Local Surgical Resection of Gangliocytic Paraganglioma of the Duodenal Papilla

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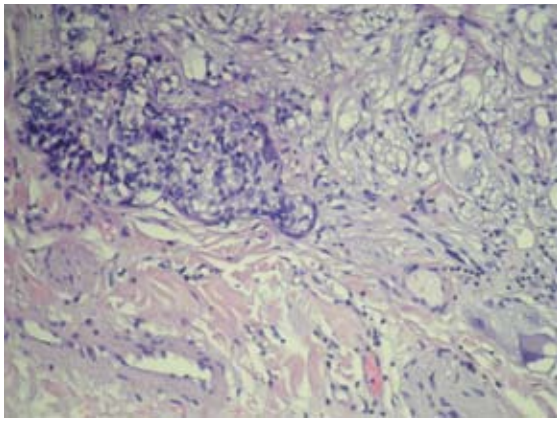
KEY WORDS: duodenum, endoscopy, gangliocytic paraganglioma, duodenal papilla

IMAJ 2009;11:311–312

Gangliocytic paragangliomas are rare gastrointestinal tumors that are found primarily in the second part of the duodenum [1]. Histologically, they are composed of three types of cells: epithelioid, ganglion and spindle cells. The more common presenting symptoms of these sessile or polypoid tumors are abdominal pain, gastrointestinal bleeding, and biliary obstruction; otherwise they are discovered incidentally [1]. These tumors are usually benign and can be cured by local excision. However, regional lymph node metastases [2] or recurrence after the incomplete excision has been reported. The endoscopic resection of periampullary gangliocytic paraganglioma has been described with minor and major papilla involvement. [3]. We describe a case of gangliocytic paraganglioma of the papilla without lymph node metastases that was asymptomatic with a history of positive occult stool blood test and mild anemia. Local resection was successfully performed, without evidence of recurrence.



The lesion is composed of three cell types: neuroendocrine cells, ganglion cells and schwannian stroma (hematoxylin & eosin, magnification x 20)

PATIENT DESCRIPTION

A 67 year old man who had suffered from psoriasis and pulmonary embolism 25 years previously presented with a 1 year history of positive occult stool blood test and mild anemia. A year prior to this he had undergone upper gastrointestinal endoscopy, which demonstrated prominent papilla. and a bleeding ulcer in the orifice of the papilla. During the preliminary consultation at the gastroenterology department he was asymptomatic. Biopsies were not taken due to fear of pancreatitis. A week later the patient underwent endoscopic ultrasound, demonstrating a very prominent ball-like papilla with normal overlying mucosa; common bile duct was seen without dilation, and no lymphadenopathy was shown. An additional upper gastrointestinal endoscopy was performed a week later and demonstrated a normal papilla with a small lesion of 15 mm lateral and proximal to the papilla. There was erosion on the surface of the lesion and biopsies were taken. Endoscopic biopsy suggested gangliocytic paraganglioma. The lesion did not obstruct the ampullary orifice.

Because of the location and limited endoscopic access to the lesion, endoscopic resection was not thought to be possible. The patient underwent local excision of the tumor from the duodenum. The postoperative course was uneventful and he was discharged on the fifth postoperative day.

HISTOPATHOLOGICAL ANALYSIS

Gross pathological evaluation of the resected specimen included a portion of duodenum without ampulla, measuring 1.5 cm in length [Figure]. The lesion measured 0.6 x 1 x 1.5 cm. It was circumscribed but encapsulated, and appeared to be covered by normal-appearing duodenal mucosa. Histologically, the tumor consisted of a complex neoplastic proliferation that included a component resembling carcinoid or islet cell tumor, admixed with a proliferation of spindled neurofibrillary cells and larger polygonal cells demonstrating gangliocytic differentiation [Figure]. The resection margins were free. No further therapy was given.

COMMENT

Gangliocytic paraganglioma is a rare neuroendocrine submucosal, typically benign tumor of the gastrointestinal tract most commonly located in the second portion of the duodenum [2,3]. A few cases involving the jejunum and pylorus have been described. Burke et al. [3] noted a slight male predominance with an average age at presentation of 54 years (range 15–80). Others have posited that there is no gender predominance [1]. The tumor usually presents with abdominal pain and gastrointestinal bleeding due to mucosal ulceration. Obstructive jaundice is less common.

Some patients are asymptomatic and tumors are found incidentally.

Endoscopy shows a sessile or polypoid submucosal lesion with normal overlying mucosa with or without an ulcer. Histologically, the tumor demonstrated the characteristic tricellular pattern of gangliocytic paragangliomas. These tumors are characteristically composed of a variable proportion of ganglion, spindle and epithelioid cells [3].

The histogenesis of this tumor is not clear. It has been suggested that the tumor arises from an embryonic celiac ganglion or from pluripotential intestinal cells, or that the lesion is due to hamartomatous proliferation of endodermal cells at the ventral primordium of pancreas, and neuroectodermally derived ganglion and Schwann cells.

These tumors are benign and submucosal and rarely recur or metastasize [4]. In most reported cases of regional lymph node involvement, the metastatic cell consists predominantly of epithelial cells [4]. In such cases the lesion is frequently larger than the tumor in the present case, ranging from 2 to 9 cm, and there is usually a local invasion or tumor emboli in the submucosal lymph nodes. Although gangliocytic paraganglioma are innocent and less frequent, they may be misdiagnosed clinically as adenocarcinoma and they should also be considered in the differential diagnosis.

The endoscopic resection attempt failed. A relatively unique element of this case is that we performed only local excision of the tumor without proceeding to pancreaticoduodenectomy or lymph node dissection. To the best of our knowledge, this is the first published case in Israel. Immunohistochemically these tumors stain positive for a variety of markers as was demonstrated in this report. Such markers include those mentioned above as well as neuron-specific enolase, pancreatic polypeptide, somatostatin, myelin basic protein and neurofilament proteins [3].

There are no data in the literature to guide clinicians on the use of adjuvant therapy despite the fact that approximately 5% of cases demonstrate malignant features [5]. Since the patient had no suspicion of lymph node involvement and is relatively old, we decided not to give any trial of adjuvant radiotherapy.

In conclusion, gangliocytic paraganglioma is a rare duodenal tumor that can present with non-specific symptoms. Positive diagnosis can be obtained histologically by observing three characteristic cell types. Although this tumor is considered benign, the regional lymph node involvement has been described. Due to the rarity of the disease, no clear adjuvant treatment strategy has been determined in cases that demonstrate regional or distant metastasis.

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