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Laparoscopic Partial Adrenalectomy for Bilateral Pheochromocytomas in a Boy with von Hippel-Lindau Disease

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Key Words

Pheochromocytoma · Von Hippel-Lindau · Laparoscopy · Children

Abstract

Objectives: In adults, increasing numbers of adrenalectomies for pheochromocytomas are performed laparoscopically. We report for the first time laparoscopic bilateral subtotal adrenalectomy for pheochromocytomas in an 8-year-old boy with von Hippel-Lindau disease.

Methods: In July 1998, an 8-year-old boy with von Hippel-Lindau disease underwent laparoscopic adrenal-sparing surgery for bilateral pheochromocytomas. The boy presented with severe hypertension and two pheochromocytomas on both sides.

Results: The child could be solely treated with laparoscopic adrenal-sparing surgery. The procedure was completed as planned. There were absolutely no intraoperative or postoperative complications. Postoperatively, catecholamine levels and hypertension went back to normal. At follow-up no residual tumor could be detected and no steroid replacement therapy was necessary.

Conclusions: In experienced hands, laparoscopic adrenal-sparing surgery for pheochromocytomas is feasible and safe. Moreover, this minimal invasive approach represents an exceptional improvement in life quality, especially in children with von Hippel-Lindau disease since surgery will probably be necessary again and again in their future life.

Introduction

Von Hippel-Lindau disease is an autosomal dominant disorder characterized by the development of hemangioblastomas in the cerebellum, spinal cord, and retina, renal cell carcinoma and cysts, pancreatic cysts, and pheochromocytoma [1–3]. Prevalence of this condition is approximately 1/40,000, and age manifestation is usually 18–30 years. The incidence of pheochromocytoma varies in large series from 10 to 19% [2, 4]. So far all the affected patients underwent multiple open surgery for recurring tumors. We were the first group to report on laparoscopic adrenal-sparing surgery in adults [5]. Many studies in adults and children have already shown that the morbidity of laparoscopy is much lower than that of open surgery [6, 7]. Nowadays adrenalectomies are increasingly performed laparoscopically especially for benign cortical adenomas of the adrenal gland [8]. Pheochromocytomas differ from adrenal adenoma. The risk of hypertensive crisis during surgery cannot be neglected although it can be reduced by preoperative treatment using a-blockers as well as β-blockers and early intraoperative venous ligation. Bilateral pheochromocytomas occur in inherited syndromes such as von Hippel-

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Lindau disease [9]. Yet there have been only a few reports of bilateral laparoscopic adrenalectomy. As an alternative, adrenal-sparing surgery should be considered, but laparoscopic partial resection has only been described by our group so far [5] and was only performed in adults. We report our experience and management in a particular case of bilateral pheochromocytoma of an 8-year-old boy with von Hippel-Lindau disease. Our results show that minimal invasive removal of bilateral pheochromocytomas without total adrenalectomy is feasible and safe in children. The residual adrenal tissue and no need for steroid replacement therapy are important since side effects of medication can be avoided and long-term quality of life remains untouched.

Patient and Methods

Patient

In July 1998 an 8-year-old boy with autosomal dominant von Hippel-Lindau disease was transferred to our department for laparoscopic adrenal-sparing surgery for bilateral pheochromocytomas (fig. 1). The boy presented with severe hypertension and two pheochromocytomas on both sides. Investigations of the VHL gene revealed a G to A exchange at nucleotide 490.

Preoperative Treatment

Five weeks prior to the planned operation we started with a certain regimen to avoid severe hypertensive complications. He received phenoxybenzamine alone for the first 4 weeks and during the last 7 days additional propanolol.

Surgical Technique

Using the laparoscopic methodology we prefer the transperitoneal route. The camera port is placed at the umbilicus. Four 10-mm trocars are needed for one side, but three tocars can be used for both sides (umbilicus, subcostal pararectal right and left side), so that five trocars suffice altogether. On the left side the colonic flexure and spleen are dissected and displaced medially. On the right side it is sufficient to incise the peritoneum along the vena cava and parallel to the liver (fig. 2). This approach has already been described previously in detail by our group [8]. Since cold is a stress factor which may indeed stimulate the sympathetic nervous system and thus induce cardiovascular changes, the carbon dioxide insufflated was prewarmed with a heat exchanger integrated in the insufflation tubing [10]. The pheochromocytomas are removed with a thin layer of uninvolved tissue. The adrenal vein and most of the connective tissue around the remaining adrenal gland are left intact to preserve the vascular supply. Hemostasis is achieved by a forceps for bipolar coagulation which is used for dissection as well, and the harmonic scalpel. Finally the cut surface is sealed with fibrin glue, which is a routine preventive measure to preclude delayed bleeding [5].

Results

The case could be solely treated with laparoscopic adrenal-sparing surgery. The procedure was completed as planned and there was no need for conversion to open surgery. Operating time for the right side tumor was 90 and 160 min for the left side, respectively. Forty-five minutes were needed to change the position on the table after the right side was finished. There were absolutely no intraoperative or postoperative complications. No blood transfusion was necessary. Postoperatively, blood pressure readings returned to normal values within the first day after surgery without any antihypertensive medication. Two months postoperatively, endocrinological reevaluation revealed normal adrenal gland function. Urine catecholamine levels went back to normal as well. A follow-up CT scan showed absolutely no residual tumor (fig. 3). So far there is no need for any steroid replacement therapy. The child went back to normal activities immediately after discharge on the fifth postoperative day. Histological examination of the tumor specimens confirmed pheochromocytomas of both adrenal glands.

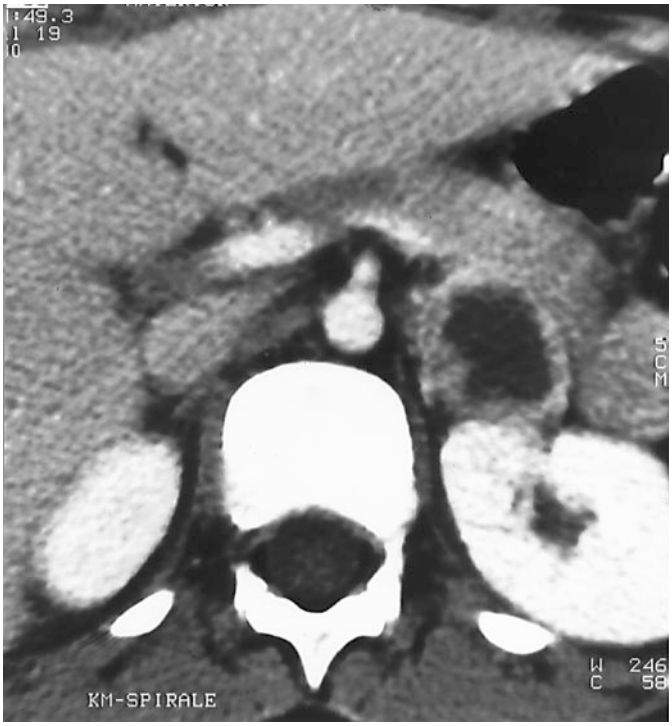

Fig. 1. Preoperative CT scan showing the extent of the pheochromocytoma on the left side.

Discussion

The clinical management of pheochromocytomas in patients with autosomal dominant von Hippel-Lindau disease is still controversial. Bilateral total adrenalectomy has been suggested to prevent risk fo recurrence and is still commonly regarded to be adequate. However, the patients and especially children then require lifelong steroid medication, which is a crucial problem during growth and puberty. An alternative is to leave the apparently unaffected adrenal gland in place. To our knowledge we were the first to report on laparoscopic adrenal gland-sparing surgery for pheochromocytoma in adult patients with von Hippel-Lindau disease [5]. Irvin et al. [11] reported in a follow-up of 7 years in 1983 that preservation of grossly normal adrenal tissue after excision of bilateral tumors may be adequate treatment. Such

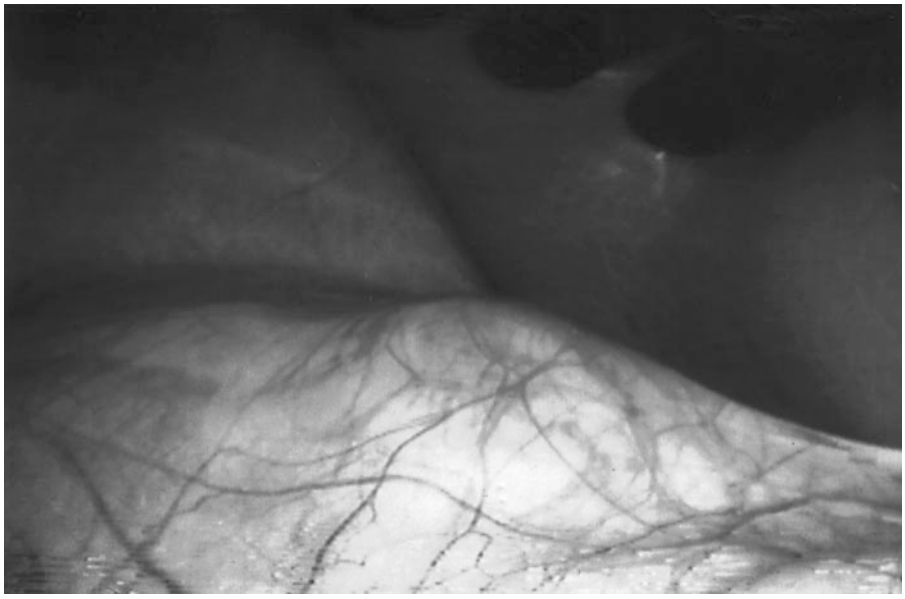

Fig. 2. Intraoperative laparoscopic appearance of the right tumor underneath the peritoneal sheath.

an approach balances the risk of later reoperation for recurrence of pheochromocytoma in the remaining adrenal tissue against the risks of chronic adrenal insufficiency in patients requiring lifelong steroid substitution therapy. This must be of special interest in children. We have to consider that removing both adrenals imposes a new burden on affected patients, as hormonal replacement can only partially substitute for the physiological circadian rhythm of hormone release. There is also the risk of an addisonian crisis in children with poor compliance and probably of osteoporosis with long-term treatment. However, if one attempts to spare the cortex there is a risk of recurrence because some medullary tissue will remain. These hazards must be carefully weighed against the benefits. The risk of recurrence is difficult to asses, since in von Hippel-Lindau disease it is different for each specific mutation and therefore, it may be kindred-specific. However, the risk is low, with 1 recurrence in a series of 29 patients with pheochromocytomas who had a mean follow-up of 7 years, at least [12]. In view of all these arguments, an increasing number of authors are now in favor of cortex-sparing surgery [11, 13–18]. Usually mean age of diagnosis of pheochromocytoma is the second and the third decades [2, 4]. Therefore our 8-year-old patient with bilateral pheochromocytomas as second manifestation of his von Hippel-Lindau disease represents a rare case, encouraging us to perform minimal invasive adrenal gland-sparing surgery. Stoba et al. [19] reported that in childhood, particularly in bilateral tumors, enucleation is the method of choice to preserve hormonal function of adrenal tissue. A crucial factor in that sort of surgery is the preservation of the vascular supply to the cortex. In a few exceptional cases, cortical tissue left in the adrenal bed may survive, although the arterial and venous vessels have been ligated. There are occasional reports demonstrating normal functioning adrenal cortex after such a procedure [15]. Subtotal adrenal resection with preservation of the adrenal vein has been proposed by others [16–18]. However, subtotal adrenalectomy does not remove all medullary tissue so that there is a 2-fold risk of tumor recurrence and postoperative adrenal insufficiency [13]. Therefore, we removed the tumors with a small margin of healthy tissue, while

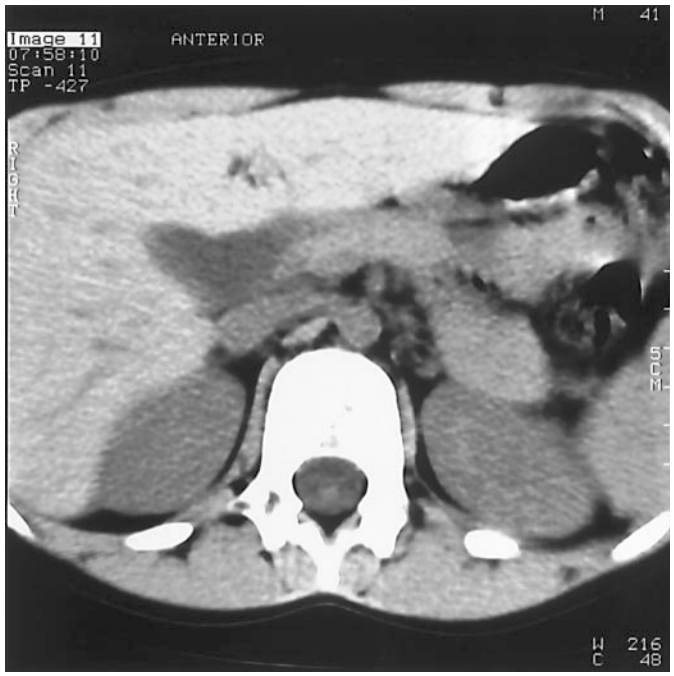

Fig. 3. Postoperative CT scan revealing no evidence of any residual tumor or local recurrence.

preserving most of the uninvolved adrenal tissue, supplying arterial branches and adrenal vein.

Laparoscopic excision of pheochromocytoma is technically demanding, since these tumors arise in the center of the gland [20]. This method has the advantage of optical magnification and, thus, provides better visualization of small vessels, which in turn allows for meticu-

lous dissection during partial resection. Of course, hemorrhage is a concern, but as in adults we were able to achive adequate hemostasis by bipolar coagulation and the harmonic scalpel in the child as well. Moreover, as laparoscopy is less traumatic than open surgery, adhesion formation is reduced so that further surgery for potential recurrences is less difficult [21]. Thus, it may be assumed that for most postlaparoscopy patients the overall morbidity of a second operation for a recurrence is likely to be a minor problem compared to the side effects of lifelong steroid replacement therapy. Of course in all the affected patients lifelong follow-up is mandatory because tumor may recur many years later. When considering hospitalization alone, the advantages of laparoscopy over open surgery may seem insignificant but if overall postoperative morbidity and time to complete recovery are also taken into account, the benefits are substantial. Moreover, this minimal invasive approach, especially in children with von Hippel-Lindau disease where surgery will probably be necessary even several times in their future life, represents an exceptional improvement in life quality, and may also render further surgery for recurrent adrenal disease less difficult.

In conclusion, our experience with laparoscopic adrenal-sparing surgery for familial pheochromocytoma in adults tempted us to perform that procedure in an 8-yearold boy with bilateral pheochromocytoma as manifestation of the von Hippel-Lindau disease. We demonstrate that this particular approach is feasible and safe. Following adequate preoperative medication using ·-blockers, good hemodynamic stability could be maintained during surgery. We show that the specific risks of surgery for pheochromocytoma are not enhanced by the laparoscopic approach [22, 23]. No major intraoperative and absolutely no postoperative complications occurred, and overall morbidity was very low in our 8-year-old boy.

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Editorial Comment

B. Guillonneau, Athens

This article is very representative of the current trends in urology, for several reasons. Firstly, in terms of the facts, it shows the possibility of conservative surgery for bilateral pheochromocytomas in a child, which is quite remarkable. This is a rare lesion and this case is already interesting per se. Secondly, the technique used illustrates the progress in laparoscopy, as the ability to perform a rare, difficult and complex procedure by laparoscopic surgery reflects the high level of technical skills acquired by the operators. This means that laparoscopic surgery can now be used not only to perform routine urological surgery, but also surgery of exceptional cases, demonstrating the maturity it has acquired over recent years in the hands of experienced operators. Laparoscopy is no longer an 'extreme' form of surgery, but a surgical modality which has become part of routine practice and which can be used to treat these extreme cases.

The urological community must be made aware of the enormous progress in the technical skill of laparoscopic operators, who are now able to perform the very great majority of urological surgical procedures. The frontiers of laparoscopic urological surgery are being progressively extended day by day.