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Case Report Open Access

Rare Case Report-Presacral Ganglioneuroma with Lymphadenopathy

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Abstract

Presacral Ganglioneuroma is an extremely rare tumor arising from sympathetic ganglion cells. As per our knowledge less than 20 cases have been reported previously in the world. We present a case reports on a Presacral Ganglioneuroma in 2 year old male child. He was admitted to our pediatric ward with complain of abdominal pain. Abdominal Ultrasound and computed tomography confirmed large pelvic mass with enlarged lymph nodes and extents of the lesion from the S1 level to the coccyx with intraspinal extension. We emphasize on radiologic appearance and pathological features of this rare entity.

Keywords: Ganglioneuroma; Presacral localization; Tumor

Introduction

Ganglioneuroma (GN) is benign slow growing tumor arising from sympathetic ganglion cells [1,2]. It is seen anywhere along the sympathetic nerve chain. At diagnosis, 60% patients are under the age of 20 years. The median age at diagnosis is 6.5 years. Only 5% of the cases are above 60 years [3]. The most common locations are the posterior mediastinum (41.5%), retroperitoneum (37.5%), adrenal gland (21%) and neck (8%) [3]. A presacral location of GN is extremely rare. As per our knowledge less than 20 cases have been reported previously in the world. Here, we present case report of presacral ganglioneuroma with lymphadenopathy.

Case report

A 2 year-old male child was admitted to our pediatric ward presenting with lower abdominal pain and pelvic mass. Neurologic examination was normal. Routine blood tests and serum tumor markers were within normal range. USG and CECT abdomen and pelvis was performed which show pelvic mass in presacral region. Then USG guided biopsy of this patient was performed and 0.5 × 0.6 × 0.4 cm^3 sized specimen was obtained which grossly shows few tiny grey white soft to firm tissue piece. Microscopically, section shows fragment of ganglion cells with neural tissue suggestive of ganglioneuroma.

USG abdomen shows large heterogenous echotexture solid lesion with echogenic calcific foci in presacral region which displaces rectum and urinary bladder anteriorly.

NCCT abdomen and pelvis shows 63 × 62 × 70 mm sized large lobulated hypodense lesion in presacral region with multiple scatter foci of calcification. On contrast enhanced study, Lesion shows heterogeneous enhancement in presacral region. Lesion extends into sacral canal from S3-4 to S4-5 neural foramina on right side with widening of neural foramina and spinal canal.

There are 16 × 11 × 41 mm sized soft tissue component is noted within spinal canal which causes erosion of posterior elements of sacrum. Lesion displaces rectum anteriorly and bladder anteriorly and superiorly. There are few enlarged nodes with foci of calcification surrounding lesion along b/l common iliac vessels, largest of which measures 11 × 17 mm in size (Figures 1-6).

Discussion

Ganglioneuroma is a benign tumor of neural crest origin that is very rarely found in the presacral region. There are less than 20 cases reported in the literature. The classification of neuroblastic tumors is based on the International Neuroblastoma Pathology Classification System [4]. According to this classification, neuroblastoma, ganglioneuroblastoma and ganglioneuroma are subdivided in seven categories. Neuroblastoma, ganglioneuroblastoma and ganglioneuroma are tumors arising from precursor cells of the neural crest that form the sympathetic nervous system and are called neuroblastic tumors [4].

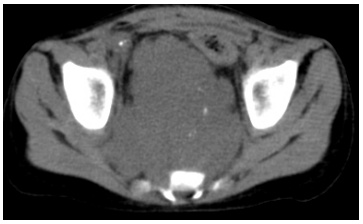

Figure 1: Axial NCCT images of pelvis shows large lobulated hypodensemass lesion with foci of calcification in presacral region.

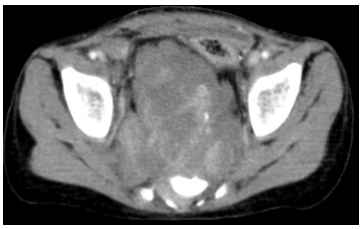

Figure 2: Axial CECT images of pelvis shows large lobulated heterogeneously enhancing soft tissue density lesion in presacral region which shows extensioninto sacral spinal canal through right sided neural foramina.

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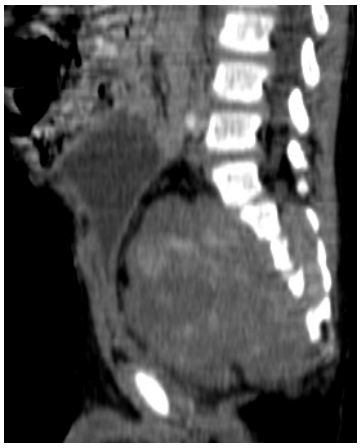

Figure 3: Sagittal CECT images of pelvis shows large solid masslesion extending into sacral spinal canal with widening of neural foramina and spinal canal.

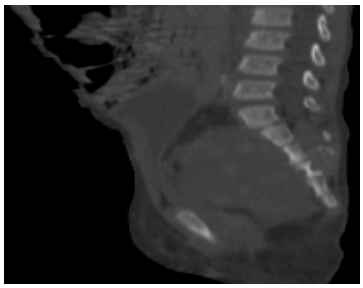

Figure 4: Sagittal Bone window CT images of pelvis showserosion of posterior elements of sacrum.

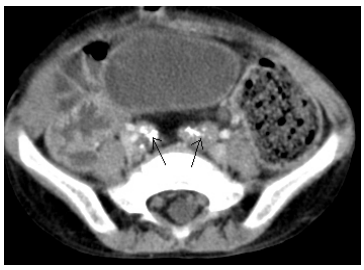

Figure 5: Axial CECT images of pelvis show few enlarged nodes with foci of calcification (black arrows) along b/l common iliac vessels.

Neuroblastoma is the poorly differentiated malignant lesion with bad prognosis, while ganglioneuroma is considered a benign tumor with excellent prognosis [5].

Ganglioneuromas are often asymptomatic, but a variety of nonspecific symptoms seen due to local mass effects on adjacent structures. For GN, median age at diagnosis is 6.5 years. For presacral location, median age at diagnosis is 35.5 years with a range from 8-70 years [3,6]. But in our case, age of patient is 2 year at which presacral ganglioneuroma is not reported yet. Usually, presacral GNs have a mean diameter of 7 cm [6].

MRI and CT are the preferred methods for imaging of ganglioneuromas. Calcifications are seen in approx. 42-60% of

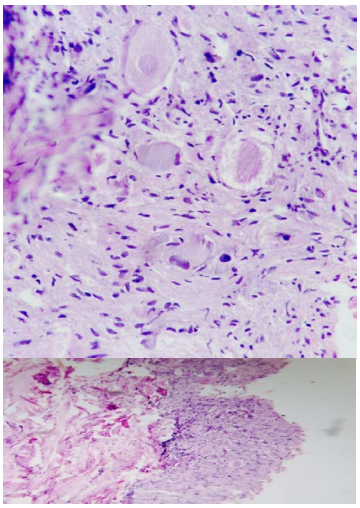

Figure 6: Section from presacral mass showing scattered mature ganglion cells surrounded by fascicles of Schwann like cells.

GN. Ganglioneuromas are similar to ganglioneuroblastoma and neuroblastoma on imaging, and therefore, it is not possible to differentiate these three tumors [7]. FNAC can be used preoperatively, but it usually does not give accurate diagnosis. Surgical resection is the optimal treatment of retro rectal tumors and provides a definitive histologic diagnosis [8].

Prognosis of GN is very good after surgery, even if it is subtotal and there are macroscopic residuals. Adjuvant chemotherapy or radiotherapy is not indicated due to the benign nature of the disease [3,6,9]. Malignant transformation of GN is found in rare cases, so follow-up needed yearly.

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