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Incidentally detected retroperitoneal ganglioneuroma after non-vehicle traffic accident: A case report of 6 year-old boy

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ABSTRACT

Ganglioneuroma is a rare benign tumor originates from neurol crest and may develop anywhere along the sympathetic nervous system is present. Nearly all patients with ganglioneuroma are older than 10 years old. In most cases ganlioneuromas are detected incidentally during work-up for unrelated conditions. Nevertheless, radiological and differential diagnosis of retroperitoneal masses may be challenging. We present management of a 6-year-old boy with ganglioneuroma that located in left retroperitoneum.

Keywords: Ganglioneuroma; incidental; pediatric; neuroal crest; retroperitoneum.

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Introduction

Ganglineuroma (GN) is a rare benign tumor originates from neural crest cells that forms part of neuroblastic tumors such as neuroblastoma and ganglioneuroblastoma. In contrast to other malignant forms, which consist of immature ganglion cells, GN includes ganglion cells, mature Schwann cells and nerve fibers [2].

It is reported that prevalence of GN in general population is one per million. Although GN occurs in all age groups, it is rare in subjects younger than 10 years, nearly all GN patients are older than 10 years old and approximately one in five of patients is over the age of 40 [3, 4].

GNs may develop anywhere along the sympathetic nervous system is present, including cervical spine, mediastinum, 2% of GNs are detected in the abdomen (49% arise in the adrenal gland and %51 extra-adrenal), 39% in the mediastinum, and the remaining 9% are detected in the pelvis or neck [5].

Despite the large size of tumor, retroperitoneal GNs are usually asymptomatic and hormonally silent. In most cases GNs are detected incidentally during work-up for unrelated

conditions [6]. Nevertheless, radiological and differential diagnosis of retroperitoneal masses may be challenging. We present a case of a large incidental tumor that located in retroperitoneum at the neighborhood of the upper pole of left kidney, in a 6-year-old boy.

Case report

In March 2015, a 7 cm-diameter mass adjacent to the upper pole of the left kidney was incidentally detected in a 6-year-old boy during examination in the emergency room for a non-vehicle traffic accident. The trauma was managed conservatively and the patient was referred to our instituon for the management of the mass.

On admission a retroperitoneal mass on the left side adjacent to upper pole of the kidney was recognized on computed tomography (CT), however due to insufficient image quality of CT, a contrast magnetic resonance imaging (MRI) of the abdomen was performed and a serum panel was ordered to determine whether the mass was endocrinologically active.

On MRI imaging, it was confirmed that the presence of retroperitoneal mass measuring 80 x 75 x 55 mm, adjacent to the upper pole of left kidney, lobular contour which pushed down the left kidney to inferior.

Mass signal was homogeneously low in T1-weighted (W) images, while T2-W images showed slight hyperintense signal. Heterogeneous contrast enhancement was detected in T1-W sequences after contrast media. Pathological lymph nodes were not visualized and the remaining abdominal structures had normal appearance [Fig. 1].

The urine and serum catecholamine and catecholamine metabolites (dopamine, vanilmandelic acid, homovalinic acid) levels were normal. Routine blood and urine analysis were also normal.

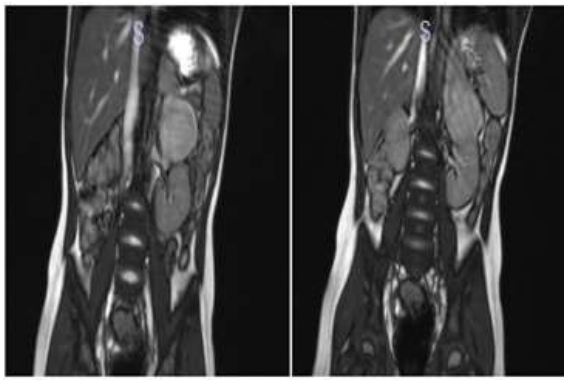

Fig. 1. MRI features of mass on T1-W images.

Excision with laparotomy was performed for removal of the tumor. During surgery, a mass covered by an intact capsule that adjacent but not attached to kidney and renal vascular structures. Left surrenal gland was also normal and very close to infero-medial part of the tumor. Retroperitoneal tumor was completely removed and left surrenal gland was preserved. Resected tumor was sent for histopathological examination [Fig. 2].

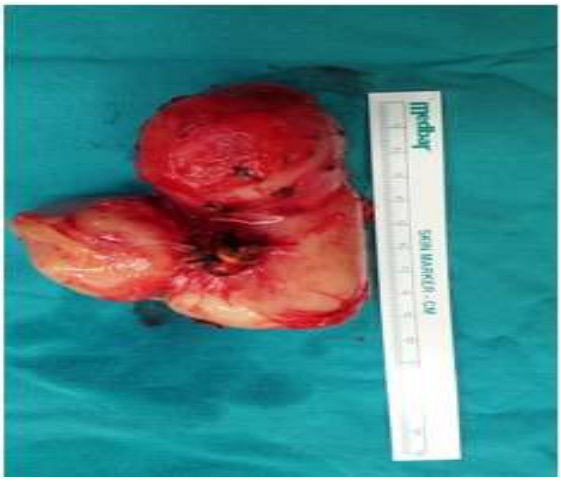

Fig 2. Postoperative macroscopic view of specimen.

Histopatological diagnosis of the tumor was ganglioneuroma that was composed of Schwann and mature ganglion cells.

The patient was followed for 9 months postoperatively. No recurrence was detected

during follow-up. However, the patient died on January 2016, due to fall from a height.

Discussion

The detection of an abdominal mass is a serious finding and is the most common presentation of a solid tumor in childhood. Wilms tumor and neuroblastoma are the most common abdominal masses in children older than 1 year. Physical examination findings, age, sex, other comorbidities, laboratory tests, and imaging methods are very important in the differential diagnosis [7,8].

Primary tumors of retroperitoneum which originate from mesodermal, neurogenic, germ cell ectodermal or lymphatic tissues are uncommon in children. Moreover, they have different spectrum and prevalence compared to those in adults.

In contrast to neuroblastoma, GN is benign and mature form of tumors that originate in neural crest. The most common location of GN is the posterior mediastinum (%40), retroperitoneum (%35), adrenal gland (%21), and neck (%8) [9]. Despite being detected in large sizes, retroperitoneal GNs are mostly asypmtomatic and found incidentally. Occasionally, GNs present with abdominal pain or the palpation of abdominal mass, and sometimes compression of the adjacent organs leads to diagnosis [1]. In the present case, a retroperitoneal mass was detected incidentally when assessed after a traffic accident at emergency department.

Radiological features of GNs on CT have been pointed out that it is well-circumscribed oval or lobulated mass along the sympathetic chains with regular borders, homogeneous content on unenhanced CT with slight to moderate delayed enhancement. GNs do not invade the adjecent vascular structures. There is no fatty or cystic component. Calcification can be detected in %20-50 of cases on CT, which is typically fine and speckled. On MRI, GNs show low signal intensity on T1-W images and high or isosignal intensity on T2-W images, depending on the proportion of stroma included in the mass [2,9]. In Figure 1; macroscopic appearance with lobulated contour of mass and T1-W MRI features has been shown.

GNs is known as asymptomatic and hormonally inactive in general. Hormone-secreting GNs, are extremely rare, have been mentioned as a few case report in literature in adults which located in adrenal gland [6]. In this case, metabolic tests were normal and hormonally inactive retroperitoneal mass has been detected.

Surgical approaches for retroperitoneal GNS have changed over the years. Open, laparoscopic or robot assisted laparoscopic modalities are being performed due to size and localization of the tumor. The complete removal of GNs provides for both effective diagnosis and treatment. There is no need for additional treatment such as chemotherapy and radiotherapy due to the nature of this benign disease [10, 11].

Conclusion

Despite being rare tumors, GNs should be considered as a differential diagnosis in the treatment of retroperitoneal tumors in childhood. Since radiological features and laboratory tests are insufficient, it may be difficult to distinguish from other lesions in retroperitoneal space.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Written informed consent was obtained from the parent of the patient.

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