

Extra-Adrenal Pheochromocytoma Presenting As Pre-Eclampsia: A Case Report

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Introduction:

Pheochromocytoma is a rare tumor of chromaffin cells most commonly arising from the adrenal medulla [1]. The peak incidence is in the third to fifth decades of life[1]. Bilateral disease is present in approximately 10% of patients. Bilateralism is much more common in familial pheochromocytoma, often found in association with the familial multiple endocrine neoplasia syndromes (MEN, types 2A and 2B). In patients with MEN type 2 syndromes, the risk of developing a contralateral tumor following unilateral adrenalectomy is approximately 50%. Other syndromes associated with pheochromocytoma include neurofibromatosis, von Hippel-Lindau disease, cerebellar hemangioblastoma, Sturge-Weber's syndrome, and tuberous sclerosis. In a series of 82 unselected patients with pheochromocytoma, 23% were found to be carriers of associated familial disorders. Therefore, all patients with pheochromocytoma should be screened for MEN2 and von Hippel-Lindau disease to avert further morbidity and mortality in the patients and their families. Extra-adrenal pheochromocytoma or functional paraganglioma occurs in approximately 10% to 15% of cases and may arise from any extra-adrenal chromaffin tissue in the body associated with sympathetic ganglia. Herein we report a case of extra-adrenal pheochromocytoma causing uncontrolled hypertension during pregnancy.

Case Report:

A 20-year-old woman came with a history of severe hypertension that was detected at 16 weeks of pregnancy. The blood pressure was not controlled even after aggressive antihypertensive therapy. Twenty-four hour Urine VMA (vanillyl mandelic acid) levels were high indicating the source of hypertension as pheochromocytoma. Pregnancy was terminated due to poor prognosis of the condition. Subsequent ultrasound was performed to identify the tumor. Ultrasound showed both adrenals to be normal however a mixed echogenicity mass with small hypoechoic areas measuring 5.4 X 4.2 cm was noted at the level of aortic bifurcation in the

retroperitoneum (Fig.1, Fig.2). A diagnosis of extra-adrenal pheochromocytoma was suggested. Contrast enhanced CT scan showed it to be an intensely enhancing retroperitoneal mass at the level of aortic bifurcation (Fig.3, Fig.4). Both the adrenals were normal confirming the diagnosis of Extra-adrenal Pheochromocytoma.



Fig1: Mixed Echogenicity Well Defined Retroperitoneal Mass At The Level Of Aortic Bifurcation (Transverse Section).

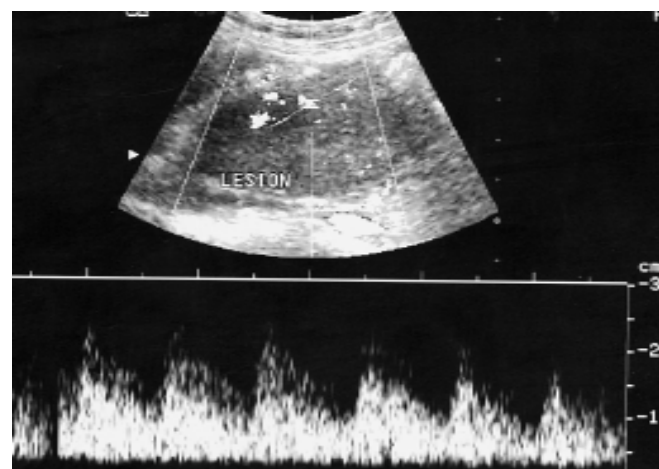


Fig2: Abundant Arterial Signals In The Mass.

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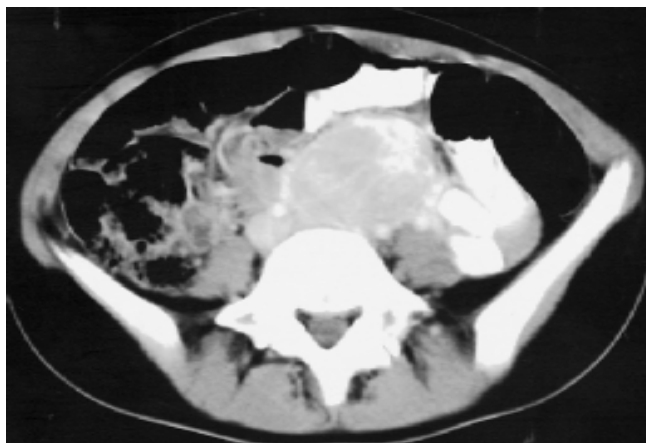
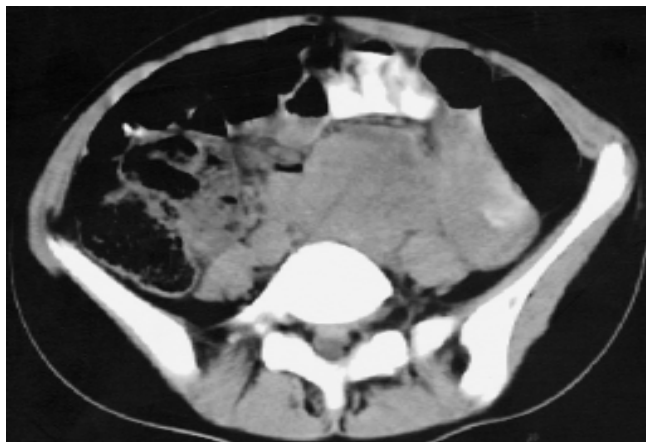


Fig3 And Fig4: Plain And Contrast CT sections show an intensely enhancing retroperitoneal mass at the level of aortic bifurcation.

Discussion:

Extra-adrenal pheochromocytoma is most often located within the abdomen and may have greater malignant potential than adrenal pheochromocytoma [2]. Extra-adrenal tumors usually have a poorer prognosis than adrenal tumors [2]. However, in one series of 73 patients referred to tertiary care centers, no difference was found in the metastatic potential or the prognosis of extra-adrenal tumors compared to adrenal tumors [5]. Due to the production and release of catecholamines, pheochromocytomas cause hypertension. However, only 0.1% to 0.5% of all hypertension patients will be found to have a pheochromocytoma. The importance of the recognition of this disease is that over 90% of patients properly diagnosed and treated are curable [3].

The hypertension caused by pheochromocytoma may be sustained or paroxysmal and is often severe with occasional malignant features of encephalopathy, retinopathy, and proteinuria. Less commonly, severe hypertensive reactions may occur during incidental surgery, following trauma, exercise, or micturition (in the setting of bladder pheochromocytoma) when the diagnosis is unsuspected. Other clinical features of pheochromocytoma include headache, sweating, palpitation, tachycardia, and severe anxiety along with epigastric or chest pain. Orthostatic hypotension is frequently present and is probably due to reduced intravascular volume following chronic adrenergic

stimulation.

The diagnosis of pheochromocytoma is established by the demonstration of elevated 24-hour urinary excretion of free catecholamines (norepinephrine and epinephrine) or catecholamine metabolites (vanillylmandelic acid and total metanephrines). The measurement of plasma catecholamines can also be of value in the diagnosis of pheochromocytoma. However, the measurement of plasma catecholamines has limited sensitivity and specificity. Plasma metanephrines have been reported to be more sensitive than plasma catecholamines [4].

Once the diagnosis is confirmed by biochemical determinations the localization and extent of disease should be determined [4]. Ninety-seven percent are found in the abdomen, 2% to 3% in the thorax, and 1% in the neck. The initial studies should be a chest film and abdominal computed tomographic (CT) scan. I131meta-iodobenzylguanidine (MIBG) has been found to be useful as a scintigraphic localization agent[1]. If the tumor is not adequately localized by these methods then magnetic resonance imaging (MRI), or rarely, vena cava catheterization with selective venous sampling for catecholamines may be indicated [4]. CT and MRI scans are about equally sensitive (98%-100%), while MIBG scanning has a sensitivity of only 80%. However, MIBG scanning has a specificity of 100%, compared to specificity of 70% for CT and MRI [3]. If extra-adrenal or metastatic disease is suspected, additional studies such as bone scan, liver-spleen scan, chest CT scan, or ultrasound may aid in determining the extent of disease. Surgical resection is the standard curative modality.

A paraganglioma or extra-adrenal pheochromocytoma is rarely observed during pregnancy with potentially lethal consequences [5,6,7]. Maternal prognosis depends on early diagnosis and multidisciplinary management prior to tumor resection [5]. Ultrasonography or MRI localizes the tumor. If MRI is not available, CT is often sufficient [5,7]. Its definitive treatment is surgical resection preceded by medical management [6].

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