Case Report

Ganglioneuroma mimicking Wilms’ tumor: Pediatric case report

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Ganglioneuroma is the tumor of sympathetic nerve fibers arising from neural crest cells. It may be present anywhere in the body along the autonomic nerve cells. This is usually a non-cancerous tumor. Its age incidence is between the 10-40 years of age. Usually tumor is asymptomatic; symptoms depend upon the location of tumor. In chest and pelvis, it may give symptoms by compression effect. Also, in the abdomen, mass may be palpable; this may be an incidental finding by radiologist. Radiological investigations may not differentiate this condition from other solid masses; misdiagnosis is common. Furthermore, the diagnosis is made on the basis of histopathology. Literature also shows that ganglioneuroma is misdiagnosed mostly.

Key words: Retroperitoneal mass, neural crest cells, autonomic nerve cells, incidentalomas.

INTRODUCTION

Ganglioneuroma is a rare entity which arises from sympathetic nerve fibers. The commonest sites are mediastinum, retroperitoneum and adrenal medulla but it may arise anywhere in the body usually along autonomic nerve cells (Erem et al., 2008). It is a benign tumor and malignant transformation is extremely rare. Age incidence is between 10-40 years but is more common in adolescents (Srinivasan et al., 2007). It is usually discovered during evaluation or treatment of another condition and the diagnosis is only histological (Oderda et al., 2011). The radiology mainly helps in its detection, as an incidentaloma (Carrión López et al., 2012).

Usually this condition is asymptomatic (Erem et al., 2009) and the symptoms depend upon its location. In chest it may cause breathing difficulty, chest pain or tracheal compression, and in retroperitoneal space, it can cause abdominal pain, mass or bloating. The pressure effects on spinal cord may range from pain to motor or sensory loss (Alimoğlu et al., 2012). Rarely, ganglioneuroma may produce certain chemicals or hormones and patients may be present with hypertension, increased body hair, sweating, diarrhea and females with enlarged clitoris. The most important differential diagnosis is with neuroblastoma in which urinary adrenaline, dopamine and VMA are raised whereas, ganglioneuroma is usually hormonally silent (Sucandy et al., 2011).

Misdiagnosis and maltreatment is not uncommon. The best diagnostic tool to identify these tumors is computed tomography or MRI of chest, abdomen and pelvis. Treatment involves surgical removal if symptomatic (Qing et al., 2010). The drastic complications include irreversible compression effect on spinal cord which may not resolve even after removal of tumor.

CASE REPORT

A 12 year old Pakistani boy was presented with history of palpable mass in left flank. The mass gradually increased in a size over a period of twelve weeks. Initially, the boy was seen and evaluated by a general practitioner. Abdominal ultrasound (USG) revealed heterogenous solid mass involving left kidney measuring 13.2x9.6 cm.
Figure 1. Axial postcontrast CT image shows the well-circumscribed mass in the retroperitonium on the left side. The mass is seen anterior to the abdominal aorta and inferior vena cava. Left kidney not visualized.

Figure 2. Per operative photo showing huge solid mass (Black arrow) adjacent to left kidney (Red Arrow).

There was no stone or hydronephrosis and impression was that of left renal mass with suspicion of Wilms’ tumor.

He was admitted at the Urology Department, Services Hospital Lahore, Pakistan which is a tertiary care hospital. Nothing was remarkable in general physical examination. However, the systemic examination of respiratory, cardiovascular and central nerve system was unremarkable. On abdominal examination, the mass was visible upon inspection with normal overlying skin and there were no prominent vessels. It was bimanually palpable, firm in consistency and partially mobile. Also, overlying skin temperature was normal, examining fingers could insinuate below costal margins since there was no bruit on auscultation. The investigations complete blood count, renal and liver functions were within normal limits.
Computed tomography (CT) abdomen was carried out and according to CT abdomen report, fairly well defined soft tissue density encapsulated minimally enhancing hypo-dense mass was seen arising from upper and mid pole of left kidney; left adrenal gland was not separately visible. Medially mass was projecting towards midline close to lateral margin of abdominal aorta, not encircling or infiltrating the vessels (Figure 1). Radiological diagnosis was in favour of the Wilms' tumor. In addition, Patient was referred to oncologist for neo-adjuvant chemotherapy but he demanded tissue diagnosis. Tru-cut biopsy was performed which was inconclusive. Explanation was made to the Parents and the boy was planned for exploration. The mass was approached through transperitoneal left Chevron incision. A solid 14x10 cm retroperitoneal mass separate from left kidney and suprarenal gland was identified and removed (Figures 2 and 3). The recovery was uneventful and patient was discharged in good condition. The biopsy report came out to be “Ganglioneuroma” (Figure 4a and b). The follow up USG at 3 months showed normal kidneys and there was no local recurrence.

**CONCLUSION**

Ganglioneuroma is usually a rare condition which can be easily confused with other pathologies. It provides a great diagnostic challenge. Computed tomography may not delineate the retroperitoneal mass from kidney. High index of suspicion is required to reach the proper diagnosis. Also, misdiagnosis can lead to misdirected...
management.

Consent

Written informed consent was obtained from patient’s parents for publication of this case report and any accompanying images.

REFERENCES


