Functional, malignant intrathoracic paraganglioma

Praveen Aggarwal¹, Jyoti Prakash Wali¹, Panangapalli Venugopal², Anil Khosla³ and Satish Kumar Tickoo⁴

Departments of ¹Medicine, ²Cardiothoracic and Vascular Surgery, ³Radiology and ⁴Pathology, All India Institute of Medical Sciences, New Delhi – 110029, India.

Summary: This paper describes a case of functional, malignant branchiomeric paraganglioma, the third such to be reported. The patient presented with malignant hypertension and symptoms suggestive of excessive catecholamine secretion.

Introduction

Paragangliomas, the tumours of the paraganglion cells, occur most commonly in the adrenal medulla. These tumours may also occur in carotid body, glomus jugulare, vagus nerve, larynx, lungs, retroperitoneum and bladder. Mediastinal paragangliomas are of rare occurrence. We describe a case of functional, malignant branchiomeric paraganglioma.

Case report

An 18 year old girl was admitted with a 2-month history of swelling in the right supraclavicular area. She had been empirically treated with antitubercular drugs elsewhere. One week later she developed visual blurring in both the eyes and the antitubercular treatment was stopped. Examination of the fundus at that time had revealed bilateral papilloedema; however, a computerized tomography (CT) scan of the brain was normal. Three weeks prior to admission, a biopsy of the supraclavicular mass was done without any apparent complications. This was reported as 'possible chemodectoma'; the slides, however, were not available for review. At this time, her blood pressure was noted to be 170/110 mmHg. In the week preceding admission, she had four episodes of headache, palpitations, sweating and dyspnoea. During one of these episodes, her blood pressure was recorded as 170/130 mmHg.

Examination at the time of admission revealed a pulse rate of 136/min and a blood pressure of 190/120 mmHg. Examination of the eye fundi showed

bilateral haemorrhages, macular oedema and papilloedema.

Investigations revealed a normal haemogram, blood chemistry and routine urinalysis. The 24-hour urinary catecholamines and vanillyl mandelic acid (VMA) levels were 2,000 µg and 43.5 mg respectively. The ultrasonography and the CT scan of the abdomen and pelvis were normal. A penetrated chest X-ray showed a soft tissue mass in the superior mediastinum displacing the trachea to the left (Figure 1). An X-ray of the cervical spine showed a collapse of the fourth cervical vertebra suggestive of a metastatic deposit. A CT scan of the chest disclosed a large moderately enhancing soft tissue mass in the superior mediastinum. A TC-90m EHDP bone scan revealed hot spots in the ninth right rib, sternum, second and third lumbar vertebrae and fourth cervical vertebra suggesting metastases at these sites.

With a diagnosis of malignant, functional intrathoracic paraganglioma, she was started on phenoxybenzamine (30 mg/day) and later, propranolol (60 mg/day) was added. On this treatment, her blood pressure and pulse rate were adequately controlled.

At thoracotomy, the mass was found to be located in the antero-superior mediastinum and was closely adherent to the superior vena cava, trachea and brachiocephalic vein. Because of the attachment of the tumour to the vital structures, only about two-fifths of it could be removed safely.

On microscopic examination, the tumour was composed of well defined nests of cuboidal cells separated by highly vascularized fibrous septae. The individual cells had a moderately abundant, somewhat granular cytoplasm (Figure 2). Some of the cells were shown to be positive for argentaffin granules by Fontana-Masson method. The adrenaline and noradrenaline

Correspondence: J.P. Wali, M.D., M.N.A.M.S.

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Figure 1 Radiograph of the cervico-dorsal junction showing a superior mediastinal mass.

Figure 2 Microscopic section of the tumour showing groups of cuboidal cells separated by highly vascularized fibrous septae. (H & E X 95).

contents in the tissue were 6.6 mg and 10.9 mg per gram of tissue respectively.

A final diagnosis of functional, malignant, branchiomeric (aorto-pulmonary type) paraganglioma was made.

Discussion

The extra-adrenal paraganglia and their tumours are classified into four categories on the basis of anatomical location, histochemical features and innervation. These are branchiomeric paraganglia (subdivided into orbital, jugulo-tympanic, intercarotid, subclavian, laryngeal, coronary, aorto-pulmonary and pulmonary paraganglia), intravagal paraganglia, aortico-sympathetic paraganglia and viscer-autonomic paraganglia. Functioning and non-functioning tumours may arise in any of these paraganglia. The term pheochromocytoma should be restricted to the paragangliomas arising in the adrenal medulla.¹

Since the first report of intrathoracic paraganglioma in 1924 by Miller,² about 152 cases of these tumours have been reported.³ Out of these 152 cases, 54 cases were of the aortico-sympathetic group, 8 were of the branchiomeric group and 12 were of the viscer-autonomic group. It has been found that the proportion of functioning tumours is much higher in the case of aortico-sympathetic tumours as compared to the branchiomeric tumours. Out of the reported cases of branchiomeric paragangliomas, only two were both functional and malignant.^{4,5} Another patient with malignant branchiomeric tumour had elevated urinary homovanillic acid levels but the patient did not have hypertension.⁶ The present case, therefore, is the third case of branchiomeric paraganglioma which is malignant as well as functional.

The treatment of malignant paragangliomas is still not well established. Radiotherapy and chemotherapy have been tried with varying results. Recently, ¹³¹I-meta-iodobenzylguanidine has been used in treating this tumour and the initial results are encouraging.

After surgery, this patient received radiotherapy to the cervical vertebral metastasis and, presently, she is on combination chemotherapy consisting of cyclophosphamide, vincristine and dacarbazine.⁷ The initial response is encouraging and her requirement of phenoxybenzamine has come down to 5 mg/day.

In contrast with the three patients with benign functional tumours reported by Dunn *et al.*,³ ours did not have a carotid body tumour.

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