^{18}F-Fluorodeoxyglucose PET/CT in a Patient with Esophageal and Genital Leiomyomatosis

Young-Sil An, MD
Deog-Yoon Kim, MD

Diffuse esophageal leiomyomatosis is a rare benign tumor, which can be associated with leiomyoma in female genital tracts involving the uterus, vagina, and vulva. Alport syndrome, an inherited disorder that includes the kidneys, eyes, and sensorineural hearing loss, is also rarely associated with these multiple leiomyomatosis. In our case, ^{18}F-fluoroseoxyglucose positron emission tomography/computed tomography was used to distinguish esophageal and genital leiomyomatosis from malignant masses.

Index terms:
Leiomyomatosis
Esophagus
Genital tract
^{18}F-FDG PET

DOI:10.3348/kjr.2009.10.6.632

Korean J Radiol 2009;10:632-634
Received March 9, 2009; accepted after revision May 18, 2009.

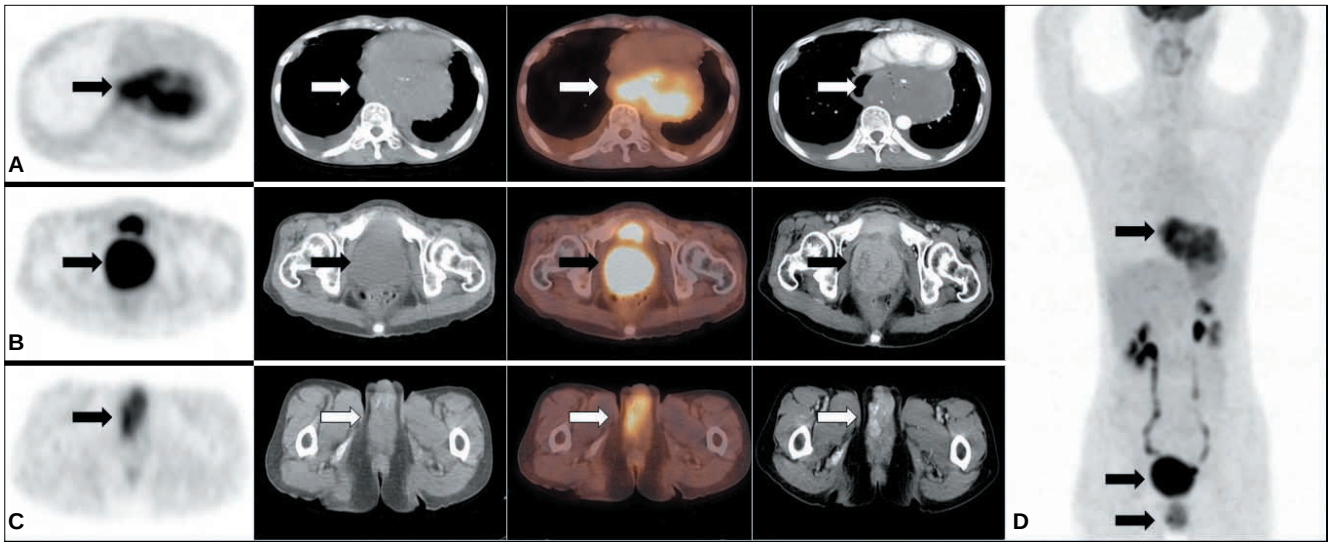
All authors: Department of Nuclear Medicine, School of Medicine, Kyung Hee University, Seoul 130-702, Korea

Address reprint requests to:
Deog-Yoon Kim, MD, Department of Nuclear Medicine, School of Medicine, Kyung Hee University, Hoegi-dong #1, Dongdaemun-gu, Seoul 130-702, Korea.
Tel. (822) 958-8215
Fax. (822) 958-8218
e-mail: petct@paran.com

Diffuse esophageal leiomyomatosis is a rare benign disease of proliferating smooth muscle that can be accompanied by leiomyoma of extraesophageal organs including the tracheobronchial and genital tracts (1–3). Moreover, diffuse leiomyomatosis can be associated with Alport syndrome in familial cases (2, 4). ^{18}F-fluoroseoxyglucose positron emission tomography (^{18}F-FDG-PET) can be useful in diagnosis since the benign masses can form lesions in the esophagus and extraesophageal organs that may be confused with malignancy. However, few reports have been published on the use of ^{18}F-FDG PET in diffuse leiomyomatosis involving multiple organs. We describe ^{18}F-FDG PET/computed tomography (CT) findings in a patient with esophageal and genital leiomyomatosis.

CASE REPORT

A 49-year-old woman was hospitalized for further evaluation of dysphagia. Esophagoscopy revealed a huge mass in the lower esophageal portion. Abdominal CT examination demonstrated a huge low-attenuated submucosal mass in the esophagus. Additional heterogeneous highly-attenuated masses were evident in the uterus and vulvar region. While diagnosis of gastrointestinal stromal tumor or leiomyoma is typical from such CT observations, preferred according to conventional CT, a malignancy cannot be fully excluded. Appropriately, the patient was referred to the nuclear medicine department for ^{18}F-FDG PET/CT to differentiate the esophageal and genital masses as malignant or benign. After a 6 hours fast, the patient was intravenously administered 370 MBq ^{18}F-FDG. At the time of injection, serum glucose level was 97 mg/dl. The patient was instructed to rest comfortably for 60 min while whole body PET/CT images were obtained using a Gemini Dual PET/CT scanner (Philips Medical Systems, Amsterdam, The Netherlands). Seven frames (2.5 min/frame) of emission PET data were acquired in a three-dimensional (3D) mode after non-contrast CT scans from the base of the skull to the upper thigh (tube rotation time of 1 second per revolution, 120 kV, 50 mA, 6.5 mm per rotation, and acquisition time of 43.52 second for a scan length of 856.5 mm). Emission PET images were



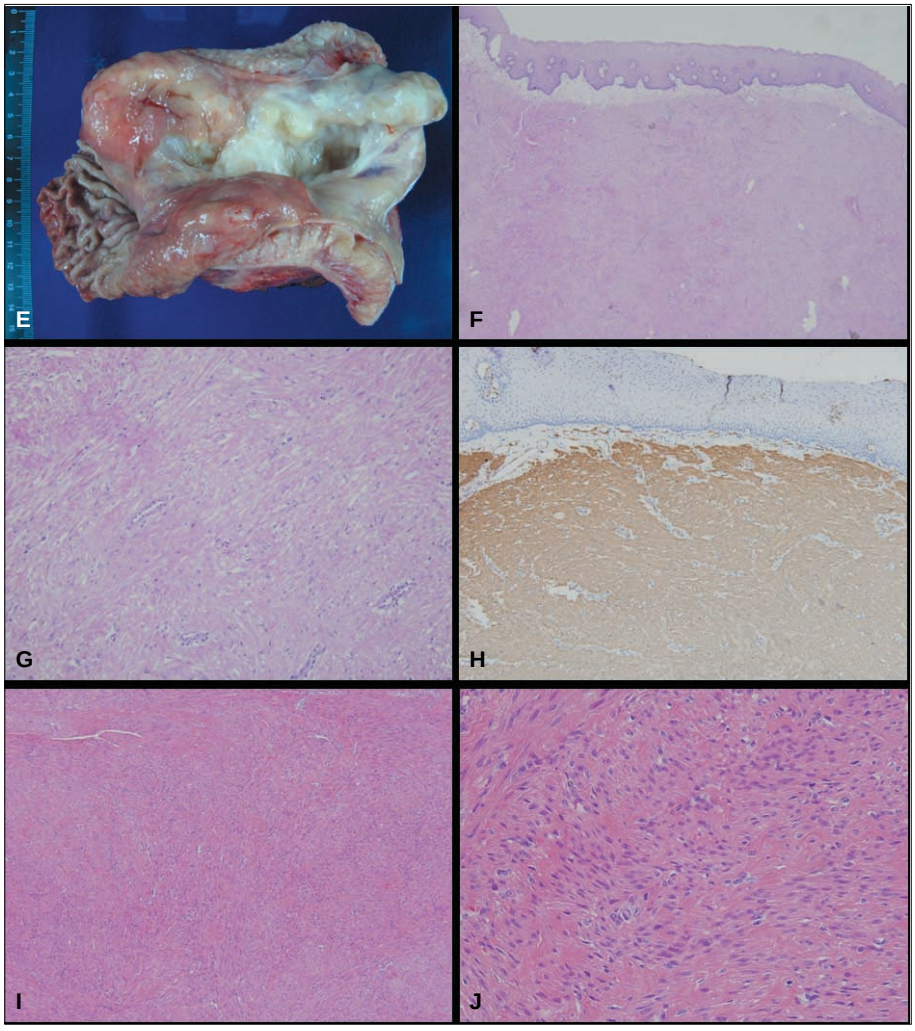


Fig. 1. Esophageal and genital leiomyomatosis

A. Huge abnormal increased ^{18}F-FDG uptake lesion (arrows) in esophagus (maximal SUV: 3.8) noted on PET/CT image (upper row). Corresponding contrast enhanced CT images demonstrate abnormal mass lesions in each organ.

B. Hypermetabolic lesions (arrows) were evident in pelvic region including uterus (maximal SUV: 6.13) (middle row).

C. Hypermetabolic lesions (arrows) were evident in vulvar region (maximal SUV: 2.81) (lower row).

D. Arrows point to lesions evident in on PET.

E-J. Pathologic examination. Grossly, 13 cm (length) × 8 cm (width) × 6 cm (depth) poorly circumscribed, sausage-like, white/pink, firm muscular mass oriented across distal esophagus and gastric cardia was evident. Diffusely thickened esophageal wall measured 2 cm in maximal thickness (**E**). Histologically, Hematoxylin & Eosin staining (× 40 in panel **F**, × 200 in panel **G**) reveals characteristics of leiomyoma. Multiple confluent well-differentiated smooth muscle proliferations were evident. Cells formed fascicles and interlacing bundles without cytologic atypia. Immunostaining for smooth-muscle actin (× 40) was strongly positive in smooth-muscle origin tumor cells with control reaction of microvillous projections in apical cell membrane (**H**). Uterus lesion also demonstrates as leiomyoma pattern in Hematoxylin & Eosin staining (× 100 in panel **I**, × 400 in panel **J**).

reconstructed with non-contrast CT using 3D-Row-Action Maximum-Likelihood Algorithm reconstruction (field of view = 576 mm, slice thickness = 4 mm, matrix size = 144 × 144, voxel size = 4.5 × 4.5 × 4 mm). Standardized uptake value (SUV) was calculated based on the injected dose and patient body weight. PET/CT demonstrated intensely increased ^{18}F-FDG uptake with 3.8 of maximal SUV in the esophagus, 6.13 in the uterus, and 2.81 in the vulvar mass (Fig. 1A-D), which was indicative of high probabilities of malignancy. Each mass was excised. Pathological testing confirmed that each mass was benign leiomyomatosis (Fig. 1E-J). The patient's son was remarkable for Alport syndrome, and had undergone removal of an esophageal leiomyomatosis nine years previously. However, the patient showed no specific abnormal sign of Alport syndrome.

DISCUSSION

Leiomyoma in the esophagus must be differentiated from malignant esophageal carcinoma, cyst, or bulbous stricture. Rarely, thickened layers of the esophageal wall can also be caused by diffuse leiomyomatosis, a benign muscular abnormality (1). Diffuse leiomyomatosis is also known as giant muscular hypertrophy with diffuse proliferation of the esophageal smooth muscle. The large tumor formation predominantly occurs in the middle and distal third of the esophagus. The most common clinical symptoms are dysphasia, vomiting, and retrosternal pain due to luminal narrowing and motor disorder of esophagus. These leiomyomatosis frequently involve extraesophageal organs such as tracheobronchial tree or female genital tract (uterus, vagina, vulva, and clitoris) and also show hyperplasia of the thoracic or genital smooth muscles (2, 3). The familiar form of leiomyomatosis can be associated with Alport syndrome, a hereditary disease that displays hematuric nephropathy and sensorial deficiencies including sensorineural hearing loss and congenital cataracts. It is transmitted as a X-linked dominant and may affect females without sign of nephropathy, which is revealed as Alport syndrome carrier status (4–7). This was presently the case. Previous reports have described increased ^{18}F-FDG uptake in leiomyomas (8–11). Leiomyoma with intense ^{18}F-FDG accumulation can be confused with a malignant lesion. The cause of hypermetabolism of this benign condition is probably due to higher levels of growth factors and increased peristaltic activity in smooth muscles (3, 8–11). Approximately 10% of leiomyomas in pre-manopausal women display focal ^{18}F-FDG uptake (maximal SUV > 3.0) (12). In fact, malignant tumors cannot be distinguished from hypermetabolic benign leiomyomas using ^{18}F-FDG PET. Thus, leiomyomatosis of multiple organs may be a cause of false-positive PET and represents a pitfall for correct diagnosis between benign and malignant lesions.

References

1. Compagnoni GM, Talamonti MS, Joob A, Ergun GA, Rao S. Esophageal leiomyomatosis in a woman with a history of vulvar leiomyoma and Barrett's esophagus: a case report and review of the literature. *Dig Surg* 2000;17:306-309
2. Kugelman A, Greif Y, Gershoni-Baruch R, Berkowitz D, Best LA, Guralnik L, et al. Pulmonary presentation of esophageal leiomyomatosis associated with Alport syndrome in childhood. *Isr Med Assoc J* 2003;5:293-294
3. Pujol J, Parés D, Mora L, Sans M, Jaurrieta E. Diagnosis and management of diffuse leiomyomatosis of the oesophagus. *Dis Esophagus* 2000;13:169-171
4. Rabushka LS, Fishman EK, Kuhlman JE, Hruban RH. Diffuse esophageal leiomyomatosis in a patient with Alport syndrome: CT demonstration. *Radiology* 1991;179:176-178
5. Garcia-Torres R, Cruz D, Orozco L, Heidet L, Gubler MC. Alport syndrome and diffuse leiomyomatosis. Clinical aspects, pathology, molecular biology and extracellular matrix studies. A synthesis. *Nephrologie* 2000;21:9-12
6. Kilian AK, Ringle T, Waag KL, Düber C, Neff KW. Pre- and postoperative MRI of esophageal and gastric leiomyomatosis in a pediatric patient. *AJR Am J Roentgenol* 2005;184:S129-S131
7. Dahan K, Heidet L, Zhou J, Mettler G, Leppig KA, Proesmans W, et al. Smooth muscle tumors associated with X-linked Alport syndrome: carrier detection in females. *Kidney Int* 1995;48:1900-1906
8. Meirelles GS, Ravizzini G, Yeung HW, Akhurst T. Esophageal leiomyoma: a rare cause of false-positive FDG scans. *Clin Nucl Med* 2006;31:342-344
9. Memisoglu E, Agarwal B, Akduman I, Prather C, Collins B, Civelek AC. Multimodality diagnostic imaging of diffuse esophageal leiomyomatosis. *J Comput Assist Tomogr* 2006;30:100-104
10. Ak I, Ozalp S, Yalçin OT, Zor E, Vardareli E. Uptake of 2-[18F]fluoro-2-deoxy-D-glucose in uterine leiomyoma: imaging of four patients by coincidence positron emission tomography. *Nucl Med Commun* 2004;25:941-945
11. Lee SM, Bae SK. Esophageal Leiomyoma with intense FDG uptake on 18F-FDG PET/CT. *Nucl Med Mol Imaging* 2008;42:410-413
12. Nishizawa S, Inubushi M, Kido A, Miyagawa M, Inoue T, Shinohara K, et al. Incidence and characteristics of uterine leiomyomas with FDG uptake. *Ann Nucl Med* 2008;22:803-810