

CASE REPORT

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Ganglioneuroma in a child with chronic constipation and abdominal pain**Burcu Guven¹, Akkiz Sahin Yasar², Burhan Beger³, Veli Avci³**¹Yuzuncu Yil University Faculty of Medicine, Department of Pediatric Gastroenterology, Van, Turkey²Yuzuncu Yil University Faculty of Medicine, Department of Pediatric Hematology and Oncology, Van Turkey³Yuzuncu Yil University Faculty of Medicine, Department of Pediatric Surgery, Van, Turkey

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Abstract

Ganglioneuromas are rare, benign, slow-growing tumors arising from the neural crest. They are most commonly in posterior mediastinum, retroperitoneum, adrenal medulla and neck. They are very rare in presacral region. A 15 year old girl presented with abdominal pain and chronic constipation. A mass was seen between the uterus and sacrococcygeal bone in abdominal CT. It is resected totally and evaluated as ganglioneuroma. Conclusion: Presacral ganglioneuroma is a benign tumor which is rarely encountered. Patients might present with abdominal pain and constipation complaints as seen in our patient. Therefore, imaging methods should certainly be used for patients with chronic constipation which is resistance to treatment.

Keywords: Ganglioneuroma, pediatric, chronic constipation**Introduction**

Ganglioneuromas are rare, benign, slow-growing tumors arising from the neural crest. When tumors in the sympathetic nervous system are examined in a spectrum, neuroblastoma represents in the malignant end of the spectrum, while ganglioneuroma in the benign end [1]. Ganglioneuromas can arise anywhere along the sympathetic chain. They are most commonly in posterior mediastinum (41.5%), retroperitoneum (37.5%), adrenal medulla (21%) and neck (8%) [2]. They are very rare in presacral region.

Here, a 15-year-old girl presented with abdominal pain and chronic constipation. Most of these cases are functional. Ganglioneuroma is a rare cause of chronic constipation.

Case Report

A 15-year-old girl was admitted to the emergency room with a 2-day complaint of abdominal pain. She was constipated for about 6 months. Neither her own nor the family history of the patient presented any specific conditions. On physical examination, there was mild abdominal distension and tenderness. In adnexal area, abdominal ultrasonography showed a 82x97 mm sized cystic mass with intensive content.

In the abdominal CT, a 9.5x11.5 cm sized heterogeneous mass with cystic degenerative areas and hypodensities was observed between uterus and sacrococcygeal region (Figure 1-2). There was no significant relationship between mass and ovary. Bone destruction was not detected in the lesion. In the colonoscopy, colon lumen and mucosa were normal; no external signs of pressure were found. A 10x9x6 cm-sized, beige-colored, smooth-faced, and encapsulated mass was surgically resected. Microscopically, mature ganglion cells scattered in the neurofibromatoid stroma were observed and evaluated as mature ganglioneuroma (Figure 3).

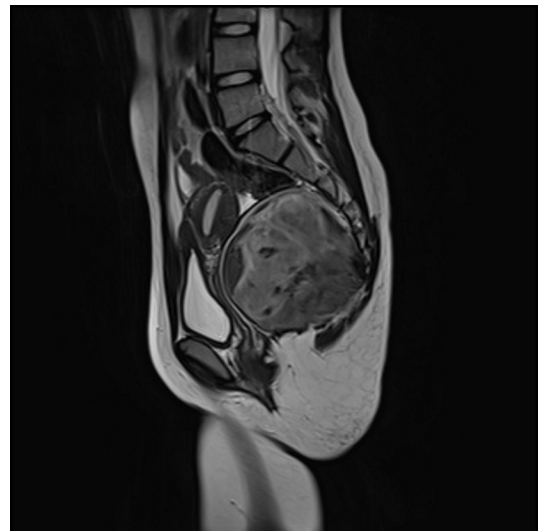


Figure 1. A heterogeneous hypointense mass between the uterus and sacrococcygeal region (Sagittal CT imaging)

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The patient who was on a four-month follow-up did not have any problem.



Figure 2. A heterogeneous hypointense mass between the uterus and sacrococcyx in posterior pelvic region (Horizontal CT imaging)

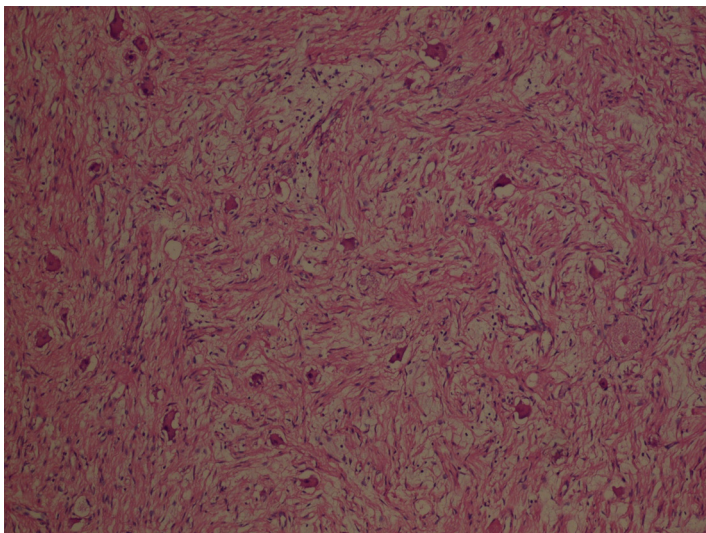


Figure 3. Mature ganglion cells with scattered localization in neurofibromatoid stroma (Hematoxylin and Eosin stain, original magnification x100)

Discussion

Peripheral neurogenic tumors are divided into three subgroups according to the degree, type, neurobehavioral differentiation, malignancy potential and development of schwann stroma, which are neuroblastoma, ganglioneuroblastoma, and ganglioneuroma. Among them, ganglioneuroma is a rare benign tumor, often originating from sympathetic ganglion cells or adrenal medulla cells. Histologically, the ganglioneuroma is composed of mature differentiation cells such as ganglion cells, schwann cells and nerve fibers [3]. It is not known whether pelvic ganglioneuromas are caused by neonatal neuroblastoma maturation or by a slow-growing ganglioneuroma [1].

Ganglioneuromes are usually seen in older children. Two-thirds of the cases are under 20 years of age. The mean diagnosis age is 7 years. In females, it is slightly more common than males (1.5: 1) [4]. They are usually slow-growing tumors. Therefore, they usually accompany with pressure symptoms or endocrinological disorders.

Ganglioneuromas are frequently localized in mediastinum, retroperineum and adrenal gland. Stout et al. reported 234 cases of ganglioneuroma [5]. Of these, 26% were located in the posterior mediastinum and 18% in the lumbar retroperitoneum [5]. Ganglioneuromas are rarely encountered in the presacral region. In previous studies, 19 cases of presacral ganglioneuroma were reported and only five of these patients were under 18 years of age. Six patients of them were diagnosed incidentally, while 10 patients had pain, 5 patients had constipation, and 1 patient had amenorrhea at the time of admission [6,7]. Our patient also presented with the complaint of abdominal pain and long-term constipation. Like our patient, a 70-year-old man presented with chronic constipation. Here, extrinsic compression in rectosigmoid segment was shown by barium enema study [8]. There was no barium enema study of our patient. In colonoscopy, it wasn't seen any extrinsic compression. If a barium enema study was done, partial obstruction could be detected. On the other hand, ganglioneuromas are usually asymptomatic. In recent years, the incidence of incidental diagnosis has also increased with the increase in imaging methods. In this case, This may have been detected incidentally.

Ganglioneuromas can be seen as space occupying lesions, rib springs or erosions in the bones, while they are seen as homogeneous hypoechoic masses in USG. While in computed tomography, they are seen as a homogenous mass with lower density compared to muscle. In T1-weighted MRI, they are seen in the form of a homogenous mass with lower density compared to liver, while in T2-weighted MRI, in the form of a heterogeneous mass with a higher density compared to liver. Discrete or punctuate calcifications are seen in 42-60% of the cases [9].

The treatment of ganglioneuromas is surgical excision. The rate of recurrence after total excision is low. However, if there is a residual mass, annual follow-up is appropriate [10]. Since it is benign, there is no need for systemic chemotherapy or radiotherapy [11].

Conclusion

In conclusion, presacral ganglioneuroma is a benign tumor which is rarely encountered. Most are asymptomatic. Rarely, patients might present with abdominal pain and constipation complaints as seen in our patient. Therefore, imaging methods should certainly be used for patients with chronic constipation, which is resistance to treatment.

Competing interests

The authors declare that they have no competing interest.

Financial Disclosure

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