

## CASE REPORT

### Incidentally detected Ganglioneuroma of adrenal gland

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#### ABSTRACT

A 44-year-old female presented with abdominal discomfort of two months duration. She had no other complaints. On clinical examination she was found to have fibroid uterus. Hysterectomy was planned. Pre-operative imaging studies incidentally picked up a large mass in the left adrenal. Areas of calcification were also noted in the adrenal mass. Excision of adrenal mass was done along with hysterectomy for fibroid. Histopathological examination of the adrenal mass revealed clusters of ganglion cells in a dominant schwannian stroma with areas of calcification. A diagnosis of ganglioneuroma of the adrenal was made. Specimen submitted as hilar lymph node also showed ganglioneuroma. Ganglioneuromas are benign tumors of the sympathetic nervous system that rarely arise in the adrenal gland.

**Key words:** ganglioneuroma, adrenal, incidentaloma, mass.

#### INTRODUCTION

Ganglioneuromas are benign tumors of the sympathetic nervous system that rarely arise in the adrenal gland. They are usually non-functional and detected incidentally following investigation for other clinical conditions. They are more commonly seen in female patients. Most common sites of origin are sympathetic ganglia, posterior mediastinum and retro peritoneum. Adrenal is a rare site. Ganglioneuromas are part of the histological spectrum of tumors comprising of neuroblastoma at the most undifferentiated end to ganglioneuroma at the differentiated end.<sup>1</sup> Ganglioneuroblastomas are group of tumors with intermediate differentiation.

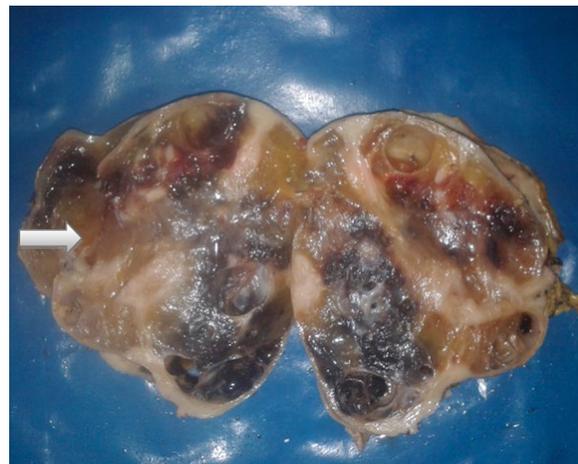
#### CASE REPORT

A 44-year-old lady presented to the local hospital with complaints of abdominal discomfort. The abdomen examination showed uterus enlarged to 12 weeks size. With a provisional diagnosis of uterine fibroid patient was worked up for elective surgery. A radiological evaluation revealed an adrenal mass in addition to uterine fibroid. With a radiological diagnosis of adrenocortical carcinoma, patient was referred to our center. Radiological evaluation done at our center confirmed left sided adrenal mass measuring 12x9x8cm with specks of calcification. The mass was abutting the spleen, pancreas and stomach but there was no infiltration into any of these organs. The endocrinology profile of the patient was within normal limits except for a reduction in cortisol level.

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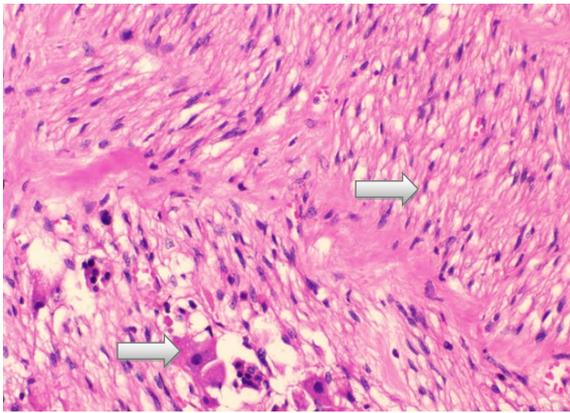
The fasting cortisol level was 0.8 microgm/dl as compared to normal range of 6.2-19.4 microgm/dl. However the patient had no symptoms related to endocrine system other than the abdominal discomfort. With a pre-operative diagnosis of an adrenal tumor patient was taken up for surgery. Excision of adrenal tumor, hysterectomy and excision of enlarged hilar lymph node was done. Specimen of adrenal tumor consisted of a large grey white myxoid mass with few cystic spaces and tiny specks of calcification. Adrenal tissue was identified at the periphery (Fig. 1).



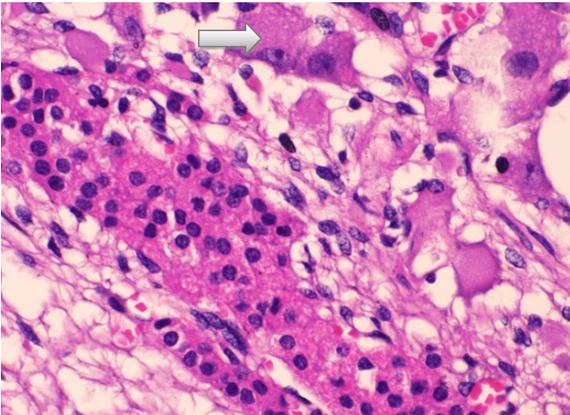
**Figure 1**, Gross specimen of adrenal tumor showing grey-white myxoid appearance.

The mass measured 10.5 x 8.5 x 3cm. The cut section of the specimen submitted as hilar lymph node also had similar appearance.

Histopathological examination of the adrenal tumor revealed a neoplasm comprising of fascicles of spindle cells resembling Schwann cells and clusters of ganglion cells (Fig. 2a, 2b).



**Figure 2a**, Section from neoplasm showing fascicles of spindle cells resembling Schwann cells and clusters of ganglion cells.(H&E x200).



**Figure 2b**, Section showing adrenal with neoplasm composed mainly of ganglion cells (H&E x400).

Focal calcification was also noted. Multiple sections studied failed to pick up any focus of neuroblastoma. Section from the mass submitted as hilar lymph node also showed histopathological features similar to adrenal mass. A diagnosis of ganglioneuroma was made. Nodal architecture could not be made out in the specimen submitted as hilar node since the entire mass was replaced by neoplasm.

Post operative period of our patient was uneventful. She is now on follow-up and is doing well.

## DISCUSSION

Ganglioneuromas are benign tumors of sympathetic nervous system arising from primordial neural crest cells.<sup>1</sup> Ganglioneuromas are at the benign end of the spectrum comprising of neuroblastoma and ganglioneuroblastoma and is characterized by the presence of mature ganglion cells. Patients with primary ganglioneuroma are significantly older than patients with neuroblastoma. Sometimes, following chemotherapy neuroblastoma can show histological maturation and differentiation into ganglioneuroma. Rarely a spontaneous regression can also occur. Ganglioneuromas thus should be examined thoroughly to exclude any foci of neuroblastoma. The adrenal gland is the most common location for a neuroblastoma, but the least common location in the sympathetic nervous

system for a ganglioneuroma.<sup>2</sup> Stowens demonstrated that 56% of ganglioneuromas develop in the mediastinum or retro peritoneum, 30% in the adrenal gland, and 14% in other sites throughout the body where sympathetic tissue is found such as the bladder and gastrointestinal tract.<sup>3</sup>

Ganglioneuroma may present as a functioning tumor. But usually they are non-functional and are noticed incidentally during investigation for unrelated conditions. In functioning tumors patient can present with weight loss, fatigue, flushing, palpitation, uncontrolled hypertension, diarrhea etc. Urinary levels of nor epinephrine, normetanephrine, vanillylmandelic acid and dopamine can be elevated. But histological confirmation is needed for accurate categorization of functioning or non-functioning tumors of the adrenal. Maweja et al suggested that a ganglioneuroma should be suspected radiologically in a non-functioning adrenal tumor, containing discrete calcifications, non-enhanced attenuation of less than 40 Hounsfield units on computed tomography (CT), a low T1-weighted signal, slightly high T2-weighted signal, and a late and gradual enhancement on dynamic magnetic resonance imaging (MRI).<sup>4</sup> Though ganglioneuromas are considered as benign they are known to produce deposits in lymph node, liver etc.<sup>5.</sup> Local recurrence and malignant transformation into malignant peripheral nerve sheath tumors have been reported.<sup>7</sup> However currently, histopathology examination is the only tool to diagnose ganglioneuroma and to differentiate it from other neural crest tumors.

## CONCLUSION

Currently, histopathology examination is the only tool to diagnose ganglioneuroma and to differentiate it from other neural crest tumors. Surgical excisions warrants complete cure in almost all the patients. Even after surgical resection it is advisable to keep the patient on regular follow-up with periodic radiological evaluation.

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