Retroperitoneal Giant Schwannoma: Difficulties in Diagnosis and Subsequent Surgical Management

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Introduction:
The term retroperitoneal tumour is usually confined to lesions arising from tissues-muscles, fat, fibrous tissue, lymph nodes, nerves (24%) and developmental remnants (75% mesodermal) of this compartment but excluding origin from retroperitoneal organs (pancreas, kidneys, ureters and adrenal glands)1. The incidence of primary retroperitoneal tumours is 0.3 to 0.3%2. Retroperitoneal swellings may be cystic or solid, benign or malignant1. Schwannomas are rare tumour arising from schwann cells of peripheral nerve sheath3. Women (54%) are more affected than men (46%)4. Most of the retroperitoneal schwannomas are benign4, locally aggressive and rarely metastasize2. It rarely form cysts, cystic changes are noted in 63% of benign and 73% malignant schwannomas resulting from alterations in vascular wall1. The main symptoms are abdominal pain (85% of all cases) and distention1. It has no imaging characteristics and very difficult to diagnose preoperatively. Diagnosis is possible by means of immunohistochemistry stain for S-100 protein which distinguish schwannomas from spindle cell tumours3. As the tumour is resistant to chemo-radiotherapy2, surgical approach should focus on complete excision of the mass5. This is a unique case of giant retroperitoneal schwannoma mimicking an adrenal tumour, encroaching upon renal hilum and posing threat to kidney. Application of all existing technological support and ground-breaking techniques of resection has evolved aiming to reduce blood loss and save the kidney.

Case presentation and management
Patient’s History: A 55 years old lady presented with pain in the left side of upper abdomen and lump in the left hypochondriac and lumbar region for eleven months. Bladder and bowel habit of the patient was normal. The patient was mildly anaemic having below average nutritional status. Hemoglobin level was 9gm/dl and other biochemical parameters were normal.
Ultrasonography of whole abdomen revealed two big mixed echoic lump in left subcostal and lumbar region, the bigger one was compressing left renal hilum without any hydronephrosis. USG guided FNAC was inconclusive. Other site of abdomen was normal. CT scan was not possible due to poor financial condition of the patient. Anaesthetic fitness of the patient was within the normal limit. Five units of blood were arranged and two units were transfused preoperatively.

*Surgical procedure:* Under GA, the abdomen was explored through long midline incision, after packing of the small intestine into right side of the abdomen the bigger lump was exposed. Prominent blood vessels were observed on the surface of the lump (Fig. 1). Entrance into the retroperitoneal space was made after stripping off the peritoneum. Finger dissection was used to separate the large vessels from the lump and divided, ligated between the artery forceps. During separation of the lump from the posterior aspect, it was found tightly adherent to the left renal hilum.

After clearing of the operative field, the renal pedicle was ligated (Fig. 3). Due to avulsion of the kidney, it was removed (Fig. 4). Another small lump (7/6 cm) was removed successfully from the left subcostal region without further damage (Fig. 4). Wound was closed in layers with a drain kept in situ after securing every bleeding points. Total duration of operation was about two hours. Two units of blood were transfused peroperatively and one unit postoperatively. Condition of the patient remained stable during operation. In post operative period the patient remain well and recovered uneventfully. Histopathological section of two tumours revealed benign schwannoma.

Discussion:
Retroperitoneal schwannoma is a very rare tumour. Preoperative diagnosis is very difficult despite modern equipped facilities. Retroperitoneal tumours usually
present without gastrointestinal or urological manifestations. Ultrasonography provided good clue to detect the origin of tumours and condition of the ipsilateral kidney. Fine-needle aspiration cytology was not helpful. CT scan was not done due to financial constrain. Accurate diagnosis is absolutely based on immunohistochemistry.

Complete excision of the tumour is the only hope of cure. Adequate preoperative evaluation is mandatory to reduce operative morbidity. Long midline incision offers adequate exposure to almost every region of the abdominal cavity and retroperitoneum. It is unsurpassed when speed is of the essence.

Laparoscopic resection is an alternative choice, but supervised training and support should be ensured. In our case, standard resection of the tumours were completed. Preservation of the left kidney was not possible. Facilities of vascular anastomotic technique might have save the kidney.

Survival rate depends upon whether tumour is benign or malignant. Complete excision can cure benign case, survival rate of malignant schwannoma is 85% of 11 years (+5). Recurrence is common in partial excision.

**Conclusion:**
Retroperitoneal schwannoma is a locally aggressive tumour. Preoperative diagnosis is difficult with CT scan or FNAB. Surgical approach should focus on complete excision of the mass. Patient undergoing complete resection tend to do well without evidence of early recurrence.

**References:**