ULTRASONIC AND COMPUTED TOMOGRAPHIC APPEARANCES OF PARAGANGLIOMA SIMULATING OVARIAN CANCER

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ABSTRACT

Paragangliomas or extra-adrenal pheochromocytomas are rare tumors affecting about 1/2000000 of the population[3]. They originate in paraganglia cells lying adjacent to the ganglia and plexuses of the autonomic nervous systems. Most of them arise from the sympathetic chain between the origin of the renal arteries and the aortic bifurcation. There is an association of paragangliomas with Von Hippel Lindau disease, neurofibromatosis and multiple endocrine neoplasia type IIa and IIb[2].

Paragangliomas usually present with large solid mass located just adjacent to the aorta in the middle age group. Sometimes, central necrosis is found. Mortality rate for surgery in the unsuspected cases is up to 50%. We report a case of an unusually large cystic paraganglioma in the left pelvis simulating an ovarian cancer. Misdiagnosis led to intra-operative hypertensive crisis and post-operative shock. These crises could have been avoided if we had searched for functional activity pre-operatively.

CASE REPORT

A 72-year-old female patient developed hypertension for 2 years. The physical examination revealed hard mass at left lower quadrant. The pelvic ultrasound revealed well-defined large cystic mass with septation about 10x12cm in the left pelvis. The cyst was homogeneous low echoic with solid papillary growth. Linear wall calcification was observed (Fig. 1). The findings were compatible with ovarian tumor.

Pelvic examination found a high positioned left pelvic mass with difficulty to be palpated. Bilateral adnexa were clear. The findings made a diagnosis of a questionable ovarian cancer so CT scan of the lower abdomen was performed. The scan revealed a well-defined, thin-walled cyst with solid papillary growth. The cyst was 10x13.5x18cm. Linear wall calcification was seen (Fig. 2). The Housfield unit intracystic part was 24-28 H.U. Following intravenous injection, intense enhancement of the solid part was observed (Fig. 3). The mass extended from the level of aortic bifurcation downward into the left pelvis. It was located adjacent to the aortic bifurcation posteriorly and the abdominal wall anteriorly. Neither evidence of adjacent organ invasion nor node enlargement was seen. Therefore theovarian tumor was supposed to be the most likely diagnosis. However, other possible diagnoses were leiomyosarcoma, malignant fibrous histiocytoma, mesenteric cyst and cystic node metastasis. The patient underwent surgical removal in the gynaecologic department. Intra-operative hypertensive crisis occured immediately after induction to anesthesia (BP 260/180mmHg) and persisted inspite of many vasodilator drugs were given. Operative finding was a large mesenteric root mass with high vascularity. The mass

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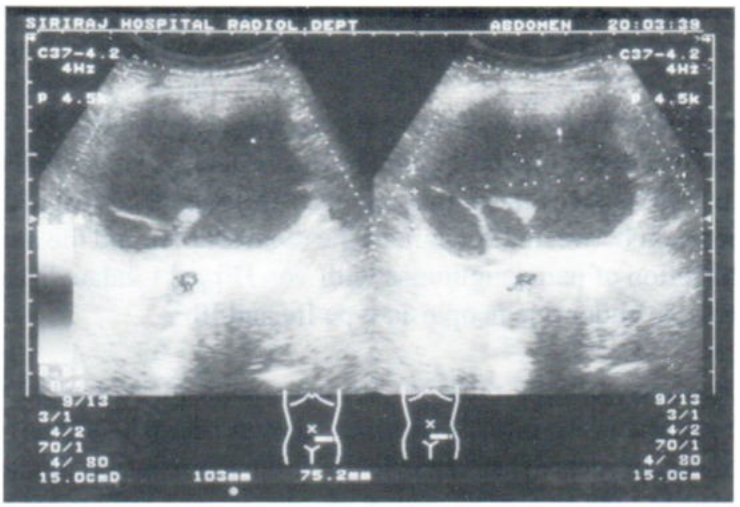
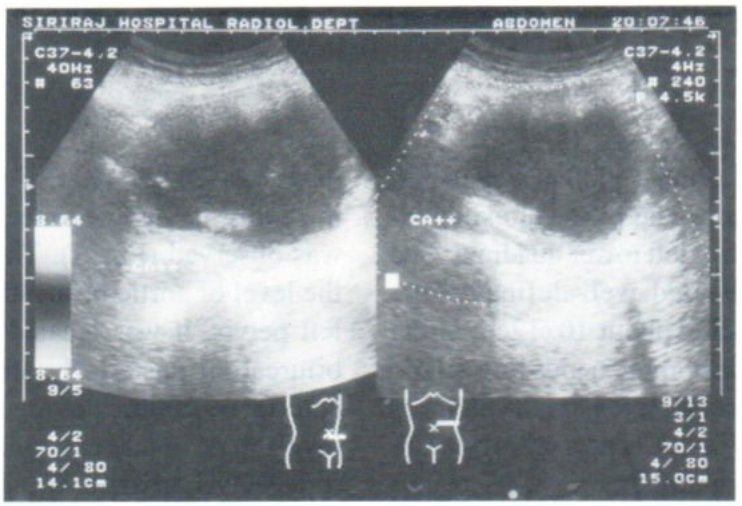
was totally removed and shown to be a large hemorrhagic cyst. The patient was shock immediately after the removal of the mass. After three hours of vaso-pressive drugs infusion, stable hemodynamic was achieved. Ten days later, the patient was discharged without complication. Pathological findings were chromaffin tumor cells with cytoplasmic vesicles containing cathecholamine compatible with paraganglioma.

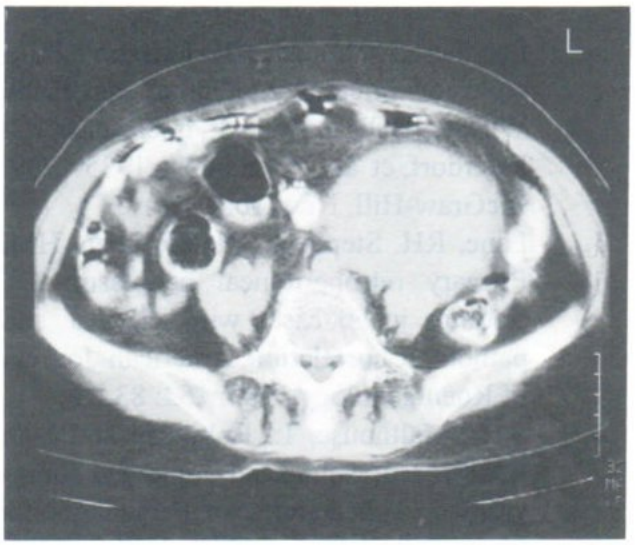


1A.

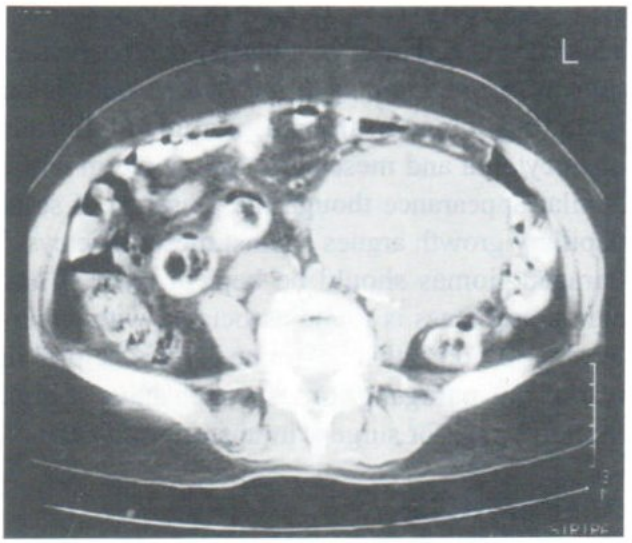


1B.

Fig.1. Ultrasound. (a) Transverse section through the left pelvis demonstrates large cystic mass with papillary growth. (b) Linear wall calcification is seen.



2A.



2B.

Fig.2. Nonenhanced CT scan. (a) Axial image through pelvis reveals well-defined, thin wall cyst 10.0x13.5x18.0cm.with solid papillary growth (b) Linear wall calcification is seen.

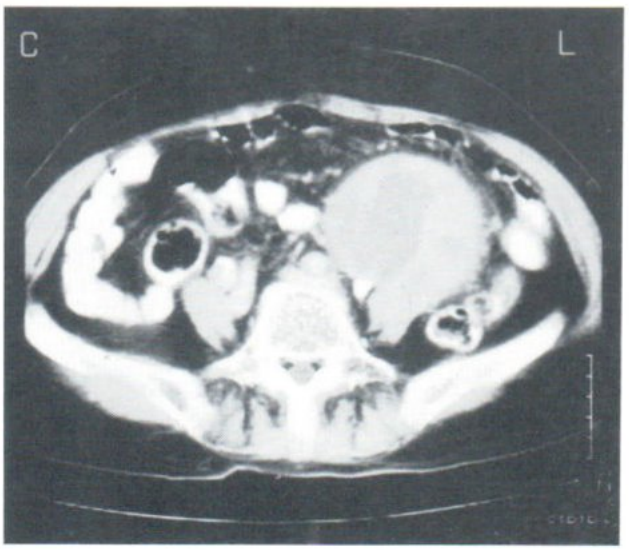


Fig.3. Contrast enhanced CT scan. Intense enhancement in solid part is noted.

DISCUSSION

Paraganglioma is a rare vascular tumor derived from neuroectodermal cells associated with ganglia cells of the autonomic nervous system. They usually lie in the para-aortic region from the level of renal arteries to the aortic bifurcation where the organ of Zuckerkaldle is found[1]. The majority of paragangliomas are functionally active[4]. Hayes et al found that 24 of 28 patients had hypertension[1]. However, cathecholamine levels were elevated in all 18 patients who were studied[1].

Paragangliomas are usually detected by CT scan as large, smooth, well-defined soft tissue mass in the immediate para-aortic region. Most small paragangliomas are homogeneous. However, larger lesions may undergo central necrosis and calcification. We report a case of an unusually large cystic paraganglioma with papillary growth simulating an ovarian cancer in an old woman. Misdiagnosis led to intra-operative hypertensive crisis, which has a mortality rate for surgery of nearly 50%[4]. Retrospective study found

a high positioned mass postulated a diagnosis other than ovarian cancer. Many other neoplasm including leiomyosarcoma, malignant fibrous histiocytoma and mesenteric cyst could give the similar appearance though the presence of solid papillary growth argues against mesenteric cyst.[3] Paragangliomas should be kept in mind if any para-aortic mass is found associated with hypertension. Blood cathecholamine level is helpful in ruling out paragangliomas which have a high mortality rate for surgery in unsuspected cases.

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