

Retroperitoneal Pheochromocytoma: A Rare Tumour Presenting as Ovarian Mass

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Abstract

A 40 year old patient suffering from vague pain in the abdomen since 1 month, came with CT scan of abdomen and pelvis showing large lobulated cystic lesion of 11 x 10.6 x 9.7cms suggesting ? Left ovarian neoplasm. On exploratory laparotomy uterus and ovaries were normal and a mass of 10 x 9 cm was seen arising from the pelvis. The Histopathological report stated pheochromocytoma. The extra adrenal pheochromocytomas are rare tumors that originate from the chromaffin tissue of sympathetic nervous system.

Key words: Retroperitoneal mass, pheochromocytoma, ovarian tumour.

Introduction

Pheochromocytomas (also known as paraganglioma) are functional and non-functional tumours of the sympathetic nervous tissue.^[1] Catecholamines are secreted by functional pheochromocytomas, resulting in paroxysmal hypertension and palpitation. The majority of the pheochromocytomas are derived from chromaffin tissue of the adrenal medulla.^[2] Extra-adrenal pheochromocytomas are rare tumours that originate from the chromaffin tissue of the sympathetic nervous system.^[2] In adults, pheochromocytomas are often called as 10% tumour because 10% occur along the diaphragm, while 10% of pheochromocytomas are extra-adrenal, 10% are bilateral, 10% are multiple, 10% are familial, 10% are malignant and 10% occur post operatively.^[3] Diagnosing pheochromocytomas is challenging due to their low incidence, and their clinical manifestations can be inconsistent. The present case report is an incidental finding of extra-adrenal non-functioning pheochromocytoma.

Case report

A 40 year old female was referred to Sri Siddhartha medical college with history of vague pain in the abdomen since 1 month with CT scan of abdomen and pelvis showing large lobulated solid cystic lesion of 11 x 10.6 x 9.7cm arising from left adnexa. Few internal septations were seen within the lesion. The lesion is extending into infra-umbilical region superior to urinary bladder which was suspected to be left ovarian

neoplasm. Routine hemogram and biochemical profile were within normal limits. CA125 was 5units/ml. Patient was prepared for surgery. Upon exploration uterus and ovaries were found to be normal, a 10 x 9 cm retroperitoneal mass was seen arising from the pelvis (Figure 1). It was clamped, cut and ligated and the mass was sent for histopathology report and it was consistent with pheochromocytoma showing trabecular arrangement of neoplastic cells with abundant cytoplasm (Figure 2). The post-operative period was uneventful.

Discussion

Pheochromocytomas are rare endocrine tumours that can present insidiously and remain undiagnosed until death. They may sometimes show onset of clear manifestations of catecholamines excess, most commonly as spells of headache, sweating, palpitation and hypertension. These tumours are no longer regarded as a uniform disease entity, but rather as a highly heterogeneous group of chromaffin cell neoplasm with different ages of onset, secretory profiles, locations and potential for malignancy ascending to the underlying genetic mutation.^[4]

Patients with pheochromocytoma may develop complicated and potentially lethal cardiovascular and

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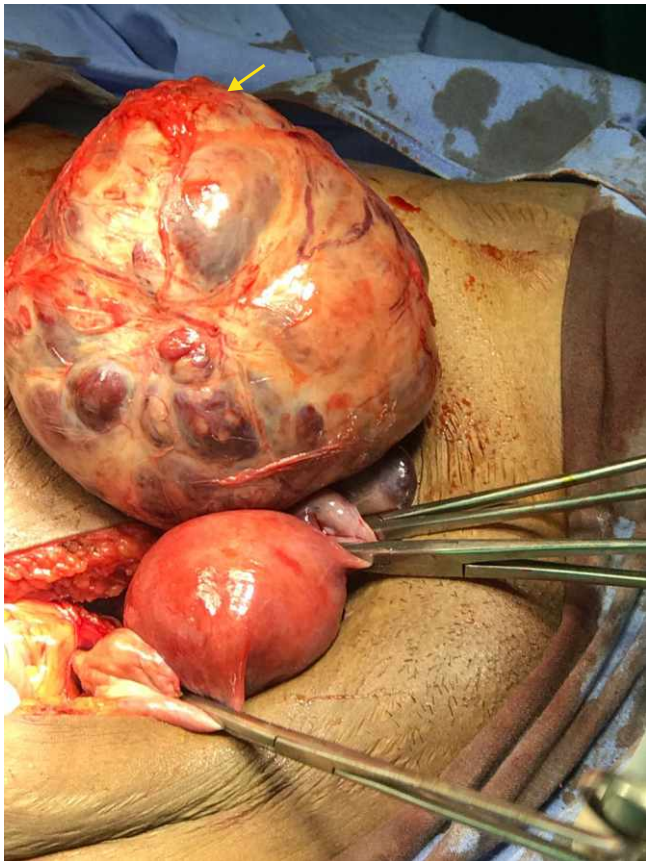


Figure 1: Gross features showing retroperitoneal mass (encircled) with normal uterus and ovaries.

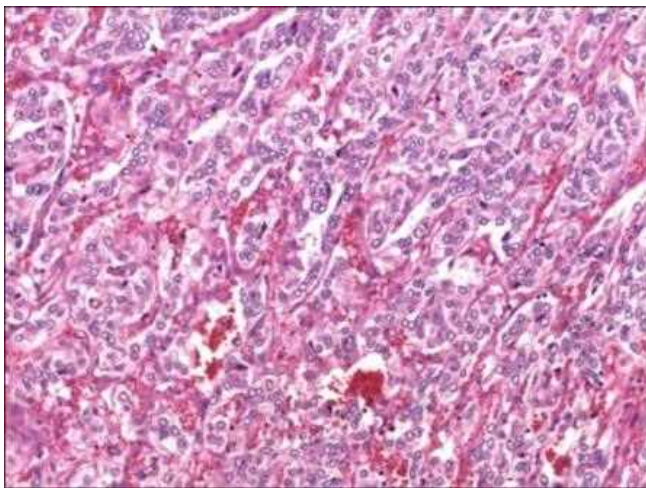


Figure 2: The H and E staining of the section shows neoplastic cells arranged in nests and trabecular pattern. cells have uniform round to oval nucleus with coarse chromatin and abundant eosinophilic cytoplasm.

other complications, especially during diagnostic or interventional procedures e.g., upon induction of anaesthesia or during surgery due to potent effect of

paroxysmal release of catecholamines.^[5] Pelvic paraganglioma may present primarily to the gynaecologist and may be treated initially as pre-eclampsia or other pelvic tumours.^[6] The patient might present with vague pelvic symptoms and may not be forthcoming with the classical symptoms.^[7] The gynaecologist might miss these important clues leading to a potentially dangerous unplanned exploration. Suboptimal imaging may reveal the mass to be related to uterus and adnexa and careful thin section imaging with 3D reconstruction might reveal the exact anatomy of the mass.

Plasma and 24 hrs urine meta-nephrines might be normal which further adds to the confusion. Sudden intra-operative hypertension upon tumour manipulation may be the first hint towards this pathology, which was not evident in our case. A detailed history, physical examination and a complete laboratory and cardiac evaluation are essential in preparation of the patient with pheochromocytoma for surgery. Normalization of blood volume minimises the possibility of protracted hypotension or shock resulting from sudden diffuse vasodilation at the time of tumour removal and if catecholamine induced hyperglycaemia is found pre-operatively, close monitoring must be initiated.^[5]

Surgery is the only curative treatment while pre-operative management has changed a little over the last 20 years, surgical procedures have evolved so that laparoscopic resection is the standard of care if feasible even for parauterine location.^[3,4] Radical surgical resection is the treatment of choice in giant extra-adrenal pheochromocytomas. Due to the possibility of neoplasm recurrence and metastasis especially after incomplete surgical excision, periodic check-ups and follow-up is a must for the patients.

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