

CT and MR Imaging Findings of Perianal Dermatofibrosarcoma Protuberans Mimicking Mucinous Adenocarcinoma Arising from Fistula in Ano: A Case Report¹

항문루에서 발생한 점액선암으로 오인된 용기성 피부섬유육종의
전산화단층촬영과 자기공명영상 소견: 증례 보고¹

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Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue neoplasm which typically originates in the dermis and spreads into the subcutaneous layer and muscle. We report an extremely rare case of a perianal DFSP mimicking mucinous adenocarcinoma arising from anorectal fistula in a 59-year-old woman. In case of a perianal mass, which is connected by a stalk to the anus and shows heterogeneous enhancement without related regional lymphadenopathy, DFSP should be included in the differential diagnosis.

Index terms

Dermatofibrosarcoma Protuberans
Mucinous Adenocarcinoma
Multidetector CT
Magnetic Resonance

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor with a low malignant potential and a high local recurrence rate after surgical excision (1). DFSP can be found anywhere on the body, but is often found on the chest and shoulders (1). DFSP arises in the dermis and invades deeper tissues such as fascia, muscle and bone (1, 2). Due to its tendency for local invasion and asymmetric spread, DFSP in the perianal area can extend to the anal sphincter and show similar imaging findings to those of a malignant tumor arising from the anorectal fistula. However, DFSP in the perianal area is extremely rare, and only one case has been reported in the English medical literature (3). To the best of our knowledge, there is no report regarding the detailed imaging findings of DFSP radiologically

mimicking mucinous adenocarcinoma arising from the anorectal fistula. We present an extremely rare case of perianal DFSP mimicking mucinous adenocarcinoma arising from fistula in ano, and focus on the CT and MR imaging findings. This report was approved by the ethics committee at our institution, and the requirement for informed consent was waived.

CASE REPORT

A 59-year-old woman presented to our hospital with a palpable mass in the left perianal and gluteal regions. The lesion appeared as a rounded, reddish, non-tender protuberance. CT examination was performed for evaluation of the mass using a 128-detector-row CT scanner (definition AS+, Siemens Medical Solutions, Forchheim, Germany). An enhanced CT of the abdo-

men and pelvis demonstrated a heterogeneously enhancing mass in the medial aspect of the subcutaneous layer of the left gluteal region and a communicating stalk between the mass and the anus, measuring 5.8 cm in size (Fig. 1A). Abdominal MR imaging was performed on a 3.0T system (Magnetom Verio; Siemens Medical Solutions, Erlangen, Germany) using a body phased-array coil, in order to obtain the differential diagnosis and initiate surgery planning. The mass showed bright signal intensity on fat-suppressed, T2-weighted images and iso-signal intensity to muscle on T1-weighted images (Fig. 1B, C). Following contrast infusion, heterogeneous enhancement in the mass and a communicating stalk adjacent to the internal sphincter of the anus were observed (Fig. 1D). On sigmoidoscopic examination, there was no evidence of a fistula opening. Excision of the mass was performed. Macroscopic examination of the excised tumor

showed a circumscribed, ovoid, pale brown mass measuring 6 cm in size, and the tumor abutted the internal sphincter of the anus without definite invasion. Photomicrography showed interwoven fascicles of cells creating a storiform pattern (Fig. 1E). Immunohistochemical staining was strongly positive for CD34 (Fig. 1F). These histologic features were compatible with a diagnosis of DFSP.

DISCUSSION

DFSP is an uncommon, soft-tissue neoplasm arising from the cutaneous tissue (1). DFSP is often found on the chest or shoulders, although it can also be found on other parts of the body (1, 2). The tumor often spreads aggressively locally into the subcutaneous tissue and occasionally involves the deeper tissues such

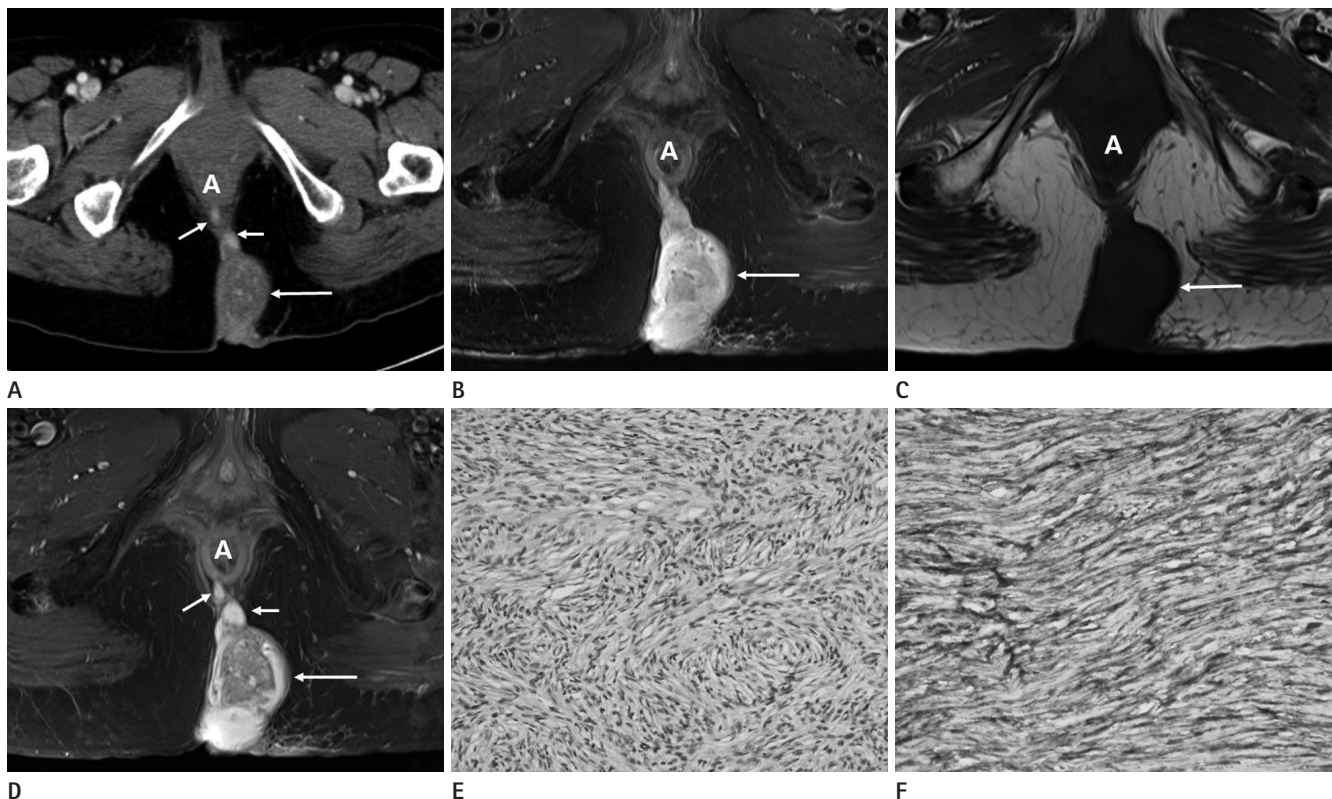


Fig. 1. A 59-year-old woman with perianal dermatofibrosarcoma protuberans (DFSP).

A. Axial enhanced CT image shows a heterogeneously enhancing mass (long arrow) in the medial aspect of the subcutaneous layer of the left gluteal region and with a communicating stalk (short arrows) between the mass and the anus, measuring 5.8 cm in size.

B, C. The mass (arrows) shows bright signal intensity on fat-suppressed, T2-weighted imaging (**B**) and iso-signal intensity to muscle on T1-weighted imaging (**C**).

D. Contrast-enhanced, fat-suppressed, T1-weighted MR images reveal heterogeneous enhancement of the mass (long arrow) and the connecting stalk (short arrows).

E. Photomicrograph shows interwoven fascicles of cells forming a storiform pattern (H&E, × 100).

F. Immunohistochemical staining is strongly positive for CD34. These findings have been considered characteristic for the diagnosis of DFSP (CD34 DAB, × 400).

as muscle and bone. It grows slowly and has a low metastatic potential, although it has a tendency to recur after excision (1). Surgical excision is the most important treatment for local DFSP, and maintaining an appropriate surgical margin can reduce the possibility of tumor recurrence (1, 4). For the complete excision of DFSP without leaving a residual tumor, preoperative CT and MRI are necessary for evaluating the exact size, extent, and invasion of adjacent structures (1, 4).

When the mass is small, DFSP appears on CT as a soft-tissue-density mass with homogeneous enhancement and a well-circumscribed margin. However, a mass larger than 5 cm in size shows heterogeneous enhancement with an irregular margin, probably due to the intratumoral degeneration and invasiveness (4-6). On MR imaging, DFSP shows iso- or low-signal intensity on T1-weighted images and intermediate or high-signal intensity greater than that of the subcutaneous fat on T2-weighted or fast spin-echo T2-weighted images (4, 6, 7). DFSP shows high-signal intensity, similar to that of water or blood vessels on fat-suppressed images (2, 7). Enhancement is generally homogeneous, but it can vary due to intratumoral necrosis or hemorrhage (2, 7). In our case, DFSP showed heterogeneous enhancement and a stalk extending to the anal sphincter. One report suggests that a stalk found between the anal sphincter and a heterogeneous mass in the perianal area is a characteristic finding of mucinous adenocarcinoma arising from fistula in ano (8). Mucinous adenocarcinoma arising from an anal fistula often shows peritumoral invasion and regional lymphadenopathy in 45% of the patients (9). In comparison, DFSP can invade the adjacent structures, although it rarely shows regional lymphadenopathy or lymph node metastasis (5, 9). The contrast-enhancement pattern of mucinous adenocarcinoma is characteristically heterogeneous (9). Mesh-like heterogeneous enhancement and more than 5-mm-sized enhancing components in the intratumoral mucin pool favor the diagnosis of mucinous adenocarcinoma (9).

For localized DFSP, surgical excision is the primary treatment, and maintaining an appropriate surgical margin is important for reducing tumor recurrence (1). Some surgical centers perform sequential removal of thin layers of the tumor with histological examination of each layer during the surgery (3). The sequential removal technique helps to achieve a completely negative margin, and thus dramatically reduces the recurrence rate (3). For mucinous adenocarcinoma, abdominoperineal resection (APR)

and colostomy is usually performed. APR is an extensive surgery performed to remove the anus, rectum, and sigmoid colon (10). Therefore, making the correct diagnosis based on the radiological and pathological findings is crucial for selecting appropriate patient management.

In conclusion, DFSP should be included in the differential diagnosis when there is a perianal mass that is connected by a stalk to the anus without regional lymphadenopathy.

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항문루에서 발생한 점액선암으로 오인된 용기성 피부섬유육종의 전산화단층촬영과 자기공명영상 소견: 증례 보고¹

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용기성 피부섬유육종은 진피 및 피하연부조직에서 발생하는 드문 연부조직 종양이다. 저자들은 용기성 피부섬유육종이 항문주변부위에서 발생하여 항문루에서 발생한 점액선암으로 오인된 매우 드문 증례를 전산화단층촬영과 자기공명영상 소견을 중심으로 소개하고자 한다. 영상소견에서 항문과의 연결성을 보이는 항문주위의 종괴가 비균질적 조영증강을 보이며 동반된 국소림프절종대가 없다면 용기성 피부섬유육종을 감별해야 한다.

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