Pelvic Ganglioneuroma – Case Report

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ABSTRACT

Ganglioneuromas are rare, benign, slow-growing tumors originating from sympathetic ganglion cells. The most common locations are the posterior mediastinum and retroperitoneum. Pelvic ganglioneuroma is very rare. The case of a 12-year-old girl with presacral ganglioneuroma is reported. The importance of considering a confident preoperative diagnosis by fine-needle aspiration biopsy is stressed.

Key words: ganglioneuroma, pelvic, child

Introduction

Ganglioneuroma is a rare neoplasm composed entirely of mature ganglion cells and Schwannian stroma. It arises anywhere along the sympathetic nerve chain. The most affected anatomic site is the posterior mediastinum, followed by retroperitoneum, adrenal gland, and neck. The clinical behaviour of ganglioneuroma is invariably benign, and complete surgical resection is considered to be curative

Herein, an extremely rare case of pelvic ganglioneuroma in a child is reported. Very few reports on the cytological appearance of this tumor exist. The utility of preoperative fine-needle aspiration in the diagnosis of ganglioneuroma, and its routine incorporation in the evaluation of pelvic masses in children is discussed.

Case Report

A 12-year-old girl presented with a complaint of lower abdominal pain for a foregoing week. Her past medical history and physical examination were unremarkable. Abdominal ultrasound (US) showed a large, solid, pseudo-capsulated, well circumscribed pelvic tumor with a few calcified foci, and the patient was hospitalized for further evaluation. The digital rectal examination disclosed a firm non-tender mass palpable just in front of the sacrum. Sedimentation rate, complete blood count, and blood chemistry tests were within normal limits, except for decreased levels of serum iron (40 mcg/dL) and ferritin (14.7 ng/mL). Serum tumor markers including neuron specific enolase, CEA, CA 19-9, CA 125, alpha-fetoprotein, and human chorionic gonadotropin were normal. Plasma and urine catecholamines as well as their metabolites homovanillic acid and vanillylmandelic acid were within the normal range. Pelvic computed tomography (CT) revealed a 9x8x7.5 cm solid, well defined mass in the presacral region with discrete speckled calcifications. Contrast-enhanced CT showed slight inhomogeneous enhancement. The uterus and rectum were compressed without invasion, and displaced ventrally and laterally. Magnetic resonance imaging (MRI) demonstrated a hypointense homogeneous tumor on T1-weighted images, and a hyperintense heterogeneous lesion on T2-weighted images. There was no extension into the sa-

Fig. 1. Large binuclear cell with prominent nucleoli and stromal fragment. May-Grunwald-Giemsa stain, x1000.
cell types is a ganglioneuroblastoma. In other words, a tumor composed primarily of neuroblasts is a ganglioneuroma, adjuvant chemotherapy or radiotherapy is not indicated. Local recurrence has been reported in about 5% of cases but in only 1% requiring a second surgical resection. The prognosis of ganglioneuroma is excellent. The relative frequency of spontaneously occurring tumors and those arising from maturing or treated malignant counterparts is unknown. There are very few reports of metastatic ganglioneuromas. These metastases are believed to be the end result of matured neuroblastomas or ganglioneuroblastomas, and these patients have an excellent prognosis.

Ganglioneuroma usually occurs in adolescents and young adults (40–60%), but individuals of all ages can be affected. The median age of occurrence is 7 years, with different studies reporting a median age of 5.5 years, 7 years, and over 10 years. There is a slight female predominance, ranging from 1.13:1 to 1.5:1. Ganglioneuroma can grow wherever sympathetic nervous tissue exists. The most common locations are the posterior mediastinum (41.5% of cases), retroperitoneum (37.5%), adrenal gland (21%), and neck (8%). Unusual sites include the spermatic cord, urinary bladder, intestine, abdominal wall, heart, bone, skin, and gallbladder. Ganglioneuromas are usually asymptomatic, and are typically discovered on a routine radiographic study such as chest radiograph. Sometimes they cause local mass effect, and patients may present with cough, dyspnea, abdominal pain, and palpation of an abdominal mass. These tumors may be hormonally active, thus manifesting with hypertension, flushing and other symptoms as a result of catecholamine secretion. It was believed that hormone production by ganglioneuroma is unusual, but there are several reports confirming that ganglioneuromas secrete catecholamines in as many as 37% of cases. Ganglioneuroma averages 8 cm in diameter and may appear encapsulated, although a true capsule is rare. Magnetic resonance imaging and computed tomography scanning are the preferred methods for imaging. Although ganglioneuroma tends to be more homogeneous than neuroblastoma or ganglioneuroblastoma, it is not possible to imaging evaluation to discriminate among these three tumors. Up to 60% of ganglioneuromas show calcifications; they are typically fine and speckled but may be coarse. Treatment consists of complete surgical excision when possible. Because of the benign nature of ganglioneuroma, adjuvant chemotherapy or radiotherapy is not indicated. Local recurrence has been re-
ported, so periodic radiological follow-up is indicated after resection. The present case describes an extremely rare presacral ganglioneuroma. A literature search has revealed only 18 case reports to date. The largest published series of presacral location includes five patients. On diagnostic evaluation of the reported patient, CT and MRI suggested neural tumor. Preoperative study was completed by fine needle aspiration cytology and histopathology. The tumor was completely resected, and postoperative histologic examination confirmed the diagnosis of ganglioneuroma.

Pediatric sacral tumors usually occur in the newborn period, with most of these tumors being sacrococcygeal teratomas. Other benign congenital tumors of the sacral region include lipomas, dermoids, and epidermoids. Ganglioneuroma, although very rare, should be considered in the differential diagnosis of nonteratomatous sacral tumors in pediatric age group. Because it shares clinical and radiological features with other benign and malignant neural tumors, accurate preoperative diagnosis is often difficult, but critical for proper management. This case illustrates the utility of preoperative fine-needle aspiration biopsy in the diagnosis of ganglioneuroma. Although technical approach may be hard, this is a rapid, simple, cost-effective, and safe diagnostic procedure. Adequate sampling, experience and caution in interpreting the specimen, as well as a close collaboration between pediatric oncologist, radiologist and cytopathologists are essential for its success.

REFERENCES


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PELVIČNI GANGLIONEUROM

SAŽETAK