F-18 FDG PET/CT Findings of Spontaneous Mesenteric Fibromatosis in a Patient with Gardner’s Syndrome

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Gardner’s syndrome (GS), a variant of familial adenomatous polyposis, is an autosomal dominant disease. Originally, Gardner described a syndrome consisting of hereditary intestinal polyposis with osteomas and multiple cutaneous and subcutaneous lesions [1] (Figs. 1 and 2). The syndrome was later modified by the addition of other features, such as dental abnormalities, abdominal fibromatosis, and a number of malignant tumors. The principal cutaneous lesions that have been described in GS are epidermoid cysts. Other cutaneous lesions include fibromas, lipomas, leiomyomas, neurofibromas, and pigmented skin lesions [2]. Fibromatoses are histologically benign, but locally aggressive fibrous tumors consisting of mature fibroblasts within an extensive collagen matrix. Most cases are sporadic, but there is a clear association with familial adenomatous polyposis and GS, suggesting a link with a mutation of the APC gene on chromosome 5q22 [3]. Fibromatosis occurs in 3.5%–29% of patients with GS, and is more likely to be multiple and to involve the mesentery and abdominal wall rather than being an isolated form [4, 5]. Clinically, fibromatosis presents as a painless firm soft tissue mass. Most cases of fibromatosis are believed to be precipitated by surgical trauma; however, a few cases of spontaneous occurrence have

Fig. 1 A 20-year-old man was admitted to oral surgery with a dental problem. A panoramic radiograph showed multiple osteomas in the right angle and anterior portion of the mandible with dental abnormalities (impacted or unerupted teeth, and supernumerary teeth). Surgery was performed, and a biopsy confirmed osteomas.
been reported [6]. In our patient, no history of abdominal surgery or trauma was present. In addition, an abdominal CT obtained 2 years ago revealed no abnormality.

Although the radiological features of fibromatosis on CT or MR have been described in the literature, F-18 FDG-PET or PET/CT findings are rarely reported [7–10]. The F-18 FDG uptake in patients with fibromatosis ranged from low to moderate grade and was generally heterogenous with a few tiny foci of relatively intense uptake or relatively homogenous. The areas of higher FDG metabolism are likely to represent more cellular and mitotically active areas [7]. Mesenteric fibromatosis has similar findings to extra-abdominal lesions [7, 9] (Fig. 3).

Fig. 3 Routine laboratory studies for dental surgery showed abnormal liver function tests. The patient was referred to the Gastroenterology Service. Computed tomography (CT) of the abdomen demonstrated a large soft tissue mass in the right lower quadrant (RLQ), suggesting a small bowel lymphoma with intussusception. A F-18 FDG PET/CT was performed to assess the tumor extent and determine the appropriate site of biopsy. (a) A maximum-intensity-projection (MIP) image showed an oval-shaped, moderate-grade FDG uptake interspersed with a few tiny foci of relatively intense uptake in the RLQ of the abdomen (arrow; SUVmax, 5.6). (b, c) Coronal (fused PET/CT) and transverse (upper image, fused PET/CT; middle image, contrast enhanced-CT; lower image, PET) images through the mass revealed an 8×6-cm, enhanced, soft tissue mass with moderate-grade FDG accumulation in the RLQ. Upper and lower gastrointestinal endoscopy revealed multiple adenomatous polyps in the colon. Laparotomy revealed a firm mass in the mesentery. Biopsy confirmed a fibromatosis (also known as a desmoid tumor). The patient was diagnosed with Gardner’s syndrome, with multiple osteomas in the mandible, dental abnormalities, mesenteric fibromatosis, multiple colonic polyps, and neurofibroma.

Conflict of Interest The authors declare that they have no conflict of interest.

References