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Presentation and Management of Bulky Adrenal Tumors in Khartoum

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Research Article

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Abstract

Introduction: The detection of adrenal masses, especially pheochromocytoma needs a high index of suspension. All patients with symptoms or signs suggestive of pheochromocytoma need thorough investigations. Patients with pheochromocytoma have a potentially curable cause for hypertension, but if undetected, pheochromocytoma presents a high risk of morbidity and mortality.

Objectives to Assess:

The modes of presentation of adrenal masses.

The available methods of investigations.

The options of surgical treatment.

Results: Seventy-two patients were treated in the urology unit of Soba University hospital in Khartoum in the period from 1996 to 2017. The mean age was 32 years. The male to female ratio was 1 to 2.7. CT abdomen was diagnostic in 93.1 percent. The commonest tumor was pheochromocytoma occurring in 75%, followed by adrenal carcinoma in 11.2% then benign tumors in 6.8% and others in 7.0%. All patients underwent open surgical adrenalectomy. The mortality rate was 1.4%. Other complications are discussed in the text.

Discussion: CT abdomen has a sensitivity of 100% in diagnosing adrenal masses. This sensitivity reduces to about 93% in detecting extra adrenal tumors. Although open surgical adrenalectomy remains a very effective method of treatment of bulky pheochromocytoma, laparoscopic adrenalectomy is becoming a good alternative to open surgery.

Keywords: Pheochromocytoma; Adrenalectomy; Laparoscopy; Adrenal Carcinoma; Catecholamine's

Abbreviations: VMA: Vinyl Mandelic Acid; CT: Computed Tomography.

Introduction

The two main concerns with regard to adrenal masses are:

- Whether it is hormonally active (functional).
- Whether it is malignant.

Benign adrenal cortical adenomas are commonly smaller than 6 cm in diameter on initial presentation, but they can be larger [1]. The treatment for hormonally active adrenal tumors is surgery [2]. In this study we present a series of patients with bulky adrenal tumors (tumors 9 cm or larger in diameter) with special emphasis on pheochromocytoma. All patients with any of the following symptoms or signs were investigates for pheochromocytoma:

- Patients requiring more than four medications to control their blood pressure.
- Patients with onset of hypertension before the age of 35 or above the age of 60 years.
- Patients with other symptoms or signs suggestive of pheochromcytoma.

Objectives to Assess

- Frequency of bulky adrenal tumors in Khartoum.
- Modes of presentation of bulky adrenal tumors.
- Available methods of investigations.
- Methods of surgical treatment.

Patients and Methods

This is a hospital based descriptive study carried out in the urology unit of Soba University hospital in Khartoum –Sudan- in the period from Jan 1996 to Dec 2017. All patients presenting with symptoms or signs of pheochromcytoma or discovered to have adrenal tumors incidentally were included in the study. All had detailed history taken, physical examination with special emphasis of frequent blood pressure recording at different times of the day. Investigations included urinalysis, Vinyl Mandelic Acid (VMA) measurements, renal function tests, chest X-ray, ECG, abdominal Ultrasonography and computed tomography of the abdomen. Selective angiography and MRI were added in few patients with suspected extra adrenal tumors.

All the 72 patients in this study had surgical treatment in the same urology unit at Soba hospital by one group of surgeons. There was close collaboration between the physician endocrinologist, the anethiologist and the urologist. All the patients with pheochromocytoma were started on an alpha blocker (Pheoxybenzamine tablets) two weeks prior to surgery. Beta blockers were added in few patients with severe tachycardia .Blood was prepared for all patients. All the patients with pheochromocytoma had invasive arterial line monitoring with a transducer. Central venous line was established following induction of anesthesia. A short acting alpha blocker was kept ready to compact with possible fluctuations of blood pressure during induction of anesthesia, positioning of patient, and during manipulation of the tumor. Blood pressure and other vital signs were monitored continuously during and in the post-operative period. The surgical approach for adrenalectomy was a posterior approach through the elevens intercostals space retero-peritoneally with the patient in the kidney position. All the surgically removed adrenal tumors were sent for histological examination. The follow up was for one year with review of recurrent symptoms or signs.

Results

A total of 72 patients were included in this study. Their ages ranged between 12 and 52 with a mean of 32 years. There were 52 females and 20 males with a male to female ratio of 1 to 2.7. The commonest presenting symptom was hypertension occurring in 42 patients (58.3%), followed by headache in 16 patents (22.2%), blurring of vision in 8 (11.1%), loin pain in 4 (5.6%), while it was incidental finding during abdominal CT for other reasons in 2 patients (2.8%). Twelve patients (16.7%) gave past history of diabetes and 4 (5.6%)were asthmatic. Clinical examination prior to surgery revealed palpable adrenal mass in two patients (2.8%) with malignant adrenal tumors. Three female patients (4.2%) had simple multinodular goitre. Severe papilledema was detected in 4(5.6%) young patients.

Computed tomography (CT) of the abdomen was diagnostic in 67patients (93.1%) including the incidentally detected cases. The Ct abdomen could locate the anatomical site, the size as well as commenting on the kidneys and other abdominal organs especially in cases of malignant adrenal tumors. Abdominal Ultrasonography was performed in 52 patients (72.0%), VMA in 6 patients (8.3%), MRI in 4 (5.6%) with recurrent tumors, while selective angiography was performed in one patient (1.4%) with difficult to locate the anatomic site of a recurrent tumor. Chest X-ray showed cariomegaly in 4 patients (5.6%). Fifty-two (72.2%) of the tumors were on the right side in close proximity to the upper pole of the right kidney, 16 (22.2%) were on the left side while 4 (5.6%) were bilateral.

All the 72 patients (100%) underwent open surgical adrenalectomy as the sole method of treatment. The surgical approach was posterior retroperitoneal through the 11th intercostals space in 68 patients (94.4%), while it was transperitoneal in 4 patients (5.6%) with bilateral tumors. There were 8 post-operative complications (11.1%). The mortality was one (1.4%) in a patient with bulky adrenal carcinoma invading the upper pole of the left kidney. other complications included recurrence within 6 months in 5 patients (6.9%), spleenectomy in 0ne (1.4%), and severe post-operative hypotension in 4 patients (5.6%) who received beta blocker as well as a short acting alpha blocker intra operatively. The blood pressure was back to normal in the immediate post-operative period in 8 patients (11.1%), while it was back to normal in all the other patients within seven to ten post-operative days .Four patients (5.6%) developed severe hypotension in the immediate postoperative period but could be resumed with adequate intravenous normal saline. Two of the four patients with papilledema resumed their vision within two weeks while it resolved gradually in the other two patients within six weeks. The VMA was back to normal within ten to fifteen days postoperatively. The histology of the tumors showed pheochromocytoma in 54 patients (75.0%), adrenal carcinoma in 8 (11.1%), benign tumors in 5 (6.9%), adrenal hyperplasia in 3 (4.2%) and tuberculosis in 2 patients (2.8%).

Discussion

Most of the patients in this study were of young age groups with a mean of 32 years. On the contrary Eisenhower, et al. reported a mean age of 50 years in patients with pheochromocytoma, while in the group with non-secreting tumors, the mean age was 42 years at the time of diagnosis. In their study which included 172 patients, pheochromocytoma tumors were diagnosed eleven years later than those with non-epinephrine producing tumors. Neumannet, et al. [3] and Manelli, et al. [4] showed that age at diagnosis varies according to tumor catecholamine phenotype. They showed that earlier age at diagnosis is hereditary than sporadic. In the current study we couldn't document hereditary link with age at presentation. There were more females than males in this study making a male to female ratio of 1 to 2.7. This is in agreement with reports by Grunbach, et al. [5] and Kloos, et al. [6]. One of the possible explanations of females being affected more than males could be attributed to the fact that females have more opportunities to receive abdominal CT compared to males. In this study most of the adrenal tumors occurred on the right side. Jonson, et al. [7] reported that the left side was affected more the right side. In the current study bilateral tumors constituted only 5.6%, while in other reports approximately 10% of the adrenal tumors were bilateral [8,9].

In this study CT abdomen was diagnostic in 93.1%. In other studies [10,11] the sensitivity in detecting pheochromocytoma ranged between 93 to 100%. They showed that sensitivity of CT decreases to approximately 90% for extra-adrenal pheochromocytoma [12]. Other studies showed MRI to be superior to CT for detecting extra- adrenal tumors [13,14]. The majority of the bulky tumors (75.0%) in this study were pheochromocytoma; however pheochromocytoma was reported to occur in only 8% in other series [15]. Some studies showed chances of having a malignant tumor correlates directly with the size of the tumor [16]. This can be explained at least in part by the fact that most of the patients in this study presented late giving more chances for the tumor to grow bigger. Incidental finding of adrenal tumor in this study was 2.8%. Other studies showed incidence of up to 4.0% in patients undergoing CT abdomen for other reasons [17]. All the patients in this study underwent open surgical adrenalectomy as the sole method of treatment. Surgery for pheochromocytoma entails several considerations. Induction of anesthesia before surgery, positioning of the patient and manipulation of the tumor or other stimulation can cause massive outpouring of catecholamines from the tumor resulting in hypertensive crisis, stroke or myocardial infarction.

To prevent these symptoms, patients with pheochromocytoma must undergo pharmacological blockade of catecholamine synthesis with phenoxybenzamine (an alpha adenceptor blocker) [18-21]. The operative approach in the majority of the patients in this study was posterior retroperitoneal through the 11th intercostals space, while open surgery was performed through a transperitoneal approach in only 4 patients with bilateral tumors. The survival rates in this study were 98.6%. In other studies the survival rates following open transperitoneal approach ranged between 97.7 to 100% [22-24]. Development of laparoscopic surgical techniques has provided an alternative to open surgical procedures [25-27]. It was shown by some authors that both open surgery and laparoscopic surgery have similar blood loss and complications [28,29]. Others reported that laparoscopic adrenalectomy is the preferred procedure for lesions smaller than 8cm [30,31]. No randomized trials have compared open surgery versus laparoscopic adrenalectomy, however open surgery is performed for bilateral tumors, large masses (masses more than 10cm, for possible malignant disease and

pheochromocytoma [32,33]. As this study involved mainly bulky tumors (more than 9cm in diameter) most of which were pheochromcytoma, all were treated with open surgery, besides that laparoscopic surgery was not well established in this unit at the time of starting this study. We look forward to see the role of laparoscopy in bulky pheochromocytoma.

Conclusion

Diagnosis of adrenal tumors, especially the pheochromocytoma entails a high index of suspension in all patients with hypertension, or other symptoms or signs suggestive of pheochromocytoma. Their management entails a close collaboration between the endocrinologist, the cardiologist, the anethiologist and the urologist. With advances in imaging, incidentaloma is expected to be detected more. With advances in laparoscopic techniques the future can witness treating bulky adrenal pheochromocytoma with laparoscopy.

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