker was implanted in January 1997. Severity and frequency of syncope episodes diminished to one to two times a month, and the duration of attacks reduced following pacemaker implantation, but they did not completely disappear during the follow-up period of 3 years. Syncope episodes were associated with hypotension without bradycardia with appropriate activation of the pacemaker. After 3 years of pacemaker implantation, frequency of syncope increased to four to six times a month. The patient complained of episodic light-headedness associated with bradycardia, and syncope that could be elicited by turning the head to the left. Physical

Figure 1. Asystole attacks during 48 hours Holter recording. The longest one is 6.1 seconds on strip 2.

examination revealed a mass on the right upper side of the neck. Bradycardia and hypotension were induced by palpation of the mass. CT of brain and neck was repeated to seek the origin of the mass. CT revealed a right carotid space mass located between carotid artery bifurcation and jugular foramen of the size of 4.0×2.5 cm with smooth margins and enhancement (Fig. 2). Surgical resection was performed via lateral cervical approach. Mass and vagus nerve were extracted en bloc. Tumoral pleomorphism and insular pattern of paraganglioma cells with chromogranin positive granulated cytoplasm were observed in histopathologic analysis of the mass (Fig. 3). Syncope episodes completely disappeared after operation. The patient is still on

follow-up without any complaint at postoperative 16th month.

DISCUSSION

Paraganglioma of the vagus nerve has not been reported as a cause of asystole and syncope attacks. The most frequent symptom of VP is a slowly growing, painless lateral neck mass that is most commonly located behind the angle of the mandible.^{3,4} Other symptoms are hoarseness, pharyngeal fullness, dysphagia, dysphonia, cervical pain, hearing loss, vertigo, cough, and aspiration.^{4,5} VPs are usually benign, slow-growing neoplasms associated with low morbidity and mortality.^{4,6} Miller et al.

Figure 2. Computerized tomography view of the right carotid space mass.

Figure 3. Tumoral pleomorphism and insular pattern of paraganglioma cells with chromogranin positive (neuroendocrine) granulated cytoplasm (HE $\times$ 400).

showed that the average duration of symptoms before presentation was 40 months (range 5–72 months).⁴ In our case, the time between the syncope attacks and the manifestation of left neck mass was approximately 3 years.

Several vagal neoplasms associated with bradycardia and syncope including schwannoma and neurilemmoma have also been reported.^{7–9} Severe bradycardia and cardiac arrest during surgical removal of these tumors have been attributed to direct vagal stimulation with manipulation of the tumor.

Vagus nerve is the afferent and efferent limb for cardiovascular reflexes.¹⁰ Cardiovascular reflexes with vagal efferents produce an inhibition of blood pressure and/or heart rate through vagal effects, which result in inhibition of sympathetic activity to the peripheral vasculature. Cicogna et al. described a group of patients presented with a pathological involvement of the parapharyngeal space, frequently by a malignant tumor and suffering from prolonged, severe, and relapsing vasodepressor syncope.¹¹ They have assumed that the compressive expansion of a tumor in the parapharyngeal space may play a role in vasovagal syncope. Glassopharyngeal nerve constitutes the main afferent nerve pathway, and vagus nerve the efferent pathway in the pathophysiology of this kind of syncope. Mass may abnormally stimulate the peripheral branches of the glassopharyngeal nerve, and possibly coincidentally the adjacent fibers of the Xth nerve. Interestingly, the intracranial section of the IXth nerve may relieve the symptoms in these cases.

Demand ventricular or even AV sequential pacemakers do not change the clinical picture of the vasodepressor syncope. Although there was a cardio-

inhibitor component in our patient, the DDDR pacemaker could not prevent the syncope episodes completely, due to the presence of the significant vasodepressor component.

We conclude that in our case, even during the early stages of the disease, an abnormal discharge from the vagal nerve or stimulation of the peripheral branches of the glassopharyngeal nerve by the mass possibly caused an intense vasovagal reaction characterized by reflex cardiovascular syncope.

REFERENCES

1. Davidson J, Gullane P. Glomus vagale tumors. *Otolaryngol Head Neck Surg* 1988;99:66–70.
2. Murphy TE, Huros AG, Frazell EL. Chemodectomas of the intravagale: Vagal body tumors, nonchromaffin paragangliomas of the nodose ganglion of the vagus nerve. *Ann Surg* 1970;172:246–255.
3. Batsakis JG. Tumors of the Head and Neck: Clinical and Pathological Considerations, 2nd Edition, Baltimore, Williams & Wilkins, 1979, pp. 369–380.
4. Miller RB, Boon MS, Atkins JP, et al. Vagal paraganglioma: The Jefferson experience. *Otolaryngol Head Neck Surg* 2000;122:482–487.
5. Eriksen C, Girdhar-Gopal Harsha, Lowry L. Vagal paragangliomas: A report of nine cases. *Am J Otolaryngol* 1991;12:278–287.
6. Leonetti JF, Brackmann DE. Glomus vagal tumor: The significance of early vocal cord paralysis. *Otolaryngol Head Neck Surg* 1989;100:533–537.
7. Wood BM, McNeil WT. Schwannoma of the vagus nerve. *Anaesthesia* 1986;41:1130–2.
8. Mukherjee DK. Neurilemmoma of the vagus nerve: A case report. *J Laryngol Otol* 1979;93:187–192.
9. Sawamura Y, Tribolet N. Vagal schwannoma associated with syncopal attacks and postural hypotension: A case report. *Neurosurgery* 1990;27:461–463.
10. Abboud FM. Integration of reflex response in the control of blood pressure and vascular resistance. *Am J Cardiol* 1979;44:903–911.
11. Cicogna R, Bonomi FG, Curnis A, et al. Parapharyngeal space lesions syncope syndrome: A newly proposed reflexogenic cardiovascular syndrome. *Eur Heart J* 1993;14:1476–83.