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Case Report
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Retroperitoneal Dumbbell Neurofibroma Presenting as Wilms' Tumour with Postoperative Spinal Cord Compression



*Govani DR[1], Mehta AR[2], Midha PK[3], Govani ND[1], Panchasara NG[1], Patel RR[1] and Patel RV[1]**

[1]*Department of Pediatrics and Pediatric Surgery, Postgraduate Institute of Child Health & Research and KT Children Govt. University Teaching Hospital, Rajkot 360001, Gujarat, India*

[2]*Formerly Head, Department of Surgery at Tata Memorial Hospital, Mumbai, India*

[3]*J. Watumull Global Hospital & Research Centre, Delwara Road, Mount Abu, Rajasthan 307501, India Affiliated to Medical Faculty of God Fatherly Spiritual University, Mount Abu, Rajasthan, India*

Abstract

A six-year-old girl presented with a left-sided abdominal mass and a non-functioning left kidney, initially diagnosed radiologically as Wilms' tumor. Intraoperatively, the kidney was found to be compressed—not invaded—by a massive, benign-appearing retroperitoneal tumor. The mass was excised with preservation of the kidney. Unrecognised extension of the tumor through the lumbar intervertebral foramen resulted in a postoperative spinal epidural hematoma, causing acute paraparesis. Emergency lumbar decompression was performed, with partial neurological recovery. This case highlights the diagnostic challenge posed by giant dumbbell neurogenic tumours and the importance of preoperative spinal imaging when retroperitoneal masses abut the vertebral column.

Keywords: Dumbbell Neurofibroma; Lumbar Spinal Tumor; Retroperitoneal Mass; Wilms' Tumor Mimic; Pediatric Abdominal Mass; Non-Functioning Kidney; Renal Compression; Spinal Extension; Epidural Hematoma; Postoperative Paraparesis; Lumbar Decompression; Neurogenic Tumour; Diagnostic Pitfall; Spinal Cord Compression; Emergency Laminectomy

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Correspondence:

Dr. Ramnik Patel, M.D., Director-Professor, Department of Pediatric Surgery, Postgraduate Institute of Child Health and Research and K T Children Government University Teaching Hospital, Rajkot 360005, Gujarat, India. Mobile: +447956896641, Phone/Fax: +441162893395; E-mail: ramnik@doctors.org.uk/ ORCID: https://orcid.org/0000-0003-1874-1715

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Introduction

Retroperitoneal masses in children are most commonly malignant renal tumours, particularly Wilms' tumour [1–4]. Neurogenic tumours of the lumbar plexus are rare in this age group and may mimic renal malignancy both clinically and radiologically. Dumbbell neurofibromas extending through the intervertebral foramina are well described in adults but are exceptional in children and seldom present as abdominal masses [5–12].

This case illustrates a rare diagnostic pitfall: a giant lumbar neurofibroma masquerading as Wilms' tumour, compressing—but not infiltrating—the kidney. The unrecognised intraspinal component created a unique postoperative risk, culminating in acute spinal cord compression from a postoperative epidural hematoma. The case provides important learning points for paediatric surgeons, radiologists, and neurologists.

Case Report

A previously healthy six-year-old girl presented in 1988 with a two-month history of progressive abdominal distension and intermittent left flank discomfort. There were no neurological symptoms, urinary tract infections, or constitutional features.

Examination

A firm, non-tender left-sided abdominal mass extended from the left hypochondrium to the iliac fossa. There were no café-au-lait spots or cutaneous stigmata of neurofibromatosis. Lower limb neurology was normal.

Investigations

Laboratory tests were normal except for anemia (Hb 8 g/dL). Chest radiograph was normal. Abdominal radiograph showed a large soft-tissue mass displacing bowel loops to the right.

Ultrasound demonstrated a large, smooth, rounded heterogeneous mass arising inferior to the lower pole of the left kidney and displacing it superiorly. Intravenous urography showed a non-functioning left kidney compressed by a well-circumscribed mass presumed to be Wilms' tumor.

CT and MRI were not available at the time. Retrospective review of the abdominal radiograph and the ultrasound suggested subtle paravertebral extension abutting the L2–L4 foramen.

Differential diagnosis included Wilms' tumour (primary working diagnosis), retroperitoneal sarcoma, neurogenic tumour (not initially suspected) and extrarenal rhabdoid tumour.

Treatment

The child was scheduled for emergency tumour excision with presumed left nephrectomy, following the National Wilms Tumor Trial protocol of that era.

Intraoperative Findings

A giant, encapsulated, benign-appearing retroperitoneal tumour was found compressing the left upper ureter and displacing a structurally normal left kidney. The tumour was not highly vascular, but decompression after excision resulted in persistent venous oozing from the capsule. To achieve hemostasis and avoid blood transfusion in this anemic child, the capsule was closed to provide tamponade.

The surgical team was unaware of the tumor's extension through the intervertebral foramen into the spinal canal.

Early Postoperative Course

The child recovered initially, passing flatus on postoperative day 2 and tolerating oral feeds. Care was handed over to the general surgical head of department, who had prior paediatric surgical experience.

On postoperative day 3, the child developed progressive left-leg weakness and urinary retention. The paediatric surgeon returned to duty and reviewed the patient. Histopathology from the abdominal tumour confirmed a benign neurofibroma.

Neurological Deterioration

Examination revealed left-sided weakness greater than right, with bladder involvement. Urgent myelography demonstrated a large intraspinal component of the dumbbell neurofibroma, an epidural hematoma extending from L2–L4 and compression of the cauda equina.

No orthopedic pediatric spinal surgeon was available. The paediatric surgeon, experienced in spinal decompression for tuberculosis in Africa and at home, proceeded with emergency surgery.

Emergency Decompression

The child underwent lumbar laminectomy, evacuation of the epidural hematoma and decompression of the spinal component of the dumbbell neurofibroma (Figure 1).

The excised specimen included a large, smooth, well-capsulated intravertebral component and the connecting bridge through which slow venous bleeding had dissected the capsule, causing delayed spinal cord compression (Figure 2).

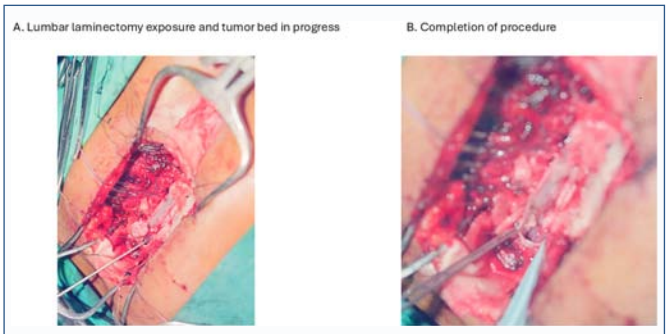


Figure 1: Intraoperative Emergency Lumbar Laminectomy.
A. Intraoperative view showing decompression in progress at the L2–L4 level. The posterior vertebral elements are being removed to access the epidural space.
B. Completion of laminectomy with full exposure of the dural sac and evacuation of the epidural hematoma and spinal component of the tumor. The spinal cord is visibly decompressed. Note the tumor bed on the left side with residual tumor extension through the intervertebral foramen which is all been excised and removed.

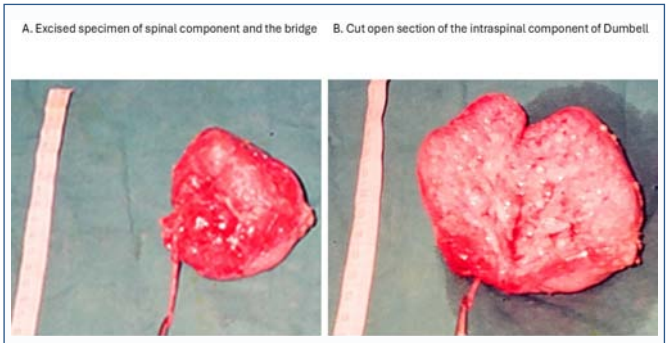


Figure 2: Gross Pathology of Excised Neurofibroma.
A. Intact excised spinal component of the dumbbell neurofibroma, including the bridge connecting the retroperitoneal and intraspinal segments. The mass is encapsulated and well demarcated.
B. Cut-open section of the tumor showing a firm, grey-white, well-capsulated mass with classical features of a benign neurofibroma, including whorled architecture and absence of necrosis or atypia.

Outcome and Follow-Up

Right lower limb power improved significantly within 48 hours. Left-sided weakness improved gradually over several weeks. At three months, she had mild residual weakness but was ambulant with physiotherapy support.

Histopathology confirmed a benign neurofibroma. Long-term follow-up MRI performed 30 years later showed no recurrence or progression of either the foraminal or retroperitoneal components.

Discussion

In pediatric cases, a giant benign dumbbell neurofibroma can present as a diagnostic challenge because its massive size and location may mimic Wilms' tumor (nephroblastoma), the most common childhood renal malignancy [1-4]. While Wilms' tumor typically presents as a painless abdominal mass, it can rarely involve the spinal canal through direct extension. This case underscores several important clinical lessons:

Neurogenic tumors can mimic renal malignancy

Large lumbar neurofibromas may present as abdominal masses, displacing the kidney and mimicking Wilms' tumor. The absence of

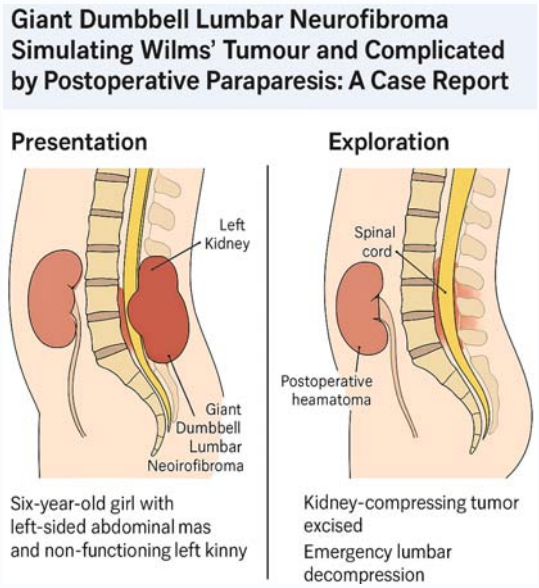


Figure 3: Graphical Abstract.
Schematic illustration summarising the case:
Left panel (Presentation): A six-year-old girl with a left-sided abdominal mass compressing the kidney, initially diagnosed as Wilms' tumor.
Right panel (Exploration): Intraoperative findings reveal a giant dumbbell neurofibroma with hidden spinal extension. Postoperative epidural hematoma causes cord compression, necessitating emergency lumbar decompression.

renal invasion and the presence of a smooth, rounded mass should prompt consideration of alternative diagnoses.

Dumbbell extension may be easily overlooked

The tumor's extension through the intervertebral foramen was not appreciated preoperatively. However, the intervertebral pedicular distances were increased and attenuated on the retrospective detailed examination of the abdominal radiographs and the ultrasound scans. The CT scan or an MRI were not available during those days. MRI is superior to CT for evaluating paraspinal masses and should be considered when a retroperitoneal tumor abuts the vertebral column. The kidney is displaced rather than infiltrated. The mass appears unusually well-circumscribed.

Postoperative hematoma risk is increased when the spinal component is unrecognised

Closure of the tumor capsule to achieve hemostasis inadvertently created a closed compartment, allowing postoperative bleeding to track into the spinal canal.

Early recognition and decompression are critical

Rapid neurological deterioration required urgent decompression. The degree of recovery correlated with the speed of intervention.

Diagnostic challenges especially when no cross imaging was available were real and beyond imagination retrospectively [6]. Simulation by rare benign tumors should be kept in the back of the mind. A large retroperitoneal or abdominal neurofibroma can occupy the flank, appearing clinically and radiologically similar to a renal tumor and can mimic a Wilms' tumor [7].

Imaging features are distinct and separate from the Wilms' tumor. Initial ultrasound or CT may show a heterogeneous mass crossing the midline. MRI is the gold standard to identify the "dumbbell" shape, where the tumor has both an extraspinal (abdominal/mediastinal) and an intraspinal component connected through the neural foramen [8].

Complications of epidural hematoma low pressure gradually over days if the tumor capsule was closed it has no escape and collected slowly in the epidural space. The gradual spinal cord compression and paraparesis followed [7].

The mechanism was not the rapid growth of the intraspinal portion of the dumbbell neurofibroma compressing the spinal cord, leading to progressive neurological deficits but slow bleeding and gradual hematoma in the epidural space which was already occupied by the spinal component [5].

Clinical presentation of this complication can be picked up early by high index of suspicion especially when the tumor was benign and histology has suggested neurofibroma, dumbbell component, intraoperative slow oozing and closure of the capsule should all alert one proactively to prevent such a catastrophic complication. It was particularly applicable in our case when the tumor itself was very benign and patient had normal life expectancy. Patients may present clinically in established cases with paraparesis (weakness of the lower limbs), gait alterations, or sensory changes [8].

Neurological risk in retroperitoneal neurofibroma is real and should be anticipated. In some pediatric cases, significant compression can exist even in the absence of severe symptoms, making early MRI critical to prevent permanent damage [9].

Management is surgical intervention as primary treatment by surgical decompression of the epidural hematoma and resection to alleviate compression and restore function. A multidisciplinary team of neurosurgeons and pediatric surgeons is often required.

Prognosis and outcome depends upon rapid diagnosis and immediate correction [10]. Following decompression and total resection, patients typically show gradual improvement in motor power and pain relief.

Differential diagnosis considerations are crucial under similar situation. Accurate diagnosis via biopsy is essential, as malignant tumors like Wilms' or neuroblastoma require chemotherapy, whereas benign neurofibromas are primarily managed surgically.

Historical Diagnostic Limitations

In 1988, cross-sectional imaging was unavailable. The diagnostic uncertainty was therefore substantial, and the mimicry by this rare benign tumour was profound.

Learning Points

- Not all paediatric abdominal masses with a non-functioning kidney are Wilms' tumors; neurogenic tumors should be considered when the kidney is displaced rather than invaded.
- Preoperative MRI is essential when retroperitoneal tumors lie adjacent to the spine or show paravertebral extension.
- Dumbbell neurofibromas may present atypically and carry a risk of postoperative spinal complications if the intraspinal component is unrecognised.
- Prompt recognition and emergency decompression of postoperative spinal hematoma are vital to optimise neurological recovery.

Conclusion

This manuscript describes a rare and diagnostically challenging presentation of a giant lumbar neurofibroma in a six-year-old girl, initially misdiagnosed as Wilms' tumor due to renal compression and a non-functioning kidney. Intraoperative findings revealed a benign retroperitoneal mass with unrecognised spinal extension, leading to postoperative epidural hematoma and acute paraparesis. Emergency decompression was performed, with partial neurological recovery. It highlights the importance of preoperative spinal imaging in retroperitoneal masses adjacent to the vertebral column, the diagnostic pitfalls of renal displacement versus invasion, and the surgical risks posed by hidden foraminal extension.

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