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Olgu Sunumu

Primary neuroendocrine tumor of the retroperitoneum

Retroperitonun primer nöroendokrin tümörü

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ABSTRACT

Neuroendocrine tumors, often referred to as carcinoid tumors, are relatively rare in children and adolescents. Especially in the retroperitoneal neuroendocrine tumors are exceedingly rare neoplasms. In this paper, a 14-year-old boy with retroperitoneal neuroendocrine tumor who presented with vomiting is reported. A comprehensive review of the literature was also performed.

Key words: Neuroendocrine tumor, retroperitoneum, children

ÖZET

Sıklıkla karsinoid tümörler olarak adlandırılan nöroendokrin tümörler göreceli olarak çocuklar ve adolesanlarda nadirdir. Özellikle retroperitonda nöroendokrin tümörler oldukça nadirdir. Bu çalışmada, kusma ile başvuran 14 yaşında erkek çocukta retroperitoneal nöroendokrin tümör olgusu sunuldu. Bu olgu aracılığıyla kapsamlı literatür gözden geçirildi.

Anahtar kelimeler: Nöroendokrin tümör, retroperiton, çocuk

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BACKGROUND

Neuroendocrine tumors (NETs) are a heterogeneous group of rare neoplasms with variable histories and biological behaviors. The most common location of NETs is the gastrointestinal, followed by the respiratory tract, while other locations are being extremely rare [1,2]. Especially in children and adolescents, NETs are exceedingly rare retroperitoneal tumors, and there have only been a few case reports in the literature [3].

Despite the diversity in their tissue location, all NETs share common features, such as growth pattern and expression of neuroendocrine markers. The neoplastic cells grow in organoid nests, and trabeculae, and have round to oval nuclei with finely granular chromatin. Small glands with a rosette like appearance are only rarely present. Vascularity is pronounced. The stroma can be heavily sclerotic or hyalinized, and may exhibit focal calcification or ossification. Occasionally, prominent nuclear pleomorphism is seen in a NET in the absence of necrosis or mitosis. Prognosis has generally been regarded as favorable and complete surgical excision appears to be the adequate treatment [1-4].

We report a very rare case of NETs of retroperitoneum who presented with vomiting symptoms and radiological findings indicative of a benign mesenchymal or neuronal tumor. In addition, a comprehensive review of the literature was also performed.

CASE PRESENTATION

A 14-year-old boy with a 3-months-history of vomiting was referred for investigation of a retroperitoneal mass. Physical and laboratory evaluation of the patient was normal except from minimal LDH elevation (LDH 401 U\L, normal range: 110-220 U\L). Computerized tomography (CT) scan and US showed a 4x3 cm-sized solid homogeneous mass with smooth boundaries without contrast enhancement. The lesion was located in front of the pancreas and dissociated from pancreatic and liver tissue (Figure 1). The chest CT scan was normal. The mass was removed surgically. Surgical exploration of the mass showed no evidence of hepatic, pancreatic or other metastatic disease and pathologic lymph node. Gastroduodenoscopic and colonoscopic examination was done during the operation. An irregularly marginated ulcer was found on the anterior aspect of the duodenal bulb during esophago- gastroduodenoscopic examination. Colonoscopic examination was normal. One biopsy specimen from the fundus and two from the antrum were taken for histopathologic examination. Histologically, the patient showed pangastritis. Because of the lack of proliferation of secretary glands, and increase in enterochromaffin cells, Zollinger-Ellison syndrome was excluded.

Gross examination of the tumor showed a sharply demarcated solid grayish-tan color lesion that was 3.5x3.5 cm in size, with punctuate foci of hemorrhage. No areas of necrosis and hemorrhage were seen. Microscopically, the tumor was composed of an organoid nests and trabecular growth pattern. Tumor cells have monotonous, round and vesicular nuclei with finely granular and dusty chromatin and multiple nucleoli (Figure 2). Immunohistochemically, tumor cells showed reactivity for pankeratin, neuron-spesific enolase, synaptophysin, S-100 and CD 56 (Figure 3). The rate of Ki67 ranges between 2, and 5 percent. Histopathological examination and immunohistochemical findings of specimen were evaluated as an neuroendocrine tumor.

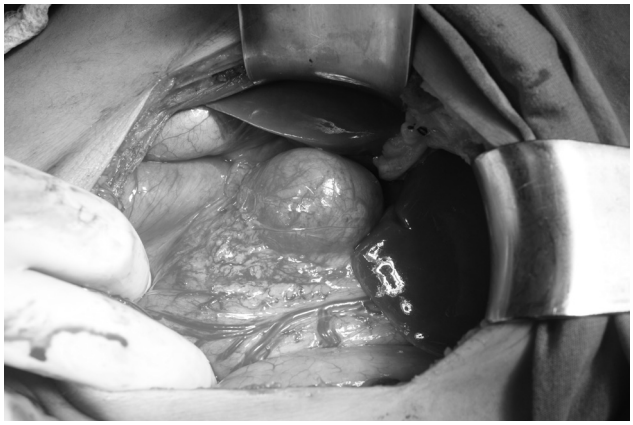

Figure 1. Tumor tissue 3.5 cm in diameter demonstrated in the retroperitoneum.

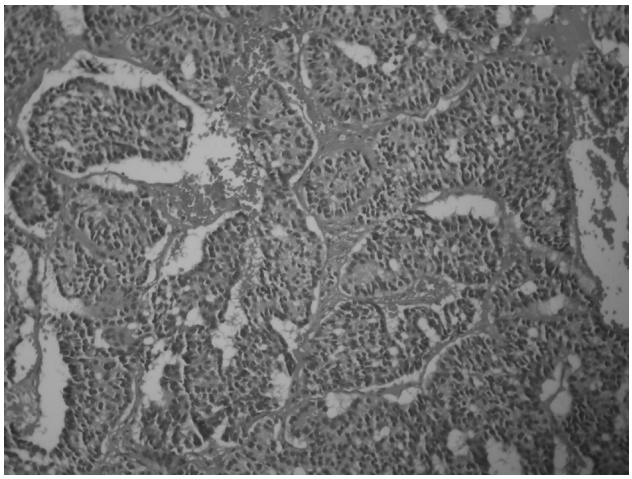

Figure 2. The tumor was composed of organoid nests (HEX200).

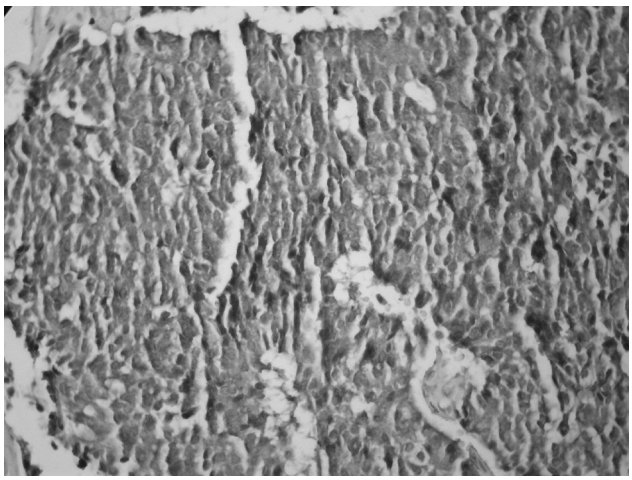

Figure 3. Tumor cells positively stained with Synaptophysin (DABX200).

After careful review of clinical, radiological (US, abdomen and thorax BT, gastroduodenoscopy, colonoscopy and MIBG scintigraphy) features, the appropriate diagnosis of primary retroperitoneal NET was rendered and no determined primary focus. So this patient had a non-secretuary tumor with occult clinical signs of secreted peptides or any specific characteristics. Patient was kept under long-term close follow-up instead of aggressive therapy.

DISCUSSION

NETs are a heterogeneous group of uncommon neoplasms with variable natural history and often indolent biological behavior. The term "neuroendocrine tumor" is better defined than the previously used "carcinoid" as it reflects the origin of these neoplasms in neuroendocrine cells [5]. NETs are rarely found in children and adults under the age of 30 years. The prevalence is 2.8 per million in adult populations which reflect significant increase over the past 15 years. Despite their low number, NETs represent the most frequent tumor of the gastrointestinal tract in children and the most frequently diagnosed primary pulmonary tumor in children and adolescents [1]. Nearly 70% of NETs develop in the gastrointestinal tract, with the appendix being the most common location, and another 25% occurring in the bronchopulmonary system. Other sites of occurrence include the thymus, gonads, breast, and other areas within the gastrointestinal tract [2,6]. Especially NETs are exceedingly rare retroperitoneal tumors. This case was presented for its rarity.

The World Health Organization has provided a recent classification of NETs, to include 5 major categories: well-differentiated endocrine tumors (benign or low-grade malignancy), well-differentiated endocrine carcinomas, poorly differentiated endocrine carcinomas (small cell carcinomas), and tumor-like lesions. This differentiation is based on the tumor's histology, tumor size, morphology, and presence or absence of local invasion or metastasis [7]. Our case was diagnosed by this classification as a well-differentiated endocrine tumor of the retroperitoneum. The prognosis of completely resected NETs appears to be very well.

NETs, neuroendocrine marker-positive (chromogranin, synaptophysin, CD56 etc.) neoplasms are considered to originate from neuroendocrine cells. Neuroendocrine cells are defined as cells that have the ability to produce neurotransmitters, neuromodulators, or neuropeptide hormones which lack axons and synapses. NETs are sporadic and nonhereditary tumours. Symptoms associated with NETs are related to the location, size, hormone overproduction, and extent of spread of the tumor [8]. Our case had a non-secretory tumor with clinical signs of secreted peptides like diarrhea, bronchospasm or flushing without any determined primary focus.

The retroperitoneum is one of the most complex regions of human anatomy as it contains a variety of organs and structures from different systems, in particular those belonging to the urinary and digestive tracts and the vascular systems. Because of this, retroperitoneal tumors represent a particular oncological pathology. No other human tumor is as deceiving and scarce in specific clinical symptoms as these tumors. Primary retroperitoneal tumors represent a variety of lesions, with different treatment and prognoses. Sarcomas were most common, followed by lymphomas, benign tumors, carcinomas and germ cell tumors but neuroendocrine tumors are exceedingly rare neoplasms in this location [9].

Of course, there are currently two explanations for what occurred in our patient. One of them is that this retroperitoneal mass could be a massively enlarged lymph node. But, after careful clinical, physiological and radiological examination, we could not find any primary focus. Therefore this mass was considered primary NETs of the retroperitoneum.

Although the prognosis for NETs of the retroperitoneum is excellent, these patients should be followed up routinely, in view of the potential for invasion, and metastasis. Certainly, follow up of patients

may provide insight into whether this mass was of primary or metastatic origin.

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