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CASE REPORT

ADRENAL GANGLIONEUROMA: RARE CAUSE OF GROSS HEMATURIA

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ABSTRACT

A Ganglioneuroma is the rarest and benign tumor of neural crest origin. It originates from the neural crest cells wherever sympathetic nervous tissue present, like in the retroperitonium, mediastinum, and adrenal medulla. They are common in children and young before the age of 20. Ganglioneuroma of adrenal gland is very rare. We are presenting rare case adrenal Ganglioneuroma in a five-year-old female child presented with large palpable left lion mass and intermittent hematuria. CT abdomen revealed, a 15x8cm-cm non-homogenous mass in the left adrenal gland compressing left kidney. Considering the size of tumor differential diagnosis of Neuroblastoma and other malignant adrenal tumor made. Patient underwent open lapromatomy for tumor removal. Histopathology examination revealed a well – encapsulated benign tumor of mature ganglion cells and Schwann- like cells arranged in fascicles.

INTRODUCTION

Ganglioneuromas are rare benign tumors arising from the neural crest cells. They arise from the sympathetic ganglion cells. The most common site is mediastinum, retro peritoneum and adrenal glands.[1] [2] Ganglioneuromas of the adrenal gland are extremely rare. These tumors affect preferentially young people with the majority reported to occur before the age of 20. These tumors are usually asymptomatic and are hormonally silent. When symptomatic, they present with non-specific symptoms related to their size or location with compression of neighboring structures.[3] Since adrenal Ganglioneuromas are extremely rare. They are found incidentally while, investigating unrelated pathology.[1] It is rare to diagnose them, presenting with lump in abdomen and gross hematuria. We are reporting first such case. Histopathological examination is the gold standard for final diagnosis and ruling out a malignant tumor.

Case Presentation

A 5-year-old female child presented to surgical OPD with complaints of mass in left lion region, and intermittent visible hematuria since two months. On physical examination, there were no noticeable findings except for palpable mass in her left lion region. She underwent routine lab work, which was unremarkable except RBC in urine. CT – Abdomen showed A 15x8cm-cm non-homogenous mass was revealed in the left adrenal gland and compressing left kidney. Endocrine work-up, including cortisol, Adrenocoticotropin hormone levels, 24 hours urinary catecholamines were within normal limit. Due to the tumor size and with consideration of the differential diagnosis of a malignant lesion made. Open abdominal surgery done for removal of tumor. While operating, tumor found to be compressing the completely left kidney and distorting the normal architecture of kidney. Complete removal of tumor along with kidney done, considering its malignant potential. The final histopathological report revealed adrenal mature Ganglioneuroma. Postoperative recovery was uneventful.

GROSS: We received a specimen of tumor of size 15x8cm along with kidney of size6x3x2cm. [Fig-1].

Fig 1 Received a specimen of tumor of size 15x8cm along with kidney of size6x3x2cm.

Microscopic Examination: shows well-circumscribed tumor comprised of bundles and fascicles of spindle cells with slender wavy spindled nuclei and scattered ganglion cells seen. (fig2A, 2B)

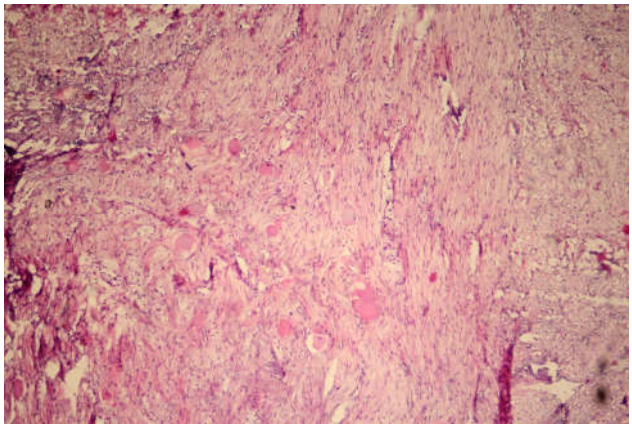

Fig.2a- Shows tumor comprised of bundles and fascicles of spindle cells with slender wavy spindled nuclei and scattered ganglion cells seen.(X10)

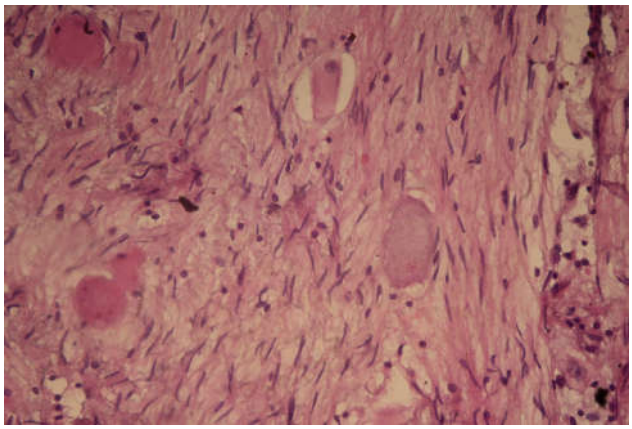

Fig. 2b- Shows tumor comprised of bundles and fascicles of spindle cells with slender wavy spindled nuclei.(X 40).

DISCUSSION

Ganglioneuromas are benign tumors originating from neural crest cells of sympathetic tissue and adrenal medulla. This tumor affects children and young people preferentially before age 20. Females are more prone than males. Ganglioneuroma is mature form of neuroblastic tumor. As Neuroblastoma differentiates, gives rise to Gangleoneuroblastoma, Ganglioneuroma. Neuroblastoma is aggressive with early recurrence and poor prognosis. On the contrary, Ganglioneuroma are benign and well-differentiated tumor. The most common location of Ganglioneuroma are posterior mediastinum and in the retroperitonium. Only in rarely occur in adrenal medulla. It is composed of ganglion cells, neuritis, Schwann cells, and fibrous tissue. They are discovered incidentally or due to non-specific symptoms caused by their mass effect on adjacent organs such as back pain and constipation.[3] [6] They are very notorious for their unusual clinical presentation. In our case patient presented with lump in left lion and intermittent gross hematuria. Clinical symptoms of GNs are non-specific, and related to their size/locations. In 2015 Jailim *et al*. reported the case of 18 year old male who presented with visible hematuria and right sided lion pain.[4] other cases reported are of microscopic hematuria by Leavittet and Fujita *et al*.[5] Ganglioneuroma are difficult to diagnose preoperatively. Imaging features of Ganglioneuroma are not discriminating. On USG, CT scan and MRI, they have different imaging characteristics.[1] [7] Modha *et al*. in their case series report, no patient was diagnosed preoperatively, in spite of imaging study in all cases and FNAC in three out of five cases.[8] The treatment for this condition is complete surgical resection through either an open or a laparoscopic approach. According to a 2004 National institutes of health State of the science statement-non - functioning adrenal masses, is to excise the lesion more than 6cm.[9] In our patient , mass was 15x8cm , and considering its unknown origin and young age of the patient, decided to go for surgery in order to resects the lesion and obtain histopathological diagnosis. After three months of surgery, there was no reoccurrence, and patient is in regular follow up.

CONCLUSION

Ganglioneuroma of the adrenal gland is an extremely rare entity and as such represents a diagnostic challenge. When young patient presents with mass in abdomen, and hematuria differential diagnosis of Ganglioneuroma should be considered. Surgical excision with histopathology confirmation of the tumor pathology remains the mainstay of management.

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