Case report

Case of functioning thoracic paraganglioma

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Accepted 31 August 2020

To cite: Paul J, Jebasingh FK, Kodiatte TA, *et al*. *BMJ Case Rep* 2020;**13**:e236440. doi:10.1136/bcr-2020-236440

SUMMARY

Functioning thoracic paraganglioma (PGL) is rare in clinical practice. We present a 33-year-old man with this pathology, who came with right-sided chest pain and was found to have a right-sided paravertebral mass. Fine needle aspiration cytology revealed a PGL. Urine normetanephrine was elevated and meta- iodobenzylguanidine scan showed increased tracer uptake in the right hemithorax, suggestive of a functioning neuroendocrine tumour. The patient was subjected to right PGL excision by video-assisted thoracoscopic surgery, after adequate preoperative preparations. The perioperative period was uneventful, except for a transient rise in blood pressure during the surgery. His blood pressure continued to be normal in the postoperative period. In any patient with a paravertebral mass, the possibility of PGL should be kept in mind even if the patient is normotensive. Making a preoperative diagnosis is important, because excision of functioning PGL without adequate preoperative preparation may be detrimental.

BACKGROUND

Pheochromocytoma (PCC) and paraganglioma (PGL) are rare catecholamine-secreting tumours originating from chromaffin cells of the embryonic neural crest. They account for 0.05%–0.1% of patients with hypertension. Catecholamine-secreting tumours arising from adrenal glands are termed as PCC and those from extra-adrenal sites are termed PGL. Nearly 80%–85% of these tumours arise from adrenal glands and the rest are extra-adrenal in origin. PGL can arise from various sites in the body. Commonly, it occurs in abdomen in relation to organ of Zuckerkandl around the origin of inferior mesenteric artery. Thoracic PGL is very rare and is often non-functional. Herewith, we report a rare case of functioning thoracic PGL and review the relevant published literature.

CASE PRESENTATION

A 33-year-old man, with no significant history, presented with persistent, right-sided dull aching chest pain of 6 months duration, with no associated symptoms like cough, haemoptysis, fever or dyspnoea. He denied history of headache, palpitation or sweating. There was no family history of early onset hypertension, early onset stroke or sudden deaths. He was referred to our hospital, as he was found to have an incidental right-sided intrathoracic paravertebral mass. He was a moderately built person with no neurocutaneous markers or marfanoid habitus. His blood pressure was 130/80 mm Hg with no postural hypotension, and had no evidence of hypertensive retinopathy.

INVESTIGATIONS

The ECG was normal and the echocardiogram did not show left ventricular hypertrophy. Chest X-ray showed a well-defined round lesion projected over the right hilum of lung (figure 1A). The contrast-enhanced computed tomogram of the thorax showed a well-defined enhancing lesion measuring 5×3.8×3.2 cm in the right paravertebral region, extending from T1 to T7 vertebral body levels (figure 1B).

He underwent a CT-guided fine needle aspiration cytology (FNAC) of the mass, which showed tumour cells positive for chromogranin, synaptophysin and GATA3 suggestive of a PGL. He did not have any hypertensive crisis during or after the procedure. The urinary normetanephrine levels were elevated (2665 mcg/24 hours (normal <600 mcg/24 hours)). A meta-iodobenzylguanidine (131I-MIBG) scan showed increased tracer uptake in the right hemithorax suggestive of a functioning neuroendocrine tumour (figure 1C).

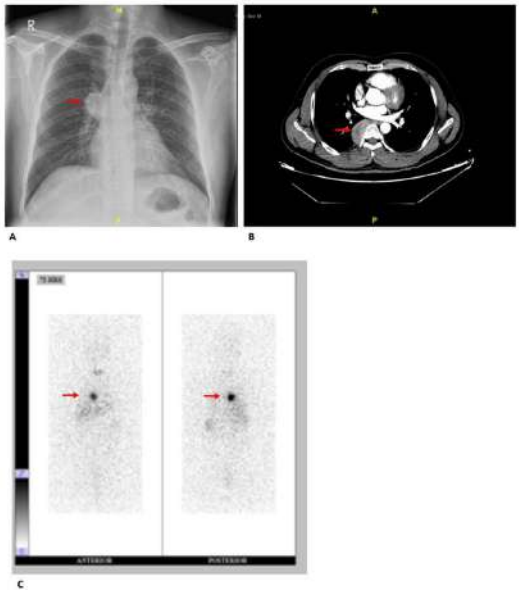

Figure 1 (A) Chest X-ray showing a well-defined round lesion projected over the right hilum of lung. (B) CECT scan of the thorax showing a well-defined enhancing lesion in paravertebral region. (C) MIBG scan at 72 hours showing increased tracer uptake in the right hemithorax suggestive of a functioning neuroendocrine tumour (shown with the arrows). CECT, contrast-enhanced computed tomogram; MIBG, meta-iodobenzylguanidine.

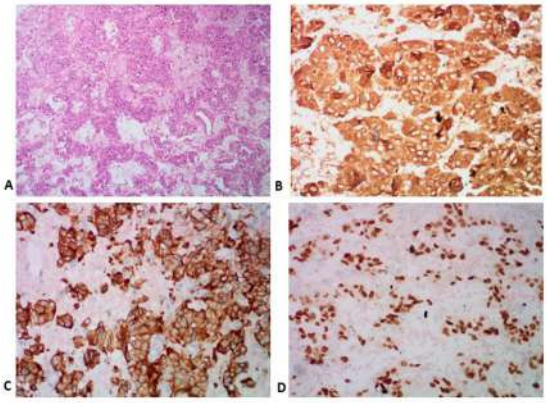

Figure 2 Microphotograph displaying (A) tumour cells arranged in nests and organoid pattern with intervening rich vascularity (H&E stain, 100× magnification), (B)–(D) expressing diffuse strong membranous and granular cytoplasmic immunopositivity for (B) chromogranin A, (C) synaptophysin and diffuse strong nuclear immunopositivity for (D) GATA-3 (immunohistochemistry, 400× magnification).

DIFFERENTIAL DIAGNOSIS

The FNAC was suggestive of PGL, urine normetanephrine was elevated and MIBG scan showed increased tracer uptake by the mass. In view of the above- mentioned results, we arrived at a diagnosis of functioning thoracic PGL.

TREATMENT

The patient was planned for the excision of the thoracic PGL and was optimised medically. He was started on extended release oral prazosin which was gradually up titrated to the maximum dose, with adequate oral salt and water loading. Preoperatively, he was also started on a betablocker metoprolol, to control the tachycardia. Complete excision of the tumour was done by a video-assisted thoracoscopic surgery. The patient was hypertensive during surgery and the blood pressure dropped to normal the moment the tumour was excised. Intraoperatively, a well-defined, well-encapsulated 5×4×3 cm lobulated tumour was seen in the right paravertebral region. The histopathology of the specimen showed a tumour composed of nests, organoid pattern and zellballen arrangement of polygonal cells with intervening rich vascularity (figure 2A) and on immunostaining was positive for chromogranin A, synaptophysin and GATA-3, suggestive of a PGL (figure 2B–D).

OUTCOME AND FOLLOW-UP

Genetic testing for succinate dehydrogenase (SDH) mutation, which is a common mutation documented with thoracic PGL was planned during the follow-up visit. His blood pressure continued to be normal during the postoperative period, without any antihypertensives.

DISCUSSION

PCC and PGL are rare catecholamine-secreting tumours arising from chromaffin cells of the neural crest. The most common aetiology for posterior mediastinal mass is neurogenic tumours such as schwannoma, neurofibroma. Thoracic PGL is rare in clinical practice. There are only 150 cases of mediastinal PGL reported in the literature.[1] Intrathoracic PGL is usually slow growing and hyper vascular tumours, having poor prognosis due to their locally invasive nature. In a study done by Fischer *et al*, of the 867 PGL, the primary site was central nervous system (39.9%), abdomen or pelvis (21.0%), head and neck (17.5%), thoracic (15.1%), bladder (3%) or unspecified (3.5%).[2] Surgical resection of the tumour is the best modality of treatment with complete excision providing the best long-term outcome. High index of suspicion is needed for preoperative diagnosis of thoracic PGL. Up to 50% of the mediastinal PGL are asymptomatic and are incidentally diagnosed.[3] In our patient, the tumour was secreting catecholamines, but the blood pressure was normal. All the functioning PGL require adequate preoperative alpha blockade to prevent intraoperative blood pressure surge. These patients need a long-term surveillance as they are known to recur.[4] A quarter to one-third of all PCC and PGL is known to have genetic aetiology.[4 5] SDH is a mitochondrial complex participating in both electron transport and the Krebs cycle. SDH mutations are seen in several familial clusters of PCC or PGL, which are described and defined as PGL syndromes PGL1 through PGL4.[6] Burnichon *et al* reported higher frequency of SDH-D and SDH-C mutations in head and neck PGL and SDH-B mutations in abdominal and pelvic PGL.[7] Both mediastinal and para-aortic PGL are usually associated with SDH-B or SDH-D mutations.[8] Some of these tumours, especially SDH-B-related tumours, carry a significant malignant potential. Biochemically, SDH-related tumour can be secretory or silent. Mutational analysis is recommended in all patients with head and neck PGLs, family members of SDH-related tumours, malignant PCC or PGL and also in early onset or multifocal disease.

Learning points

- PGLs are rare causes of paravertebral mass lesions.
- Functional thoracic PGLs can exist in normotensive patients.
- Making an accurate preoperative diagnosis and preparing the patient adequately for surgery will avoid perioperative complications due to catecholamine surge.

Acknowlegements We acknowledge the Department of Nuclear Medicine, Christian Medical College, Vellore for providing us with images of the meta-iodobenzylguanidine scan.

Contributors JP wrote the manuscript, KFJ, TAK and GBR provided critical inputs and corrections. All authors reviewed the final manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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