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Retroperitoneal Ganglioneuroma: A Rare Case Presenting As Right Ureteric Colic

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ABSTRACT

Background: Ganglioneuromas are rare benign tumors arising from the autonomic nervous system, and they are composed of well-differentiated Schwann and ganglion cells.

Case Report: An 11-year-old girl presented with colicky right loin pain that had occurred on and off for the past 1 year. On evaluation, she was found to have a retroperitoneal mass of 6 cm x 4 cm just below the right renal hilum. Her laboratory parameters were normal. The mass was hormonally silent. Surgery was performed and the mass was resected completely. The mass was closely opposed to the inferior vena cava with tributaries to it. Fibrous attachments were also noted on the spine, which were freed. Her histopathology and immunohistochemistry confirmed the mass to be a ganglioneuroma. Her neuron-specific enolase marker was positive. She was symptom free at 3 months post surgery, and her imaging was normal.

Conclusion: Ganglioneuromas have an excellent prognosis. This case is presented for its rarity and unique presentation as right colicky pain.

CASE PRESENTATION

An 11-year-old girl presented with complaints of right loin pain for the past 1 year. The pain was colicky in nature. It was associated with nausea but there was no vomiting. There was no fever. She did not have any bowel or urinary complaints. There was no hematuria. Her general examination was unremarkable. Her blood pressure was normal. She did not have any lymph nodal enlargement. There was no renal angle tenderness on the right side.

On evaluation, an ultrasound displayed a retroperitoneal mass lesion. The ultrasound revealed a 6 cm x 4 cm well-defined hypoechoic lesion medial to the right kidney. There was mild hydroureteronephrosis noted on the right side up to the upper ureter. Her laboratory parameters were normal. The urinary catecholamine levels were normal. The contrast-enhanced computed tomography (CT) scan revealed a 6 cm x 4 cm homodense lesion just below the right renal hilum and medial to the right ureter. There was minimal hydroureteronephrosis noted up to the upper ureter on the right side. It was closely opposed to the inferior vena cava (IVC). There was no calcification or necrosis noted within the mass. No enhancement was noted, even in the delayed films. Magnetic resonance imaging (MRI) was not done due to cost constraints.

She was advised to have an excision biopsy of the retroperitoneal mass lesion. Intraoperatively, under general anesthesia, she was placed in the right flank position and a subcostal incision was made. The retroperitoneal mass was excised in toto through the retroperitoneal approach, taking care not to injure the renal vessels and the ureter. Perioperatively, the kidney was displaced above and laterally by the mass. The mass was lying on the IVC with tributaries to the IVC (Figure 1). The tributaries were isolated, ligated, and cut. The IVC was not injured during the surgery. There were fibrous attachments to the spine, which were freed, and the mass was excised completely (Figure 2). The operation lasted for a little over 3 hours. The postoperative period was uneventful.

KEYWORDS: Ganglioneuroma, retroperitoneal mass, retroperitoneal ganglioneuroma, ureteric colic

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Figure 1. Intraoperative picture showing the mass lying on the IVC.

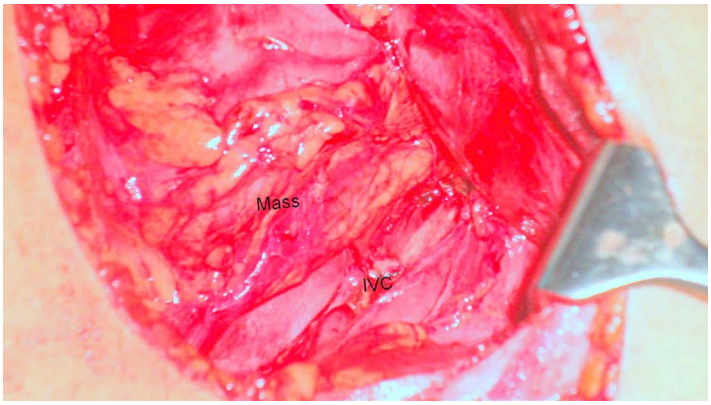
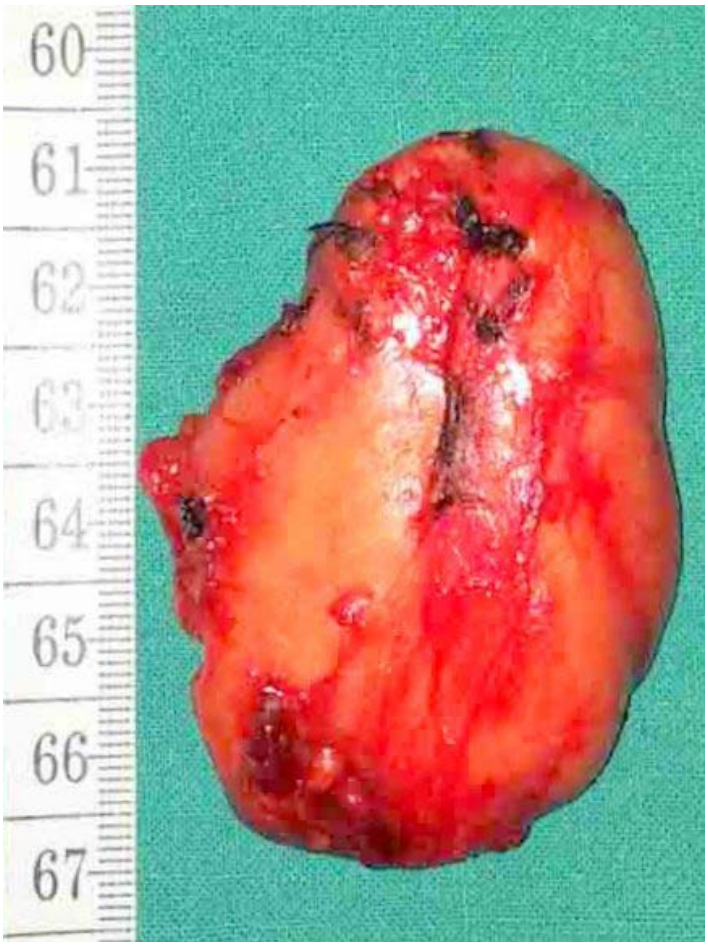


Figure 2. The excised specimen.



Histopathology of the lesion confirmed a ganglioneuroma. There were well-differentiated Schwann cells, with eosinophilic cytoplasm and ganglion cells. Immunohistochemistry was done for neuron-specific enolase, which was positive. The Ki-67 proliferation index was 2 to 3%. The patient recovered well after the procedure and was discharged on the fourth postoperative day. Her symptoms were completely relieved. She remained asymptomatic at her 3-month follow-up, and an ultrasound of the abdomen was normal.

DISCUSSION

Loretz first described ganglioneuroma in 1870. Ganglioneuroma is a rare, benign neuroblastic tumor arising from the immature cells of the sympathetic nervous system. These tumors consist of mature Schwann cells, ganglion cells, and nerve fibers.
It is commonly seen in the second and third decade of life, with a slight preponderance in females. The most common locations, in descending order, are the retroperitoneum, the mediastinum, the adrenal medulla, the cervical region, and the presacral area. Rarely, they can be associated with neurofibromatosis type I. They usually arise in relation to the sympathetic chain or adrenal medulla. It usually grows slowly to a large size before presenting with symptoms, which occur predominantly due to the compression of adjacent structures. Our patient had symptoms due to intermittent compression on the right ureter.

Usually, ganglioneuromas are hormonally silent. Rare cases have been found to be functioning, with the secretion of catecholamines or vasoactive intestinal peptides [5]. An hourglass extension into the spine had also been observed [2]. Very rare associations have been reported with seminoma [4] and pheochromocytoma [6]. Very rarely, malignant transformation has been noted [3]. The CT description is usually of a homodense lesion with well-defined margins and delayed contrast enhancement [1,7]. Discrete or punctate calcification may be noted in up to 20% of cases [7]. The mass can also be heterodense due to necrosis, when a mass outgrows its blood supply. The peculiarity in our case was that there was no enhancement at all, even in delayed films.

MRI usually reveals a homointense lesion with low signal intensity on the T1 weighted images. On the T2 weighted images, they are hyperintense, but the extent depends on the ratio of myxoid stroma to the cellular components [7]. MRI would be better than CT to predict intraspinal involvement. A nuclear bone scan had been suggested, rather than CT, for the detection of bony erosions [8].

The diagnosis of retroperitoneal ganglioneuroma is difficult to clinch preoperatively, and they are usually diagnosed by histopathological examination. Nevertheless, they should be considered in the differential diagnosis of retroperitoneal and

posterior mediastinal masses. Ganglioneuromas are usually cured with surgical excision. Some masses are not amenable for complete excision due to the location adjacent to or circumscribing major vessels. Even with subtotal resection, they usually do not recur. No adjuvant therapy is indicated. The prognosis is excellent. These patients do not require long-term follow-up.

CONCLUSION

This case is presented due to the rarity of the situation. The peculiarities about this case are the mode of presentation, with symptoms due to compression of the right ureter and the total absence of delayed enhancement on the CT scan.

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