

Ganglioneuroma of pelvis – an unique presentation in a young man

Nirmal Lamichhane¹ and Hari Prasad Dhakal²

¹Department of Surgical Oncology, ²Department of Pathology, B P Koirala Memorial Cancer Hospital, Bharatpur, Chitwan, Nepal.

Corresponding author: Nirmal Lamichhane, Dept. of Surgical Oncology, B P Koirala Memorial Cancer Hospital, Bharatpur P O Box: 34, Chitwan, Nepal. email: nlamichhane@hotmail.com

ABSTRACT

Ganglioneuromas presented as a pelvic tumor around the pelvic organs is a rare entity. A case with unusual presentation is reported. Young man of 18 years old presented with a complaint of lower abdominal mass increasing in size for last 3 years. It was treated with partial resection for debulking purpose after the conformation during surgery with frozen section. Debulking surgery with preservation of organ functions is feasible in these slow growing tumors for better quality of life.

Keywords: Ganglioneuroma, pelvis, surgery.

INTRODUCTION

Ganglioneuromas are neurogenic tumors arising from sympathetic nerve ganglions. These tumors are commonly seen in young population.¹ They are highly differentiated benign tumors and are compatible with long-term disease free survival even though surgical treatments are unsatisfactory.² Although retroperitoneal localization is relatively frequent for these tumors, presentation as a pelvic mass is also seen.¹ Here, we present a case of ganglioneuroma arising in pelvis going up into the retroperitoneum, which was undiagnosed before operation.

CASE REPORT

The young man was 18 years old male coming to us with complains of painless lower abdominal mass gradually increasing in size for last 3 years. He had history of occasional per rectal bleeding for last 6 months of visit. The bleeding was fresh in nature and occurred at the end of defecation. There was no mucous in the stool. He was well oriented with a good appetite and unaltered bowel and bladder habits.

On physical examination his vital signs were normal. He looked pallor. There were some soft elongated nodes at the posterior triangle of neck on both sides, size varied from 0.8 to 1.2 cm in diameter. Chest was clinically clear. On per abdominal examination, about 15x15 cm² nontender, non fluctuating mass that can be moved a little bit was palpated at lower mid abdomen. No hepatosplenomegaly was there. No shifting dullness was there. Bowel sounds were existing and normal. Bilateral hernial orifices were normal. Bilateral testicles were normal in the scrotum. On per rectal examination mucosa was intact but there was a soft nodular thickening (like a pad) of the rectal wall palpated around it. There was no growth or hemorrhoids seen through the proctoscopy.

On Investigations hemoglobin was 6.5 g/dl, other blood parameters were within normal limit. Chest x-ray revealed nothing, Liver Function Tests and Renal Function Tests were normal. Computed Tomography (CT) scan of the abdomen and pelvis was taken that showed a mass of homogenous density was arising from the pelvic retroperitoneum encircling the rectum covering the urinary bladder from posterior wall coming towards dome and anteriorly. It was spreading into the retroperitoneum at abdomen and bilateral iliac vessels (Fig 1,2,3). Other abdominal viscera looked normal. Occult blood was negative and Sigmoidoscopy revealed nothing.

Based on above examination and with the suspect of lymphoma we did a excision biopsy of neck nodule at right side that came to be a neurofibroma. We did fine needle aspiration cytology examination of the lesion that revealed soft tissue tumor. As all were inconclusive for management, laparotomy was done for exploration and biopsy. It revealed a 15 x 10-cm² soft to medium consistency mass was arising from the pelvic peritoneum posterior to the bladder. The tumor was extending to the retroperitoneum and was extending downwards encircling the rectum as well.

Pouch of Douglas was pretty much elevated upwards, sigmoid colon was short. It does not look like lymphoma, so we sent a frozen section of the piece of mass. That revealed it to be a Neural tumor possibly Ganglioneuroma with myxoid changes but not a lymphoma. We decided to debulk the tumor as total removal needed total pelvic exenteration that would be too much mutilating for him. Urinary Bladder was opened from its anterior wall, bilateral ureters were cannulated. Tumor with bladder dome and bulk of mass above it and mass posterior to it was excised and about 1000 gram of tumor tissue removed. The bladder is repaired. On cross section the tumor was white in color with multiple nodules of varying sizes (0.5 to 1.0 cm) in it. Ascites was nil. No peritoneal seedling of tumor was there. Post-operative period was uneventful, patient was discharged on 8th postoperative day. Final permanent section conformed the diagnosis of Ganglioneuroma (See Fig. 4). As he had same kind of tissue removed by biopsy of the neck nodule, it was diagnosed to have Ganglioneuromatosis.

DISCUSSION

Neuroblastomas, ganglioneuroblastomas and ganglioneuromas are tumors of the sympathetic nervous system that arise from the neuroectodermal cells derived from the neural crest cells.¹ Among these, ganglioneuromas are the most common neoplasm of the sympathetic nervous system in adults.³ Ganglioneuromas are thought to be the fully differentiated counterpart of neuroblastomas. They may occur spontaneously or during the therapy for neuroblastomas with either chemotherapy or radiation therapy.²

Ganglioneuromas common in 10 to 30 years old age group with more predominance in pediatric age group. The reported incidence of this disease is one per million population. They are mostly sporadic but there are a few reports of ganglioneuromas associated with neurofibromatosis type II and multiple endocrinologic neoplasia type II.⁴ Ganglioneuromas can be found in the central nervous system or peripherally in the sympathetic system.⁵ The most common localization is the posterior mediastinum followed by the retroperitoneal space.¹ Among the primary retroperitoneal tumors, they constitute only a small percentage of 0.72 to 1.6.² Pelvic retroperitoneal localization like in our's is even rarer. There are a few reports of ganglioneuroma involving the lesser pelvis and pelvic floor.⁶⁻⁸

Retroperitoneal ganglioneuromas are usually non-functioning and asymptomatic until they reach large sizes in which case they cause symptoms due to local expansion and pressure on adjacent structures.² Although symptoms of autonomic dysfunctions are usually seen in patients with hormone secreting ganglioneuromas, such symptoms may also be seen in patients with paravertebral ganglioneuromas compressing the autonomic fibers of the lumbosacral plexus.⁹ Also, there are functional ganglioneuromas that were found to release peptides such as vasoactive intestinal peptides (VIP), somatostatins and Neuropeptide Y (NPY) in the literature.^{1,10} These tumors may cause some symptoms like diarrhea, sweating and hypertension related to those peptides. Diarrhea in this patient can be caused by this kind of intestinal peptides. Since ganglioneuromas may release catecholaminergic peptides, surgeons should be aware of the possibility of hypertensive crisis during the surgery.^{1,10,11}

Preoperative diagnosis of retroperitoneal ganglioneuroma is often difficult and the diagnosis is usually based on histopathological findings after surgical excision of the tumor. Although in some cases aspiration cytology with fine needle has been reported to be useful in the preoperative diagnosis of adrenal ganglioneuroma, since the tumoral tissue can contain fractions of less well differentiated areas, surgical exploration is required to achieve a definitive diagnosis and risk assessment.^{4,10}

Radiological examination also has no diagnostic value in most cases. In a study of thirteen retroperitoneal ganglioneuromas, it was concluded that unless typical CT or MRI findings are present, diagnosis of ganglioneuromas is difficult radiologically.² A well-circumscribed mass with a tendency to partially or completely surround blood vessels without compressing the lumen was the mainstay of these findings.⁵ Visualization of this close relation perhaps may be the most important gain of imaging techniques before attempting a large excision for the surgeons. For the pure ganglioneuromas, heterogeneous high signal intensity on T2 weighted MR images may be helpful in the differential diagnosis of other retroperitoneal masses but more studies are needed especially for the mixed pathologies such as the ganglioneuroma-pheochromocytoma combination.^{5,12} Because of the rarity of retroperitoneal ganglioneuromas and absence of any

characteristic radiologic features, imaging of these tumors is not reliable and diagnostic.¹

In some studies, certain tests for elevated serum or urinary hormonal levels were studied as a screening test in patients with retroperitoneal tumors but larger series are needed to postulate any universal marker for the specific and differential diagnosis of ganglioneuromas and neuroblastomas.^{1,2,3,13}

Grossly, they are large, encapsulated masses of firm consistency with an homogenous, solid, grayish white cut surface. Areas with different color or consistency should be sampled for microscopic examination with the suspicion of less differentiated foci. They can be multiple and or associated with other independent types of neurogenous neoplasms such as neuroblastoma and pheochromocytoma.³ Microscopically, it consists of a spindle cell tumor resembling a neurofibroma but shows numerous ganglion cells. Microscopically ganglioneuromas have two subtypes. The mature subtype consists of a spindle cell tumor resembling a neuroblastoma but have fascicles composed of neuritic processes, Schwann cells and perineural cells and show numerous ganglion cells. The maturing subtype has a similar stroma but with ganglion cells of differing maturation, from fully mature ones to neuroblasts. Immunohistochemically they are characterized by reactivity with S100 and neuronal markers such as NSE and synaptophysin.¹⁴

Ganglioneuromas are typically slowly growing, benign tumors and have a tendency to remain clinically silent for a considerable time. They can occur as a result of the necrosis of immature neuroblasts from the malignant neuroblastomas in the primary or any metastatic site. The probability of reoccurrence of neuroblastoma from the ganglioneuroma is not known but at least one case has been reported 15 years after conversion to ganglioneuroma. Most patients have prolonged survival without any evidence of progression.²

According to many authors, surgical excision is sufficient for the treatment.¹⁵ Preoperative or postoperative chemotherapy or radiotherapy have no value in the treatment except it was associated with ganglioneuroblastoma changes whenthere might be some role of chemotherapy.² Even with the residual disease; cessation of all other treatments and a close follow-up may be adequate. If any progression of the tumor is seen during follow-up, re-biopsy or laparotomy may be indicated.

With the knowledge of the tumors biology, surgeons may attend to less radical operations in the aim of total resection of the tumor. Due to the close relations to large vessels, some potentially life-threatening complications may be seen during the surgical resection. In this particular case, if we go for total surgical removal, it definitely needed a total pelvic exenteration. It definitely raises a Quality of life (QOL) issue for he has no bowel or urinary symptoms so far. According to our knowledge, this is the first diagnosed case of ganglioneuroma of abdominal cavity in our hospital.

Although pelvic ganglioneuromas are very rare lesions, it should be kept in mind for the differential diagnosis of pelvic masses. As it is a slow growing tumor, gross total surgical removal with preservation of organ functions is a feasible surgical option.

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Figures:

Fig. 1. CT scan of abdomen showing Retroperitoneal widening of soft tissues. GI tract contains 1.0% barium.

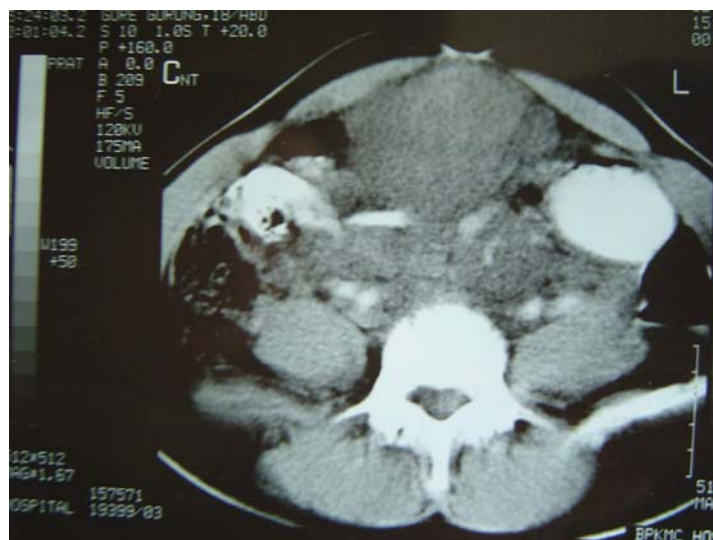


Fig. 2. CT pelvis showing Bulk of the tumor with some contrast media in the the Urinary bladder.

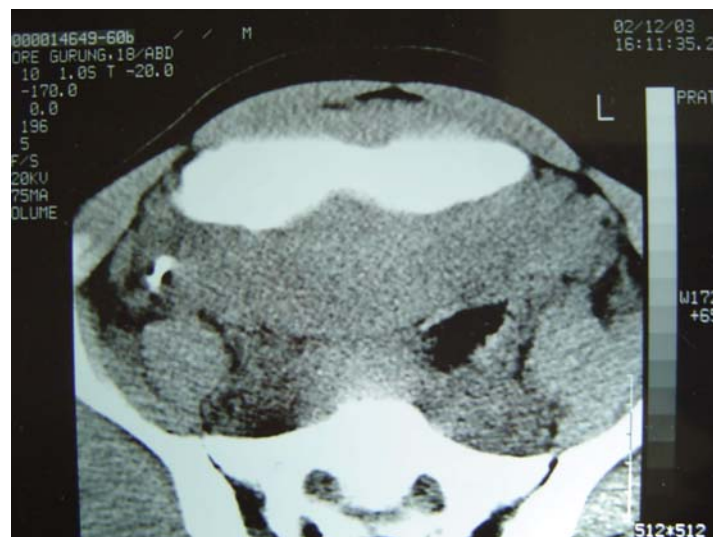
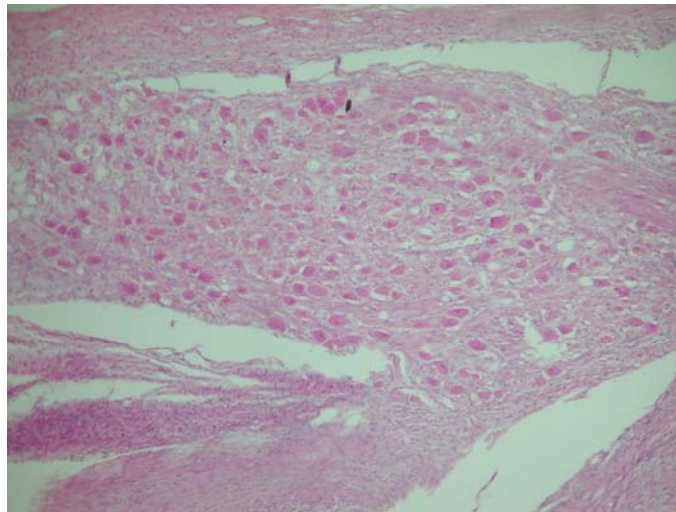


Fig. 3. CT pelvis showing soft tissue shadow around the rectum.



Fig. 4. Microphotograph of the histopathology slide on H&E Staining showing ganglion cells on X 10 magnification.



Addendum:

Published as an abstract in the first national oncology conference at bharatpur, chitwan, NEPAL.
Dec 4-6, 2004.