



Sporadic bilateral carotid body paragangliomas

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DESCRIPTION

A 57-year-old woman presented with complaints of swellings on both sides of her neck for the past 4 years which were gradually increasing in size. They were painless and she did not complain of any difficulty in breathing or swallowing. She had no complaints of blushing or palpitations in the past. She did not have any history of residency in hilly areas or exposure to chronic hypoxic conditions. There were no other swellings in the body.

On examination, a 3×2 cm hard, round and mobile swelling was noted on the right side and a 2×3 cm hard, mobile swelling on the left side. Vertical mobility of both masses was restricted compared with horizontal mobility (Fontaine sign).¹ Oral examination did not reveal any pharyngeal wall swelling. Cranial nerve examination was normal. Other system examinations were unremarkable.

No abnormalities were noted on chest X-ray or ECG. Duplex ultrasound of the neck revealed vascularised hypochoic masses at the carotid artery bifurcation on either side. Intravenous contrast-enhanced CT of the neck and cerebral region showed bilateral, intensely enhancing soft tissue density lesions in (both) carotid space of 4×3×3 cm in size in the left carotid space and a similar lesion of 1.5×1.5 cm in size in the right carotid space (figure 1A–C). MRI of the head and neck showed lesion in both the carotids with characteristic salt and pepper appearance (figure 1D).

Abdominal ultrasonography, urine vanillylmandelic acid, and plasma epinephrine and norepinephrine levels were normal. There was no suggestive family history of neck masses. No further genetic evaluation could be done due to unavailability of such facilities in the hospital. The patient was taken up for excision of bilateral carotid body lesions under general anaesthesia and the postoperative specimen was sent for histopathological examination, which

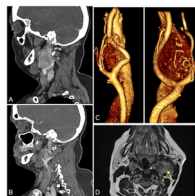


Figure 1 (A) CT angiography showing splitting of both carotids due to left carotid paraganglioma. (B) CT angiography showing splitting of both carotids due to right carotid paraganglioma. (C) Three-dimensional reconstruction of both the carotid body paragangliomas. (D) T1-weighted MRI showing salt and pepper appearance (arrow) of left carotid body paraganglioma.

Patient's perspective

I am an agricultural laborer and I don't often feel sick. I was having swelling in the sides of neck for last four years, initially only in the left, and then also in the right. They were slowly growing. When the right sided swelling appeared in the last 6 months, I took medical advice from the primary health centre, and I was referred to the present hospital where Dr. Anandhi has examined me. Following some scans, the doctor informed me that I had a tumor sort of a thing in my neck region and that is the reason for swelling. I was worried that it might be cancer and I was in serious trouble. But immediately the doctor had reassured me that it was a localized lesion and the chances of the disease being spread to other areas is very less. She said we will have to do two more tests to know more about the disease. One test was longer than the other test and I was worried a little at the loud noises the machine had made during the scan. The technician was talking to me and helped me keep calm during the procedure. After the report, Dr Anandhi has said that the diagnosis was nearly confirmed. I was explained about the surgical procedure and the risks associated. I gave my consent to the procedure. After I regained consciousness after the surgery I saw dressings over neck. I had some pain and discomfort on the first day. I recovered fast and I was discharged after 3 days. I was overwhelmed by the care given by the doctors and nursing staff in the hospital and thank them all.

Learning points

- ▶ Occurrence of sporadic bilateral carotid body paragangliomas is rare.
- ▶ Preoperative fine needle aspiration cytology of the tumour is discouraged owing to its highly vascular nature.
- ▶ CT angiography is sufficient to establish the diagnosis and surgical excision with or without vascular reconstruction.

confirmed the diagnosis of carotid body paragangliomas. The postoperative period was uneventful, and the patient was discharged in a stable condition.

Paragangliomas of bilateral carotid glands account for 3% of all paragangliomas in the body.² They manifest as slow-growing, painless masses located mostly at the hyoid bone level, anterior to



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the sternocleidomastoid muscle. Occasionally carotid bruit can be heard, which was absent in our case. Most of the sporadic cases occur as unilateral masses, but familial tumours tend to occur bilaterally. The vascular nature of this tumour stands as a potential risk for an uncontrolled bleeding if an incisional biopsy is attempted. CT angiography forms the mainstay of diagnosis of carotid body tumour, which shows a mass at the carotid artery bifurcation that causes splaying of the internal and external carotid arteries (Lyre sign). This should be differentiated from the pseudo-Lyre sign, which is caused by paraganglioma arising from the vagus nerve.³

Preoperative embolisation with polyvinyl alcohol and gel foam can be used for highly vascular tumours larger than 5 cm.⁴ However, a few reports state an increased incidence of complications such as cerebral ischaemia and cranial nerve palsies with this embolisation.⁵ Surgical excision of the tumour with vascular reconstruction is the main treatment modality in these cases. In cases of recurrence or large malignant lesions, radiotherapy can also be considered as a modality of treatment.⁶

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