Case Report
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Plexiform Schwannoma of the Stomach in Neurofibromatosis Type 2: A Case Report[1]

제2형 신경섬유종증 환자에서 발생한 위의 얼기형신경초종: 1예 보고[1]

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Plexiform schwannoma is a relatively rare benign subepithelial tumor arising from the peripheral nerve sheath, and associated with Neurofibromatosis type 2 (NF2). There are a few reports of plexiform schwannomas arising from the gastrointestinal tract, and to our knowledge, there is no report of it arising from the stomach in a patient with NF2. Here we present the first case of a plexiform schwannoma of the stomach in an NF2 patient a submucosal tumor on radiologic finding.

Index terms
Plexiform Schwannoma
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INTRODUCTION

Plexiform schwannoma is a benign neoplasm of the peripheral nerves. It is generally localized in the skin and subcutaneous tissues of the head, neck, arms, and chest. Visceral localization of plexiform schwannoma is very uncommon (1). In particular, gastric involvement of plexiform schwannoma is extremely rare.

Neurofibromatosis type 2 (NF2) is a dominantly inherited tumor-prone disorder characterized by the development of multiple schwannoma and meningiomas (2). The clinical features of NF2 typically include nervous system tumors and ocular abnormalities (3).

We present a case in an 18-year-old man affected by NF2 with a gastric plexiform schwannoma, which was confirmed by endoscopic biopsy. To our knowledge, the case we present is the first report of a patient affected by NF2 with extremely rare case of gastric schwannoma.

CASE REPORT

An 18-year-old male presented with right lower quadrant abdominal pain. He was previously diagnosed bilateral vestibular schwannoma, left cerebellar extraaxial nodule, multiple intra- and extradural nodules of the spine and calcification of the left choroid plexus. He also had a mass on his eyelid that was confirmed to be a neuroma after excisional biopsy. NF2 because bilateral vestibular schwannomas on MRI or CT are the hallmark and definitely diagnostic for NF2 (3).

For evaluation of his abdominal pain, the patient underwent abdominal CT scan with 5-mm section thicknesses using a multidetector CT scanner. Abdominal CT revealed fluid-filled dilatation of the appendix with appendiceal wall thickening and periappendiceal infiltration, suggestive of acute appendicitis. Additionally, a 2.3 cm oval-shaped, hypoattenuated mass-like lesion abutting the stomach body was noted (Fig. 1A). The

attenuation of the mass was 25 HU on unenhanced CT (Fig. 1B), and 40 HU on contrast-enhanced CT. The margin of the mass was well-demarcated, and a thin peripheral wall connected with adjacent stomach mucosa was suspected (Fig. 1C). There was no calcification within the mass. Therefore, we initially diagnosed the patient with a submucosal tumor of the stomach.

Sequential endoscopy and endoscopic ultrasound (EUS) was performed for characterization of the mass. Endoscopy revealed a submucosal tumor at the lesser curvature of the gastric lower body (Fig. 1D). EUS was also able to show a well-marginated hypoechoic mass with hyperechoic strands arising from the third layer of the gastric wall, which suggested the possibility of a neurogenic tumor (Fig. 1E).

For final diagnosis, endoscopic biopsy was performed. As the histopathologic aspect of the mass, the spindle cells were relatively bland looking with pointed ends and palisading arrangement (Verrocay bodies) (Fig. 1F). Low power view showed several fragments of a multinodular growing neoplasm, separated by fibrous capsule (Fig. 1G). These findings were consistent with

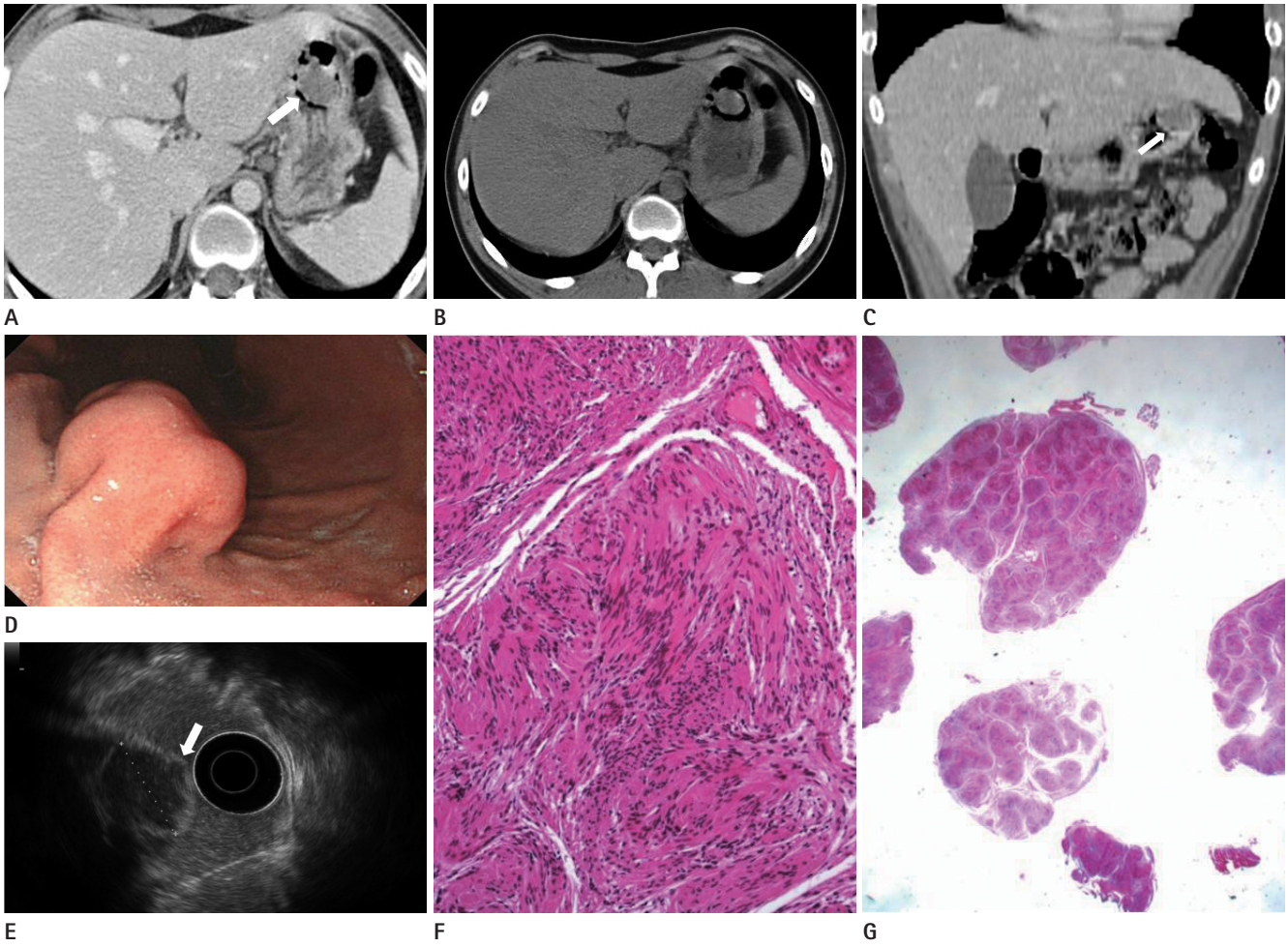

Fig. 1. Plexiform schwannoma of the stomach associated with NF2 in an 18-year-old male.
A, B. Axial MDCT scan shows a well-defined, ovoid-shaped, homogeneous soft tissue density lesion surrounded by mucosa in the gastric body (arrow) **(A)**. The attenuation of the mass is 25 HU on unenhanced CT **(B)**, and 40 HU on contrast-enhanced CT.
C. Contrast-enhanced coronal reformatted CT scan definitively shows that overlying mucosa is connected with adjacent stomach mucosa, indicative of a submucosal tumor (arrow).
D. Endoscopy shows a submucosal tumor at the lesser curvature of the gastric lower body.
E. EUS shows a well-marginated hypoechoic mass with hyperechoic strands arising from the third layer of gastric wall (arrow).
F. Photomicrograph (H&E staining × 200 magnification) shows that the spindle cells are relatively bland looking with pointed ends and palisading arrangement (Verrocay bodies), compatible with schwannoma.
G. Photomicrograph (H&E staining × 10 magnification) shows several fragments of multinodular growing neoplasm separated by fibrous capsule.
Note.—EUS = endoscopic ultrasound, MDCT = multidetector computed tomography, NF2 = neurofibromatosis type 2

a plexiform schwannoma pathologically.

DISCUSSION

Plexiform schwannoma is a widely documented variant of schwannoma (1) and a benign peripheral nerve sheath tumor composed exclusively of schwann cells arranged in a plexiform pattern (4). These lesions tend to present in early adulthood, lacking obvious sex predilection (5). Almost all plexiform schwannomas have been reported as dermal or subcutaneous tumors, and most commonly appeared as multinodular, well-circumscribed tumors (3-5).

NF2 has an autosomal dominant pathology. It is a separate entity from neurofibromatosis type 1 (NF1), and its clinical characteristics include 1) schwannoma of acoustical nerves, 2) central nervous system tumors (meningioma, astrocytoma, ependymoma), 3) juvenile subcapsular cataract, and sometimes 4) café au lait spots (recurring less frequently than in NF1) (1).

Several reports demonstrated an association between plexiform schwannoma and neurofibromatosis type (4). Solitary plexiform schwannomas are generally considered to be unassociated with NF1 (5), and rare associations with NF2 have been described (6). According to previous reports, patients with plexiform schwannoma associated with NF2 had acoustic neurinomas, but the association between these two diseases is still controversial, including the hereditary conditions (1).

Most plexiform schwannoma cases associated with NF2 showed multiple, dermal, and subcutaneous locations (6). Visceral location of plexiform schwannoma is extremely rare. The first case of a solitary plexiform schwannoma in the visceral organ was described in 1997 by Hirose et al. (7). Agaram et al. (6) assert that plexiform schwannoma with visceral localization occur more often in females than in males and have a high risk of recurrence but do not show malignant features or metastatic spreading. In our case, the patient was male and there was no evidence of metastatic spreading, but we could not know about recurrence and malignant transformation because follow up was not done. More generally, it is not yet clear whether plexiform schwannoma in unusual localizations have the same behavior as skin and subcutaneous lesions (1).

According to our literature search, only eight cases were revealed as plexiform schwannoma of the gastrointestinal (GI) tract (1, 4-10). The location of the lesions were three in the esophagus, one in the small bowel, one in the ascending colon, two in the sigmoid colon, and one in the rectosigmoid colon, respectively. Among these cases, one case in the esophagus of a pediatric patient and one case in the sigmoid colon of a second patient were associated with NF2. Therefore, to our knowledge, this is the first report that demonstrated stomach localization of a plexiform schwannoma associated with NF2.

In the GI tract, the differential diagnosis of plexiform schwannoma includes gastrointestinal stromal tumor with plexiform growth pattern. In this situation, a negative immunohistochemical stain for CD 117, along with strong and diffuse staining for S-100 protein, would be helpful in confirming the diagnosis for a plexiform schwannoma (6). Also, neurogenic tumors of the GI tract that must be distinguished from plexiform schwannoma include conventional schwannoma, neurofibroma, malignant peripheral nerve sheath tumor, ganglioneuroma, and ganglioneuromatosis (7).

The distinction among plexiform schwannoma, plexiform neurofibroma, and malignant peripheral nerve sheath tumor (MPNST) is crucial for the correct clinical management of the patient (1). While plexiform schwannoma is just a benign form, plexiform neurofibroma presents a 2% to 5% risk of malignant transformation, and MPNST is, by definition, a malignant neoplasm (1). The strong connection between NF1 and plexiform neurofibroma, and between NF2 and plexiform schwannoma is established in literature (1). This notion could be helpful in diagnosing the nature of a tumor.

The diagnosis of GI tract schwannoma is difficult preoperatively, as these lesions appear as subepithelial tumors. To our knowledge, there are no typical endosonographic features of GI tract schwannoma (9). Because of their plexiform pattern of growth, it is relatively common for benign schwannomas to traverse several layers of bowel wall and even extend into the surrounding adipose tissue (9). Definitive treatment requires complete surgical resection.

In summary, we have reported with a case of plexiform schwannoma of the stomach in an NF2 male patient. If a subepithelial mass of the stomach or GI tract is noted in a patient with NF2, although it is rare, the possibility of plexiform schwannoma should be considered.

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제2형 신경섬유종증 환자에서 발생한 위의 얼기형신경초종: 1예 보고[1]

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얼기형신경초종은 말초신경초에서 기원하는 드문 양성 종양으로 대개 상피하종괴로 발현되며 제2형 신경섬유종증과 연관성이 있는 것으로 알려져 있다. 위장관에 발생한 얼기형신경초종에 대한 증례는 매우 적으며, 지금까지 제2형 신경섬유종증 환자에서 위에 생긴 증례는 보고된 바가 없다. 본 증례에서 얼기형신경초종은 제2형 신경섬유종증 환자의 위에 점막하종괴로 발현하였으며, 지금까지 제2형 신경섬유종증환자에서 위에 발생한 얼기형신경초종을 보고한 예가 없기에 이의 영상의학적 소견을 기술하고자 한다.

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