

# Incidentally Detected Rare Retroperitoneal Tumor: Ganglioneuroma

Syed Khalequezzaman<sup>1</sup>, Sania Ahsan<sup>2</sup>, Dilruba Khandker<sup>3</sup>, Biva Shrestha Khan<sup>4</sup>, Atia Huda<sup>5</sup>

1. Consultant, Department of Radiology and Imaging, Evercare Hospital Dhaka
2. Sr. Consultant- Neuro Radiology, Department of Radiology and Imaging, Evercare Hospital Dhaka
3. Associate Consultant, Dept. of Radiology & Imaging, Evercare Hospital Dhaka
4. Sr. Consultant, Dept. of Radiology & Imaging, Evercare Hospital Dhaka
5. Ex. Sr. Consultant, Dept. of Radiology & Imaging, Evercare Hospital Dhaka

### Address for Correspondence:

Dr. Syed Khalequezzaman  
Consultant, Department of Radiology and Imaging,  
Evercare Hospital Dhaka  
syed.khalequezzaman@evercarebd.com

### ABSTRACT

A primary extra-adrenal retroperitoneal ganglioneuroma was found incidentally in a 22-year-old woman during screening for suspected pregnancy by transvaginal ultrasonography (TVS) and subsequently by magnetic resonance imaging (MRI). TVS and MRI revealed well defined solid mass in retroperitoneal at the level of L5 to S2 vertebrae and displace right common iliac and external iliac vessels. Limited cuts of the region of Computed tomography (CT) were taken and no calcification was identified. Histological examination showed that the lesion was an extra-adrenal retroperitoneal ganglioneuroma composed of mature ganglion cells and subsequently confirmed by immunohistochemistry

**Key words:** *Ganglioneuroma, retroperitoneal*

## INTRODUCTION

Ganglioneuromas are rare benign neoplasms arising from sympathetic ganglia. They belong to the group of neurogenic tumors. Though the ganglioneuromas is a rare tumor, it has to be taken into consideration in the differential diagnosis of retroperitoneal tumor. This tumor is usually asymptomatic and in the majority of cases is detected incidentally. Although the characteristics of ganglioneuroma on computerized tomography and magnetic resonance imaging have been well described, the exact diagnosis is difficult. Histopathological examination is currently the mainstay of diagnosis. Ganglioneuromas have a very good prognosis with surgical removal. We report the case of a young female patient with an incidentally identified retroperitoneal ganglioneuroma diagnosed histopathologically and confirmed by immunohistochemistry after successful operation through the abdominal approach in toto. Follow up MRI revealed no residual tumor.

## CASE SUMMARY

22 years young recently married lady reported to Gyne & Obs OPD with history of missed period and suspecting of having pregnancy. The referring physician advised to do TVS and sent the patient to Radiology & Imaging department.

TVS was done and an incidentally detected well outlined solid ovoid heterogeneously hyperechoic round retroperitoneal mass of about 9.5 x 5.4 x 8.0 cm, vol- 280 cc was noted anterior to the right hemi sacrum and lower lumbar vertebrae and it showed sparse internal vascularity (Fig.1). The mass indented and pushed back the uterus. MRI of lower abdomen was advised for further characterization.

Following day, she underwent MRI of pelvis with contrast. It showed a well-defined mass in the median and right para median pelvis extending from L5 to S2 levels, which was iso- intense to muscle on T1 W and mildly hyper- intense on T2W sequences. The mass was broad based at S1 level and extending to right S1 neural foramen (Fig.2a) and displacing right internal and external iliac vessels laterally (Fig.2c). The mass

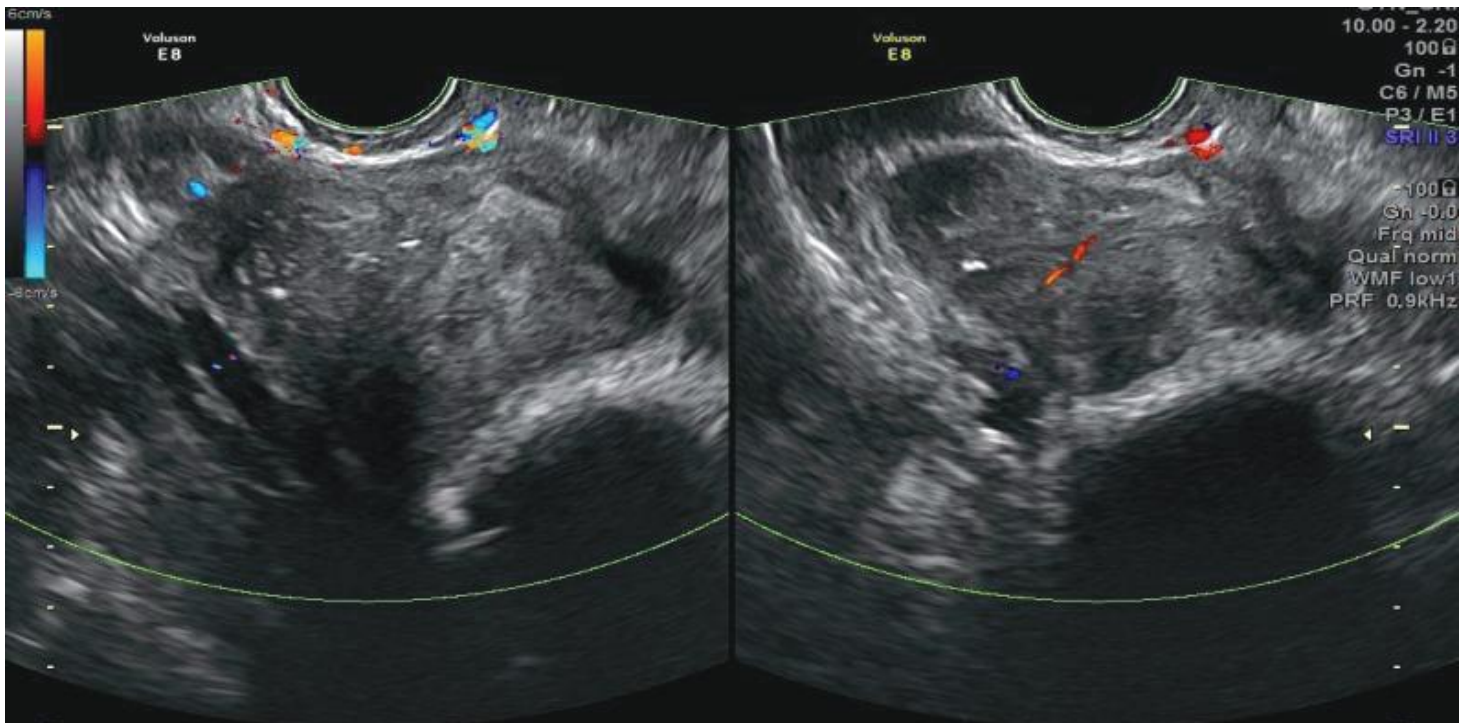
## Case Report

was not diffusion restricted (Fig.2b) and showed minimal post contrast enhancement (Fig.2c). MRI also showed polycystic appearing ovaries which was confirmed by observation during surgery (Fig.2d). The mass was separate from broad ligament and ilio-psoas muscle. Calcification is not sensitive by MRI, hence plain CT scan was done at the same setting for detection of any macro or microcalcification. However, no calcification within the mass was detected (Fig.3). She was advised to get admission for surgical management. After admission, her routine blood and urine test, liver

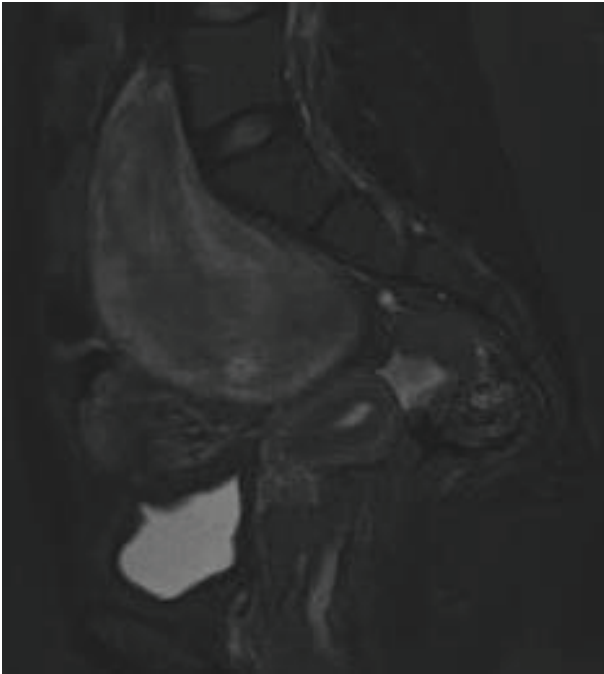
function test, and X ray chest were normal. Following day, she underwent surgical removal of a large well encapsulated retroperitoneal mass which was sent for histopathology.

Histopathology diagnosis was ganglioneuroma which was advised for immunohistochemistry which confirmed the diagnosis. She was discharged with healthy wound after the 3<sup>rd</sup> post-operative day.

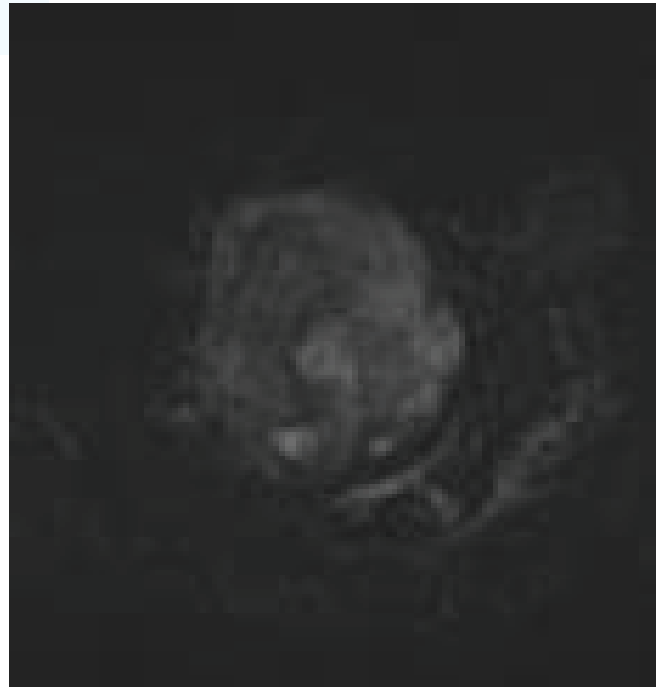
After 6 months, follow up MRI with contrast revealed no evidence of recurrence or residual tumor (Fig.4), however, retroverted uterus was present till then.



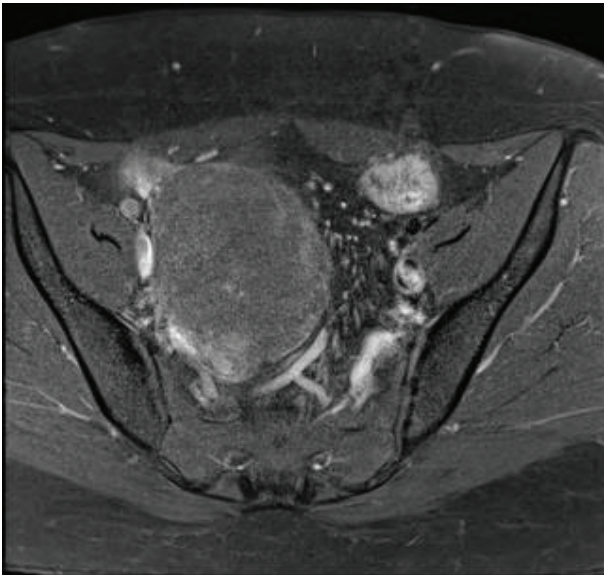
**Fig. 1: TVS: heterogeneously hyperechoic mass with scarce vascularity**



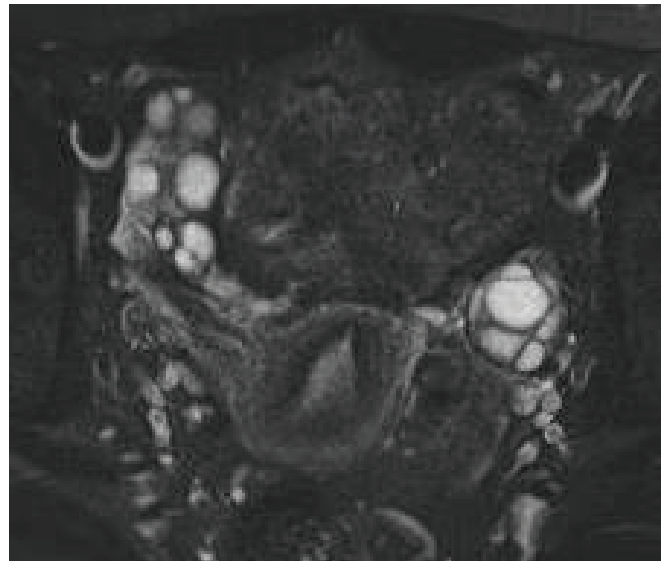
**Fig. 2a**



**Fig. 2b**

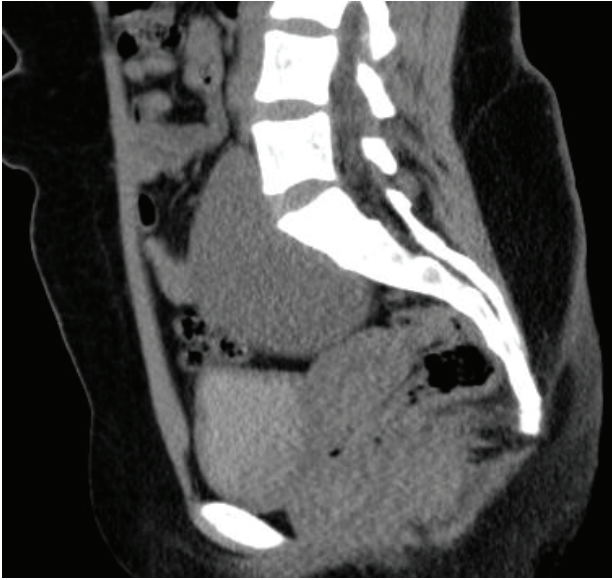


**Fig. 2c**

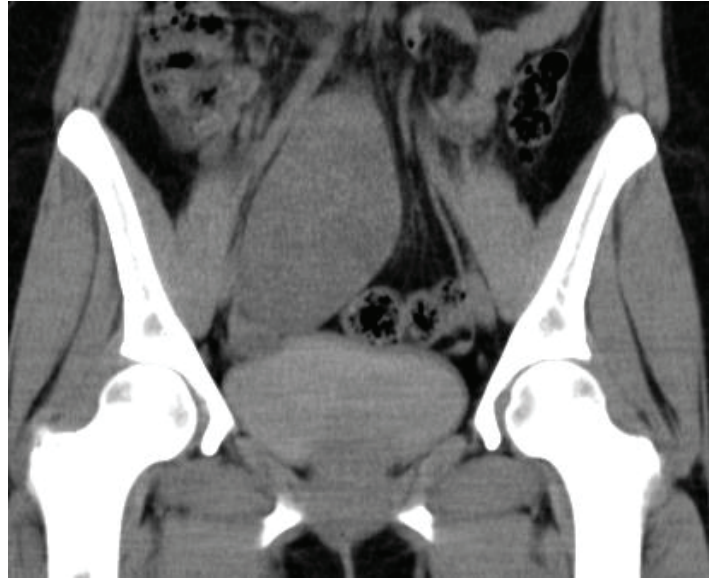


**Fig. 2d**

**Fig. 2** a. T2 sag fatsat: broad based hyperintense mass extending from L5 to S2 level, indenting and displacing the uterus posteriorly. b. DW image: no restriction of diffusion. c. T1 axial post gad fatsat: mild heterogeneous enhancement. d. T2 axial fatsat: polycystic appearing ovaries.

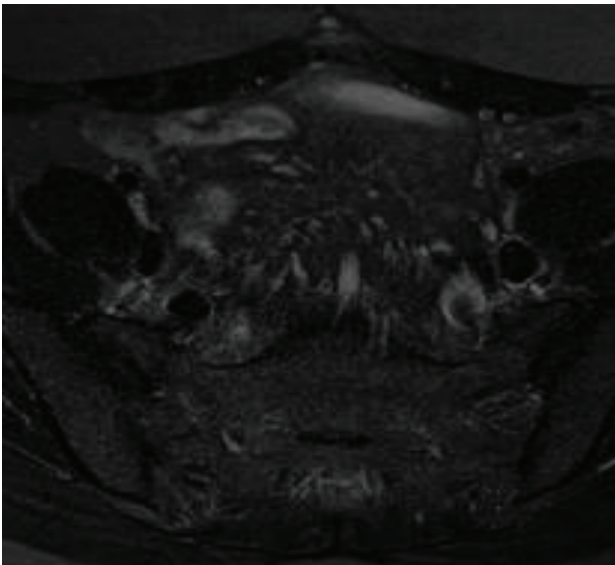


**Fig. 3a**

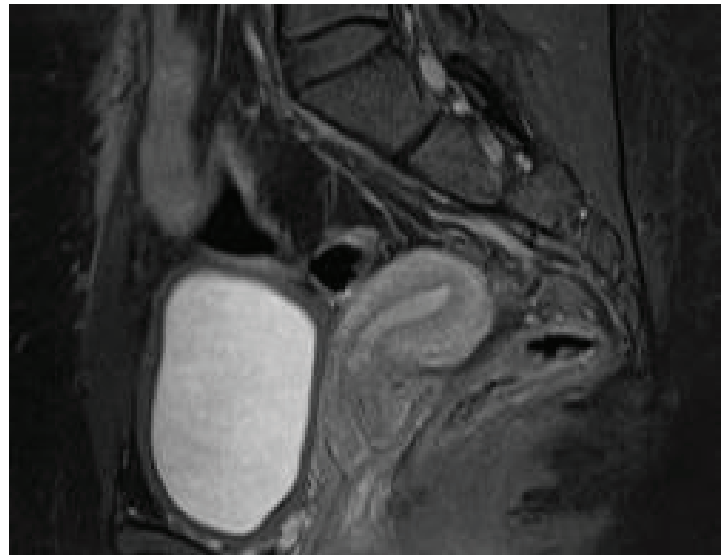


**Fig. 3b**

**Fig.3** CT scan of pelvis sag (a) and cor (b) images: no evidence of calcification within the mass



**Fig. 4a**



**Fig. 4b**

**Fig. 4.** Follow up MRI after 6 months, (a) T2 axial fatsat and (b) T1 sag post gad fatsat images show no evidence of recurrence or residual. However, retroverted uterus still persists.

## **DISCUSSION**

Neuroblastomas, ganglioneuroblastomas and ganglioneuromas are tumors of the sympathetic nervous system that arise from the neuroectodermal cells derived from the neural crest cells. Neuroblastoma tend to be aggressive and occur in younger patients (Average 2 years), whereas ganglioneuromas occur in older children (Average 7 years) <sup>1</sup>. They are mostly sporadic but there are a few reports of ganglioneuromas associated with neurofibromatosis type II and multiple endocrinologic neoplasia type II<sup>2</sup>. 56% of ganglioneuromas develop in the mediastinum or retroperitoneum, 30% in the adrenal gland and 14% from a variety of other unusual sites such as the mandible, pharynx, bladder, uterus, ovary and gastrointestinal tract<sup>3</sup>.

Most of the ganglioneuromas are found incidentally on abdominal imaging. The reported symptoms in pelvic ganglioneuromas especially in presacral location are pain, constipation or amenorrhea, though mostly clinically silent<sup>4</sup>. In our patient, pelvic ganglioneuroma was incidentally detected on abdominopelvic ultrasonography performed to confirm her pregnancy. When they have metabolic activity caused by catecholamines, vasoactive intestinal polypeptides, or androgenic hormones, hypertension, diarrhea, and virilization may be seen<sup>5,6</sup>. Compressive symptoms-related mass size may occur<sup>7</sup>.

Ultrasound is the initial diagnostic modality in screening and detecting retroperitoneal and pelvic ganglioneuromas. US reveals a homogenous, hypoechoic mass. Ganglioneuromas are reported as hypovascular tumours<sup>8</sup>; however, there is lack of information about the Doppler ultrasonographic features of these lesions in the literature. In our case, Doppler ultrasonography displayed sparse vascularity. On unenhanced CT, ganglioneuromas are of low attenuation and homogenous mass. Fine and speckled calcification may be seen in about 20% of cases<sup>9</sup>. They tend to surround or displace major blood vessels, but without compression or occlusion. Because of the myxoid matrices in the tumor, delayed heterogenous enhancement occur post contrast. The MR imaging characteristics of ganglioneuromas reported previously were hypointensity on T1WI and hyperintensity on T2WI. Heterogeneous signal intensity depends on the

combination of myxoid material and ganglion cells. As at CT, ganglioneuromas usually demonstrate delayed enhancement<sup>9,10</sup>.

The differential diagnosis list of retroperitoneal ganglioneuromas with pelvic extension include neuroblastoma, ganglioneuroblastoma, schwannoma, meningioma, or other cystic lesions<sup>4</sup>. FNA can be used preoperatively, but it usually leads to inconclusive diagnosis and hence was discouraged in our case opting for open biopsy.

Surgical resection represent the choice for treatment and no need for adjuvant systemic chemotherapy or local radiotherapy. All pelvic ganglioneuromas above the third sacral vertebral body level should be excised by anterior abdominal approach. When ganglioneuromas are located below the middle of S3 vertebra body level, posterior sacral transection or combined anterior and posterior approach is the ideal treatment, unless there is concomitant pelvic viscera involvement<sup>11</sup>.

## **CONCLUSION**

In conclusion, the ganglioneuroma arises from the sympathetic ganglion. It is a very rare disease and difficult to distinguish from other tumors since specific findings are not sufficient to diagnose it, diagnosis is always made histologically. However, it must be included in the differential diagnosis of retroperitoneal masses.

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