Rare Peripheral Nerve Schwannoma

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Abstract

Introduction & Objective: Schwannoma, a benign tumor of nerve sheath origin, is commonly found in the head and neck, as well as flexor surfaces of the extremities. It can rarely occur in the breast and stomach with only a few cases being reported.

Case Reports: The cases are: 1) a 57-year- old man with breast mass, 2) a 58 -year -old man with gastrointestinal complaints who were finally diagnosed as peripheral nerve schwannomas.

Conclusion: Clinical suspicion and histopathological confirmation are the keys to schwannomas diagnosis. Surgical excision is the treatment of choice for such patients.

Keywords: breast schwannoma, stomach schwannoma, diagnosis

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Introduction

Peripheral nerve tumors with low incidence and various features are mostly benign and involve various degrees of parent nerves [1]. Schwannoma is one of the few truly encapsulated peripheral nerve neoplasms, which was first described by Verocay. Benign schwannoma are commonly present as solitary lesions. Multiple lesions may be seen in association with neurofibromas in von Recklinghausen's disease, or as the syndrome of schwannomatosis [2].

The most common locations of Schwannomas are the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, and posterior spinal roots. Schwannoma rarely occur in breast and stomach and can arise from the parasympathetic or sympathetic division of the autonomic nervous system present in these organs [3]. The diagnosis and management of these rare types of Schwannomas are not completely introduced.

The purpose of this study was to regard schwannoma as a possible differential diagnosis of breast and gastric masses in suspicious patients and to introduce the diagnosis and treatment features of these rare tumors.

Case 1

A 57-year-old man was presented with a palpable mass in the right breast. The patient had found an ill defined mass in the breast 17 years ago but he did not have any diagnostic evaluation. The mass grew gradually in the past 3 years. He did not have any history of breast cancer in his family and never used anabolic agents. Physical examination revealed a painless, mobile, smooth, elastic-soft mass. Axillary lymphadenopathy was not observed and chest x ray was normal. Mammography showed a circumscribed and high dense "oval-shaped" tumor without microcalcifications or chest wall invasion with cystic parts. All other laboratory results were normal.

A well encapsulated mass was completely excised. The mass measured 2.5*3*4.5 cm and the cut surface was pinkish, devoid of degenerative changes and cystic areas.

Histological examination revealed a tumor entirely enclosed within a thick, fibrous capsule with a spongy biphasic pattern, corresponding to Antoni A and B areas. Spindle cells, often arranged in a palisading fashion or in an organoid arrangement (Verocay bodies), and a few large blood vessels with hyalinized walls were noted (figure 1). The tumor cells showed strong immunoreactivity for S-100 protein.

After the tumor excision, the patient was free of the disease and its symptoms recurrence for seven months.

Case 2

A 58 year-old man with the history of dyspepsia within last two years and melena as the current complaint was admitted to our hospital. During last the two months, his dyspepsia was exacerbated. He did not have vomiting but he mentioned early satiety and weight loss. His family history for malignancy was negative and he did not have any pervious surgeries or NSAIDS uses. In the physical examination, he was not pale and didn't have lymphadenopathy. We detected fullness in epigastric palpation. All other laboratory data were normal.

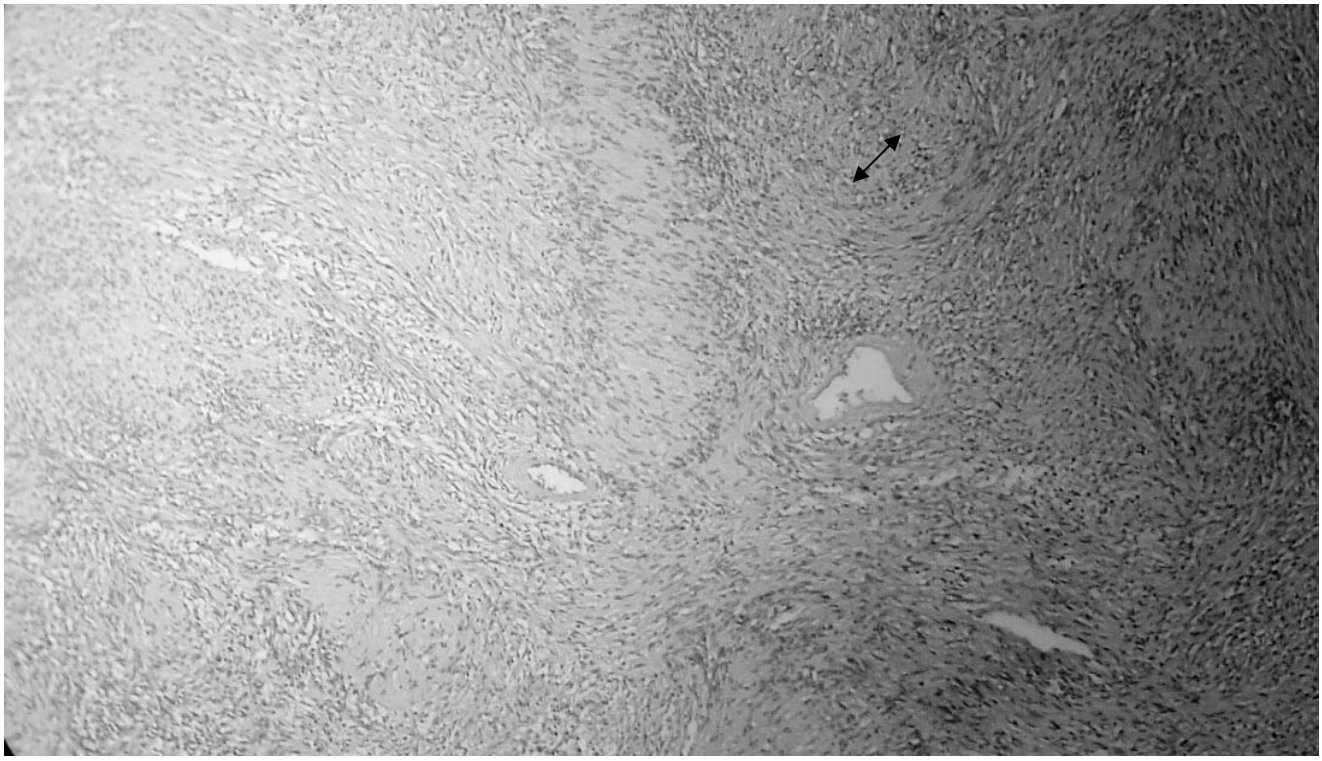

Fig 1: *Antoni A area with Verocay body and Antoni B. note large blood vessel with hyalinized walls (arrow) (hematoxylin and eosin X 400).*

In ultrasonography, a mobile 4 cm hypoecogenous mass was observed in the posterior wall of the stomach (figure 2A). The upper gastrointestinal endoscopic examination revealed a moderate inflammation in duodenum with 1*1 cm ulcer in antrum of stomach without any mucosal mass. Ureas test for helicobacter pylori was negative. The biopsy report only showed chronic inflammation without any malignant cells.

Upper gastrointestinal endosonography revealed hypoechoic mass lesion, 3.7*3.6 cm at the posterior wall and lesser curvature outside the stomach that seemed to be originated from muscularis properia. Further computed tomography scanning of the upper abdomen showed a 3.5*3.6 cm mass arising from the posterior wall of the stomach (figure 2B) that suggested tumoral lesion.

Intra-abdominal metastases were not observed. Carcinoembryonic antigen (CEA) was in normal range.

Through laparotomy approach, gastric wall tumor excised with 2 cm margin and gastric wall was repaired in two layers. A round tumor measuring 3.5*3.5 cm in size, located in the posterior wall of the body of the stomach, was observed (figure 2C). The tumor was limited to the gastric wall and didn't have any adhesion to other organs.

In pathologic finding, a 3.5*3.5 cm yellowish and lobulated mass revealed a picture of spindle cell arranged with Antoni A and B areas (figure 2D) compatible with schwannoma without any signs of increased mitoses, necroses and cellular atypia. The tumor was strongly positive for S-100 protein, and non-reactive for CD34, CD117 and desmin. The postoperative course was uneventful.

The patient was discharged 5 days after the operation. During the 6 months of the regular follow up, the patient did not have any signs of recurrence.

Discussion

Schwannomas are the most common peripheral nerve tumors. They mostly occur in the third decade of life. These benign tumors arise from Schwann cells. The most characteristic presentation is a mass lesion with point tenderness and shooting pains on direct palpation. The eccentric location and discrete encapsulated nature of these tumors often allow total resection without significant damage to the parent nerve [1].

Breast Schwannoma: in our patients breast Schwannoma was a slow growing mass without any pain. Mammography showed a circumscribed and high dense "oval-shaped" tumor without microcalcifications with cystic changes. This cystic

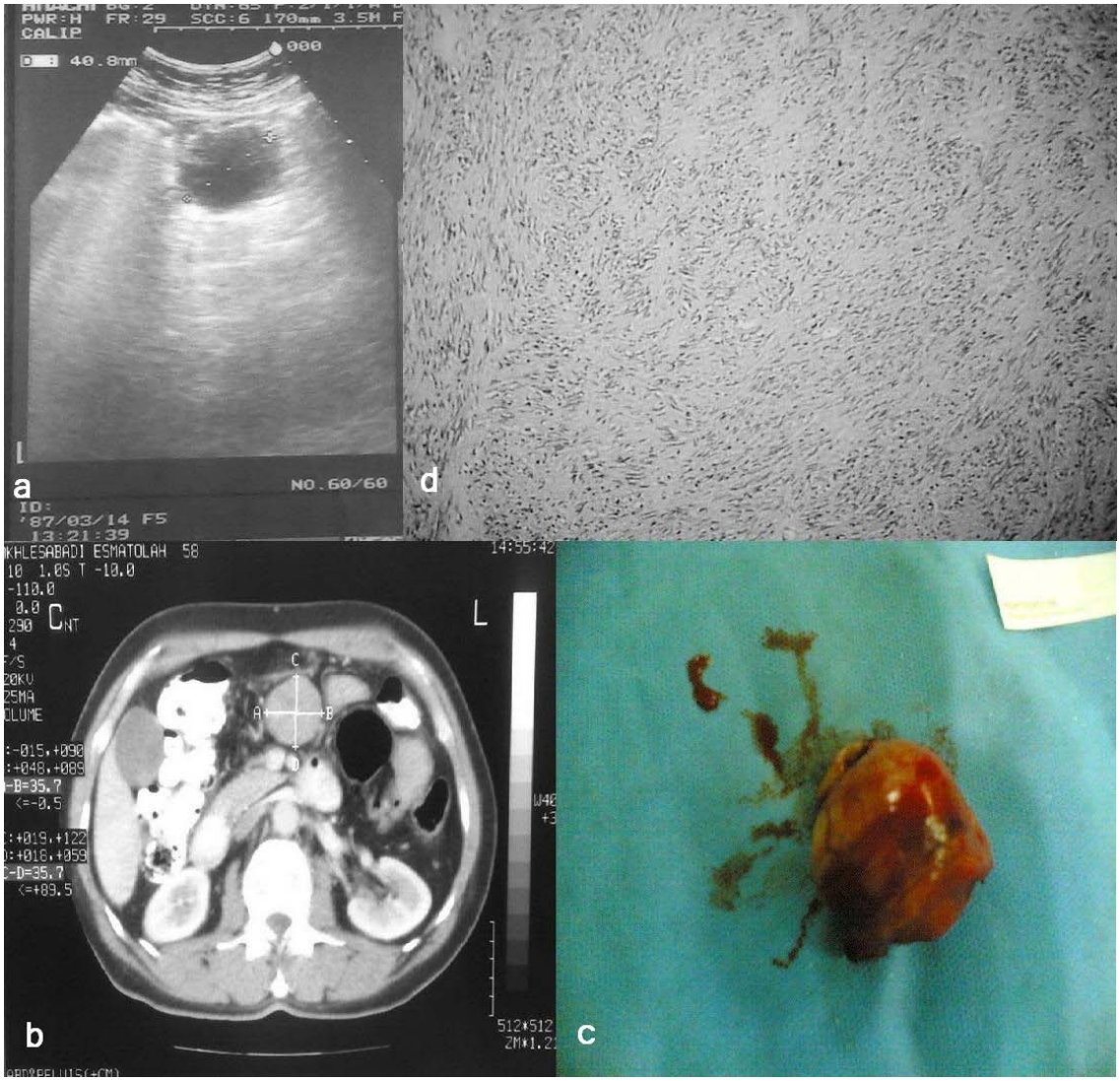

Fig 2: *A) ultrasonography show mobile 4 cm hypoecogenous mass in posterior wall of stomach. B) CT scanning of the upper abdomen shows the 3.5*3.6 cm mass arising from the posterior wall of the stomach body. C) The mass after excision. D) Spindle cell hinolaryngol Relat Spec.1995; 57(5): 273-8.*

change had not yet been reported and it might be due to the chronic period of the disease. Common findings of breast schwannoma in mammography are absence of a mass, round-oval and equally dense nodule or an ill-defined area of dense soft tissue that had been reported previously [4, 5]. These tumors show no worrisome mammographic or ultrasonographic features, and a preoperative diagnosis would be very difficult to make. In addition, we could excise tumors without any significant neurological deficits.

Cytology finding can help us to make differential diagnosis from other spindle cell lesion [6].

Gastric Schwannoma is a rare gastrointestinal mesenchymal tumor, which represents only 0.2 percent of all gastric tumors and 4 percent of all benign gastric neoplasms.This tumor arise from the nerve sheath of Auerbach plexus or less commonly Meissner plexus. They are slowly-growing encapsulated tumors composed of Schwann cells in a collagenous matrix. As the tumor enlarges, it displaces the nerve to the periphery of the tumor, preserving neural function [7].

Gastric schwannomas occur more frequently in the fifth to sixth decade of life and commonly in female patients [8]. Our patient was male and diagnosed with gastric mass in 5th decade.

Gastric schwannoma are often asymptomatic and can be discovered incidentally at laparotomy or radiographically. The most common presenting

symptom is an episode of upper gastrointestinal bleeding. Burneton case series revealed that most patients presented with abdominal pain and bleeding [9]. Hemorrhage is thought to be secondary to the submucosal mass and cause low blood flow to the gastric mucosa and gastric acidity [10]. In our case, the mass grew extraluminally and presented palpate mass at the time of admission. To our knowledge, this form of presentation has not yet been reported.

Computerized tomography can demonstrate the extent of invasion and help to determine the appearance of a benign versus malignant lesion [9]. Computed tomography scanning of the abdomen showed the 3.5*3.6 cm mass arising from the posterior wall of the stomach body without any invasion to the other organs.

Microscopically, the Schwannoma consists of two alternating components: an organized cellular component consisting of long bipolar cells that often form a pallasiding arrangement known as Verocay bodies (Antoni A area) and a loose hypocellular component (Antoni B area) [11]. In immunohistochemical staining, we can differentiate schwannoma from other mesenchymal tuomors. Positive desmin and muscle actin stains indicate leiomyoma or leiomyosarcoma ; positive CD34 and CD117 indicate GIST and positive S-100 indicates schwannoma. In our cases, the tumors revealed spindle cells with Antoni A and B areas and were strongly positive for S-100 stain, and were non-reactive for CD34 and CD117. These findings suggestive of Shwannoma.

Adequate treatment for solitary benign Schwannoma is surgical resection [7]. Total excision of these tumors requires sacrifice of the parent nerve. The choice of subtotal resection, nerve preservation, and observation, versus total resection with nerve sacrifice depends on tumor histology and the functions of the parent nerve [1]. For any benign gastric mass, we can perform wide local excision (2 cm margin). Benign gastric masses larger than 4 cm in diameter or invasion of the adjacent organs need subtotal or near-total gastrectomy. We detected 3.5*3.5 cm mass in our patient and then we excised the mass with a -2 -cm margin.

Prognosis for patients with solitary schwannoma following resection is excellent. Malignant transformation of a solitary lesion is rare.

Conclusion: we have presented two cases of rare schwannoma. Any Physician should be aware that these benign tumors might simulate a malignant neoplasm. A final diagnosis of Schwannoma was established by histopathological examination of the excised mass.

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