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Case report

Pelvic Ganglioneuroma – Case Report

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A B S T R A C T

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 in invariably benign, and complete surgical resection is considered to be curative[1,2].

Herein, an extremely rare case of pelvic ganglioneuroma in a child is reported. Very few reports on the cytological appearance of this tumor exist[3]. 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-

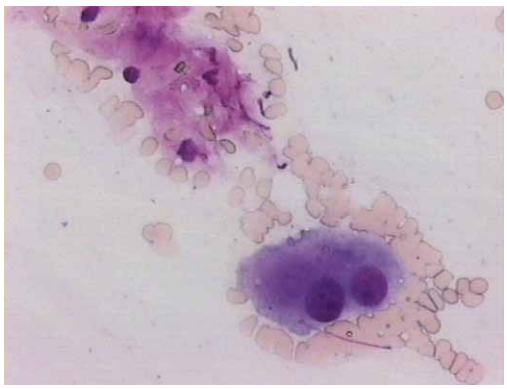

Fig. 1. Large binuclear cell with prominent nucleoli and stromal fragment. May-Gruenwald-Giemsa stain, x1000.

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cral foramina. These radiological findings were suggestive for the neural tumor.

US-guided fine needle aspiration was performed. The smears contained rare large cells with one to three nuclei and prominent nucleoli, and stromal fragments. These cells had features of ganglion cells (Figure 1). CT-guided biopsy was done. The histologic examination proved the lesion as ganglioneuroma composed of both mature ganglion cells and spindle Schwann cells (Figure 2). A complete surgical resection was performed using an anterior approach. The diagnosis of ganglioneuroma was subsequently confirmed on postoperative histopathologic examination. At a 3-year follow-up, the patient is asymptomatic and without evidence of disease.

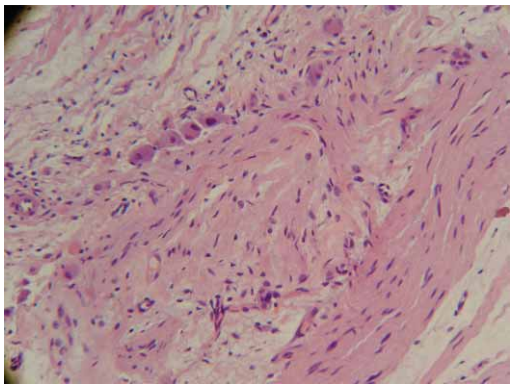

Fig. 2. Ganglioneuroma. Fascicles of spindle Schwann cells along with mature ganglion cells, some of them binucleated. Hemalaun-eosin stain, x200.

Discussion and Conclusion

Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma are tumors of the sympathetic nervous system that originate from primordial neural crest cells. These precursor cells may remain undifferentiated, referred to as neuroblasts, or may mature to ganglion and Schwann cells[4]. A tumor composed primarily of neuroblasts is a neuroblastoma, a tumor composed of mature ganglion cells and other mature tissue is referred to as ganglioneuroma, and a tumor with both immature and mature cell types is a ganglioneuroblastoma. In other words, these tumors collectively known as a group of neuroblastic or neurogenic tumors, define a spectrum of sympathetic tissue tumors, ranging from undifferentiated neuroblastoma to the mature ganglioneuroma. The presence of immature tissue in neuroblastoma and ganglioneuroblastoma indicates malignant or potentially malignant behavior; ganglioneuroma is considered benign tumor[2,4].

Neuroblastic tumors are classified according to the International Neuroblastoma Pathology Classification (INPC), established in 1999 by adopting the system of the original Shimada Classification and partly modified in 2003 (7 categories: neuroblastoma, Schwannian stroma-poor; ganglioneuroblastoma, intermixed, Schwannian stroma-rich; ganglioneuroma, Schwannian stroma-dominant; ganglioneuroblastoma, nodular; neuroblastic tumor, unclassifiable; neuroblastoma, not otherwise specified; ganglioneuroblastoma, not otherwise specified)[5]. INPC classification has been used as one of the most powerful front-end prognostic factors for patient stratification and protocol assignment in this disease[6].

Ganglioneuromas are rare, fully-differentiated neuroblastic tumors that contain mature ganglion cells, mature Schwann cells, fibrous tissue, and nerve fibers. These tumors have no immature elements, such as neuroblasts or intermediate cells, atypia, mitotic figures, or necrosis[2]. Ganglioneuromas may arise de novo, or result from the maturation of neuroblastomas and ganglioneuroblastomas. They can also develop from neuroblastomas that were treated with chemotherapy[2,6]. 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 neuroblastoma or ganglioneuroblastoma metastases, and these patients have an excellent prognosis[2,7].

Ganglioneuroma usually occurs in adolescents and young adults (40–60%), but individuals of all ages can be affected[2]. 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[7]. There is a slight female predominance, ranging from 1.13:1 to 1.5:1[7,8]. 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[2]. 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[7]. These tumors may be hormonally active, thus manifesting with hypertension, flushing and other symptoms as a result of catecholamine secretion[9]. 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[2,7]. Ganglioneuroma averages 8 cm in diameter and may appear encapsulated, although a true capsule is rare[4]. Magnetic resonance imaging and computed tomography scanning are the preferred methods for imaging[10]. Although ganglioneuroma tends to be more homogeneous tumor than neuroblastoma or ganglioneuroblastoma, it is not possible at 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[2,11]. Treatment consists of complete surgical excision when possible. Because of the benign nature of ganglioneuroma, adjuvant chemotherapy or radiotherapy is not indicated[1,2]. Local recurrence has been re-

ported, so periodic radiological follow-up is indicated after resection[12].

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[13]. 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[14]. 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

1. DE BERNARDI B, GAMBINI C, HAUPT R, GRANATA C, RIZZO A, CONTE M, TONINI GP, BIANCHI M, GIULIANO M, LUKSCH R, PRETE A, VISCARDI E, GARAVENTA A, SEMENTA AR, BRUZZI P, ANGELINI P, J Clin Oncol, 26 (2008) 1710. — 2. JEDYNAK AR, SCHWARZ RA, Ganglioneuroma and ganglioneuroblastoma, eMedicine Radiology, accessed 18.06.2009. Available from: http.//www.emedicine.medscape.com/article/340723. — 3. DOMANSKI HA, Diagn Cytopathol, 32 (2005) 363. — 4. JOSHI VV, Pediatr Dev Pathol, 3 (2000) 184. — 5. KOBAYASHI C, KANEKO M, SHIMADA H, J Pediatr Surg, 38 (2006) 621. — 6. OKAMATZU C, LONDON WB, NARANJO A, HOGARTY MD, GASTIER-FOSTER JM, LOOK AT, LA QUAGLIA M, MARIS JM, COHN SL, MATTHAY KK, SEEGER RC, SAJI T, SHIMADA H, Pediatr Blood Cancer, 53 (2009) 563. — 7. GEORGER B, HERO B, HARMS D, GREBE J, SCHEIDHAUER K, BERTHOLD F, Cancer, 91 (2001) 1905. — 8. HAYES FA, GREEN AA, RAO BN, Cancer, 63 (1989) 1211. — 9. LUCAS K, GULA MJ, KNISELY AS, VIRGI MA, WOLLMAN M, BLATT J, Med Pediatr Oncol, 22 (1994) 240. — 10. SCHERER A, NIEHUES T, ENGELBRECHT V, MOEDDER U, Pediatr Radiol, 31 (2001) 106. — 11. ICHIKAWA T, OHTOMO K, ARAKI T, FUJIMOTO H, NEMOTO K, NANBU A, ONOUE M, AOKI K, Br J Radiol, 69 (1996) 114. — 12. CERULLO G, MARRELLI D, RAMPONE B, MIRACCO C, CARUSO S, DI MARTINO M, MAZZEI MA, ROVIELLO F, World J Gastroenterol, 13 (2007) 2129. — 13. MODHA A, PATHY P, BILSKY MH, J Neurosurg Spine, 2 (2005) 366. — 14. LAM CH, NAGIB MG, Spine (Phila Pa 1976), 27 (2002) 284.

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PELVIČNI GANGLIONEUROM

SAŽETAK

Ganglioneuromi su rijetki, benigni, sporo rastući tumori porijeklom iz simpatičkih ganglijskih stanica. Najčešće su lokalizirani u stražnjem medijastinumu i retroperitoneumu. Pelvična lokalizacija je vrlo rijetka. U radu je prikazan slučaj 12-godišnje djevojčice s presakralnim ganglioneuromom. Naglašena je važnost pouzdane preoperativne dijagnoze aspiracijskom biopsijom tankom iglom.