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Metachronous Pheochromocytoma in urinary bladder and adrenal

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Abstract: Pheochromocytomas and paragangliomas are chromaffin cell tumours that embryonically derive from the neural crest and functionally they produce store and secrete catecholamines. While pheochromocytoma arises from the adrenal medulla, paraganglioma develop from both sympathetic and parasympathetic paraganglia. Synchronous or matachronous presentation of paraganglioma and pheochromocytoma are very rare. Here we report a case of 32-years - old male with multifocal functional paraganglioma of urinary bladder followed by pheochromocytoma of right adrenal seventeen years later which were surgically removed with successful outcome.
Keywords: pheochromocytoma, paraganglioma, chromaffin cell.

INTRODUCTION

Paraganglia are the cells of neuroendocrine origin which lie near or in the autonomic nervous system and extend anywhere from the skull base to the pelvic floor where the sympathetic or parasympathetic ganglia exists [1]. Paraganglioma can arise synchronously or metachronously and usually less functionally active than pheochromocytoma [1]. Though the majority of paraganglioma are sporadic, a genetic association has been identified <50% of cases [2]. Common sites of extra adrenal paragangliomas are the paraaortic region at the level of renal hila, at the organ of Zuckerkandl , thoracic paraspinal region, bladder and head and neck [1] but rarely within kidney, bladder, urethra, prostate, spermatic cord, testis, genital tract, and liver. Bladder paraganglioma constitutes less than 0.06% of all bladder tumors and less than 1% of pheochromocytoma [3]. Isolated functional bladder paraganglioma are reported many times but metachronous adrenal pheochromocytoma in a patient who had diagnosed as a functional bladder paraganglioma 17 years back is not reported so far previously to the best of our knowledge.

CASE REPORT

A 32-years-old man incidentally detected as right adrenal mass on noncontrast CT scan (NCCT) of abdomen while evaluating for left ureteric calculus. He was on antihypertensive for hypertension diagnosed three years back. He had a past medical history of bladder pheochromocytoma diagnosed by clinical features of severe headache with sweating and palpitation at the end of micturation seventeen years back. He had undergone partial cystectomy at that time and was lost for follow up. Again he became hypertensive with features of sweating and palpitation six years back for which he was worked up by ultrasonography, CT scan of bladder and pelvis, and urinary catecholamine levels .Imaging studies with ultrasonography (UDG) was normal but urinary catecholamine level was raised. Patient was advised to evaluate for pheochromocytoma and MIBG scanning but he denied and continued his antihypertensive (prazosin, amlodepine and atenolol). He presented with left ureteric colic and on evaluation found to be a case of left ureteric stone with and shrunken right kidney. Stone was expelled spontaneously within a few days which were confirmed by noncontrast CT scan, incidentally showing right suprarenal mass of a size 7cmX5cmX3cm and weighing 94.4 gms along with small contracted right kidney (Fig.-1).

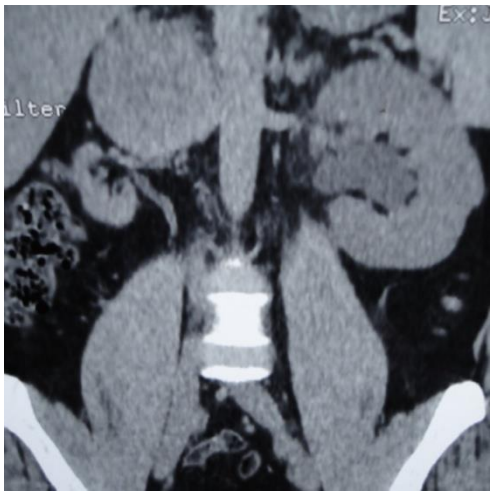

Fig-1: Noncontrast CT scans showing right small contracted kidney with right suprarenal mass

Physical examination was unremarkable. He was normoglycemic with normal renal biochemical parameters. A healthy scar of pfannestiel incision for the previous operation was present. Biochemical examinations showing raised urinary metanephrines-249.4 micro GM/gm of creatinine ;(Normal level- 27-155microG/gm of creatinine), normetanephrines level >5000microGm/gm of keratinize Urinary Vanilemandelic acid (VMA) - 6.17microG/gm of creatinine (normal range- 1.6-4.2 microG/g of creatinne), with normal thyroid profile, cortisol level, calcitonin and serum calcium levels. DTPA renogram suggested 3% function of right kidney. Laperoscopic right nephrectomy with right adrenelectomy (Fig.-2) was performed with uneventful recovery.

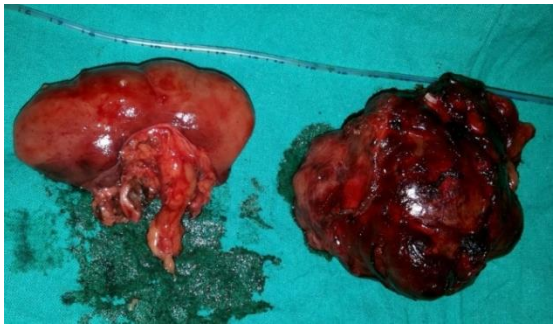

Fig.-2: The resected specimen of right kidney with right adrenal gland.

Histopathology showed the adrenal composed of large polygonal cells with centrally placed round or oval nuclei with abundant eosinophilic cytoplasm arranged in nests or 'Zellballen' around an elobarate vascular network(Fig.-3A & 3B). Cells with bizzere hyperchromatic nuclei with mitotic figures were seen. Focal areas of capsular invasion was present. Till one year of follow up the patient is symptomless without any antihypertensive drugs.

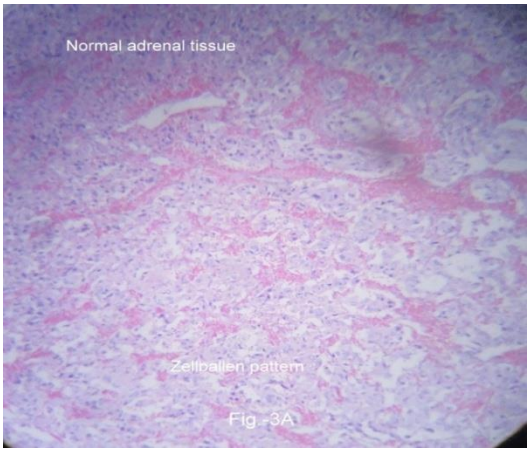

Fig-3: AHistopathology showing normal adrenal tissue in the upper part and picture of pheochromocytoma in the lower part. (H&E X 400)

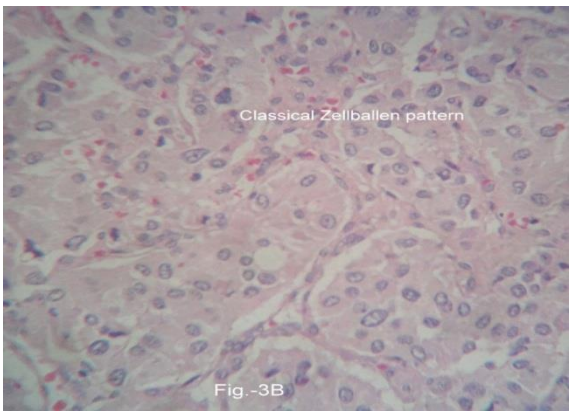

Fig-3; Histopathological picture showing classical Zellballen pattern (H&E X 400)

DISCUSSION

Paraganglioma are extra-adrenal chromaffin tumors that develop from the neuroectodermal cells of the autonomous nervous system that constitutes about 10% of pheochromocytomas [4]. Bladder paraganglioma constitutes 0.06% of all bladder tumors and 6% of all extra adrenal paragangliomas [3,4]. and most of them are high grade and functional in comparison to retroperitoneal paragangliomas [5]. Paragangliomas are seen in adults in age group of 30-50 years and are mostly benign. The patient presented with signs and symptoms related to catecholamine secretion and included paroxysmal hypertension with anxiety, sweating, chest pain, headache, and facial pallor or flushing during the attack. USG is the first line of investigative modality to detect the silent as well as functioning ectopic lesions [6]. Biphasic CECT abdomen is the modality of choice to diagnose these tumors as the entire sympathetic chain can be evaluated. Small tumors show arterial enhancement and appear homogeneous as compared to larger tumors which show heterogeneous enhancement as was observed in our case. Areas of calcification or hemorrhage can also be seen. On magnetic resonance imaging (MRI), these tumors are iso to hypointense as compared to liver parenchyma and markedly hyper intense on T2 weighted images. Although CT and MRI having excellent sensitivity for detecting most catecholamine releasing tumors but functional imaging I231 labeled metaiodobenzylguanidine (MIBG) scintigraphy PET Octreoscan highly effective for diagnosis and has a sensitivity of 100% in the diagnosis of extra adrenal pheochromocytoma [6]. These functional studies helps in determination of the extent of the disease, presence of multiple tumors or metastasis which are important for management [7]. The only definite sign of malignancy is distant metastasis to organs like bone, lung, liver and lymph nodes [2]. Histopathologically these tumor contain neuroendocrine cells surrounded by sustenacular cells in a classical 'Zelballen' pattern. Features that predict malignancy include extra-adrenal location, confluent tumor necrosis, vascular invasion,

local invasion, coarse nodularity and absence of hyaline globules [8].

Surgery is the mainstay of the treatment. Either an open or laparoscopic approach can be performed depending on the experience of the surgeon. In case of suspected malignancy an open approach is recommended to protect the capsule of the tumor and to avoid spillage to avoid recurrences [2] but the laparoscopic surgery are advantageous to reduce post operative pain, an early recovery and a shorter hospital stay in comparison to open approach. Radiation therapy has been advocated for tumors with unresectable tumors [7].

CONCLUSION

The presented case is rare in presentation having metachronous origin of paraganglioma in urinary bladder with a late presentation of pheochrocytoma in adrenal after seventeen years. Our case emphasizes thorough work up to pick up synchronous or metachronous lesions in other organs. Life-long follow-up is necessary to detect late recurrences. We suggested that there should be at least an annual follow-up for these patients if they are asymptomatic or whenever clinically indicated.

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