Diffuse Abdominal Uptake of Technetium-99m-HDP after Colectomy in Gardner's Syndrome

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A 37-yr-old man presented with increasing abdominal girth and multiple palpable intra-abdominal masses 3 yr after colectomy for polyposis coli. Whole-body skeletal scintigraphy performed prior to laparotomy demonstrated diffuse abdominal uptake of ^{99m}Tc-HDP consistent with mesenteric fibromatosis confirmed at surgery. When diffuse abdominal uptake of skeletal imaging agents occurs in patients with prior colectomy for polyposis coli, mesenteric fibromatosis as a manifestation of Gardner's syndrome should be suspected. This case illustrates another cause of diffuse abdominal uptake of skeletal imaging agents.

Key Words: skeletal scintigraphy; technetium-99m-HDP; Gardner's syndrome; mesenteric fibromatosis; polyposis syndromes

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Gardner's syndrome is characterized by premalignant gastrointestinal adenomatous polyposis associated with various skeletal/dental and soft-tissue abnormalities such as mesenteric fibromatosis (intra-abdominal desmoids) (*1–4*). Since colonic polyposis uniformly leads to colorectal adenocarcinoma, colectomy is performed prophylactically. Unfortunately, mesenteric fibromatosis occurs frequently after colectomy, causes significant morbidity due to aggressive local invasiveness and is difficult to eradicate. We present a case of Gardner's syndrome in which skeletal scintigraphy demonstrates diffuse abdominal uptake of ^{99m}Tc-HDP following colectomy for polyposis coli.

CASE REPORT

A 37-yr-old man presented with a 1-yr history of increasing abdominal girth and multiple nodular intra-abdominal masses. He presented with anemia and occult gastrointestinal bleeding at age 22 which led to subtotal colectomy for polyposis coli at age 34. He denied abdominal pain, early satiety, nausea, vomiting, skeletal pain or constitutional symptoms and continued to have regular bowel movements. There was no family history of the condition.

Physical exam was remarkable only for marked abdominal distension with multiple nodular intra-abdominal masses and mild cachexia. Laboratory tests revealed mildly reduced hemoglobin and hematocrit of 12.0 and 37.2, respectively. A CT scan of the abdomen demonstrated multiple nonenhancing nodular masses throughout the mesentery and anterior abdominal wall without organ involvement (Fig. 1). Skeletal scintigraphy was performed with a dual-head gamma camera 3 hr postinjection of 740 MBq ^{99m}Tc-hydroxymethyl diphosphonate (HDP). The study demonstrated diffuse abdominal uptake of radiotracer with otherwise physiologic distribution (Fig. 2).

The patient underwent laparotomy in an attempt to surgically debulk the abdomen. A larger anterior wall tumor was excised (Fig. 3), while the massive mesenteric desmoid tumor burden was found to be unresectable. The patient's postoperative course was unremarkable. He was discharged and started on tamoxifen.

DISCUSSION

A syndrome characterized by a triad of intestinal polyposis, various soft-tissue tumors and multiple osteomas was first described in 1912 (*1*). Approximately 40 yr later, Gardner et al. established a genetic basis for the syndrome and described additional manifestations, including dental anomalies, desmoid tumors and various skeletal lesions. This syndrome became known as Gardner's syndrome (*2–5*).

Gardner's syndrome belongs to the dyshistogenetic syndromes characterized by hamartoma, hyperplasia and a propensity for neoplasia. All three germ layers are involved (*6*). Two entities, familial ademtous coli and Gardner's syndrome, are in this category. Although they were initially thought to be independent clinical syndromes, current evidence suggests that both are caused by different mutations of the same gene located on the long arm of the fifth chromosome. There is much overlap and less distinction between familial adenomatosis coli and Gardner's syndrome (*4–7*).

Gardner's syndrome occurs in approximately one in 14,000 individuals. Approximately two-thirds of cases are familial, and the remainder occur as sporadic mutations. Transmission of the syndrome is as an autosomal dominant with nearly complete penetration but markedly variable expression. Described manifestations include: (a) multiple adenomatous polyps of the colon, (b) mesenteric fibromatosis, (c) multiple skeletal and dental abnormalities, (d) cutaneous and ocular lesions and (e) increased

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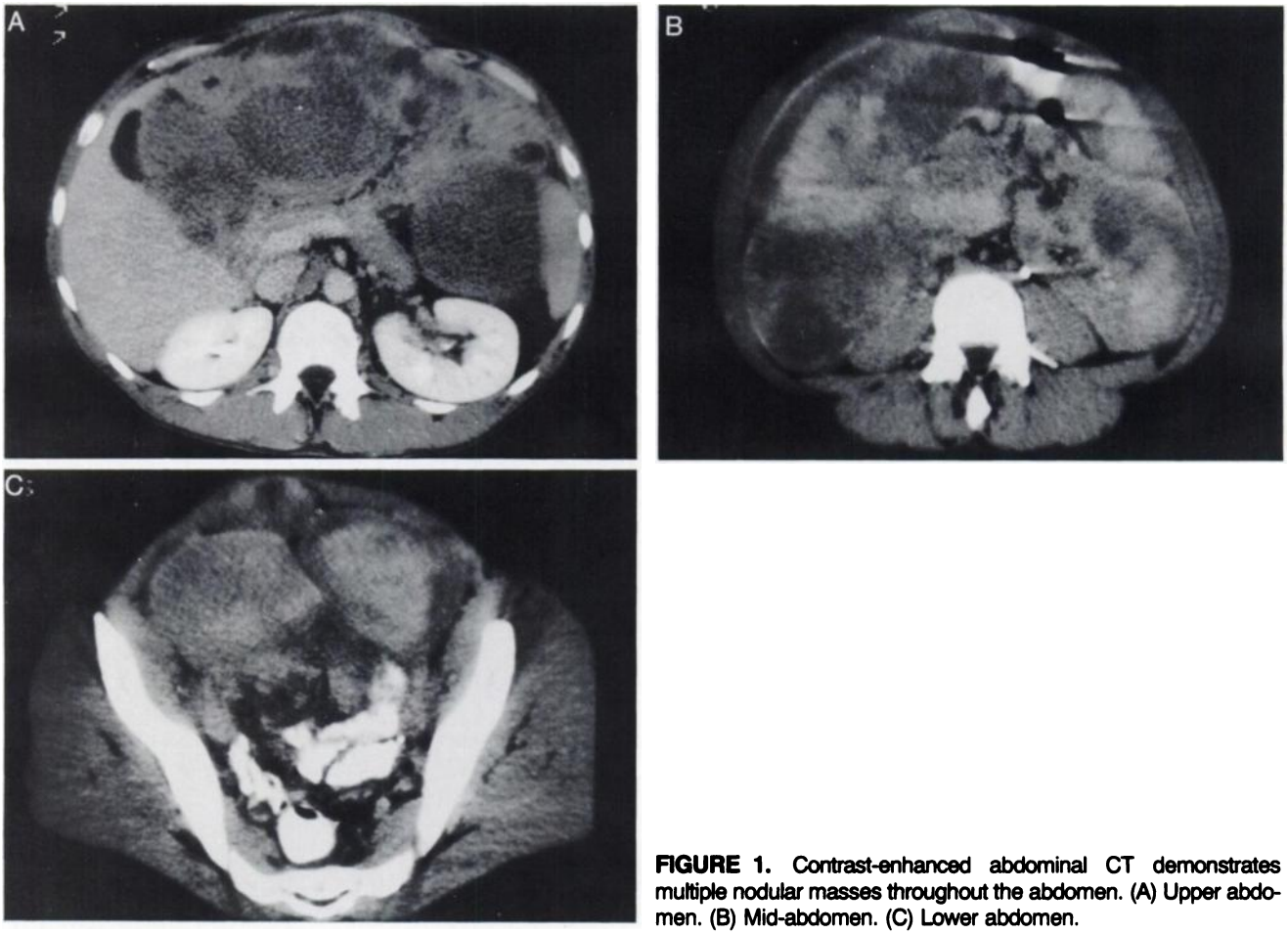

FIGURE 1. Contrast-enhanced abdominal CT demonstrates multiple nodular masses throughout the abdomen. (A) Upper abdomen. (B) Mid-abdomen. (C) Lower abdomen.

incidence of gastrointestinal and extra-gastrointestinal neoplasms.

The adenomatous polyps represent the major hallmark of the disease, as well as the most significant abnormality. They occur classically in the colon but may occur anywhere from the stomach to the terminal ileum as well. They typically manifest themselves in the first or second decade of life and approximately 100% result in malignancy approximately 10–15 yr after onset, thus making early colectomy the primary treatment. Mesenteric fibrosis typically occurs 1–2 yr after polyps surgery, but it may occur spontaneously. The fibromatosis may be asymptomatic, but also may cause pain or intestinal obstruction. Rapidity of growth is variable. These masses are important because they may involve rectal structures. Surgical resection must be complete since the lesion is extremely aggressive. Recurrence is common.

The bony lesions consist primarily of osteomas which typically occur in the craniofacial region. In the long bones, the manifestation is more commonly cortical hyperostosis rather than osteomas. These lesions are important in that their discovery may predate the gastrointestinal lesions. Dental lesions consist of supernumary and unerupted teeth, dentigerous cysts, hypercementomas and multiple caries. Sebaceous cysts about the face and neck are also common and predate the polyposis. Pigmented lesions of the ocular fundus also occur.

Interestingly, there is an increased incidence of non-gastrointestinal malignancies. Thyroid and basal cell carcinomas, as well as a variety of sarcomas, have also been reported. Fibrosarcomas have been reported but may in fact represent misinterpretation of aggressive fibromatosis. Central nervous system tumors, glioblastomas and medulloblastomas have been reported as well (Turcot's syndrome).

Both skeletal and ^{67}Ga scintigraphy have been used in the evaluation of Gardner's syndrome (*12*). Both may localize and identify desmoid tumors and skeletal lesions. Since, as mentioned above, extra-intestinal manifestations may precede the usually symptomless colonic polyposis, which is uniformly fatal without colectomy, early diagnosis may be life saving. Discovery of multiple osteomas, cortical hyperostosis of the long bones and desmoid tumors on routine skeletal or whole-body ^{67}Ga scans should lead one to suspect Gardner's syndrome.

Most commonly, scintigraphy may be used to localize

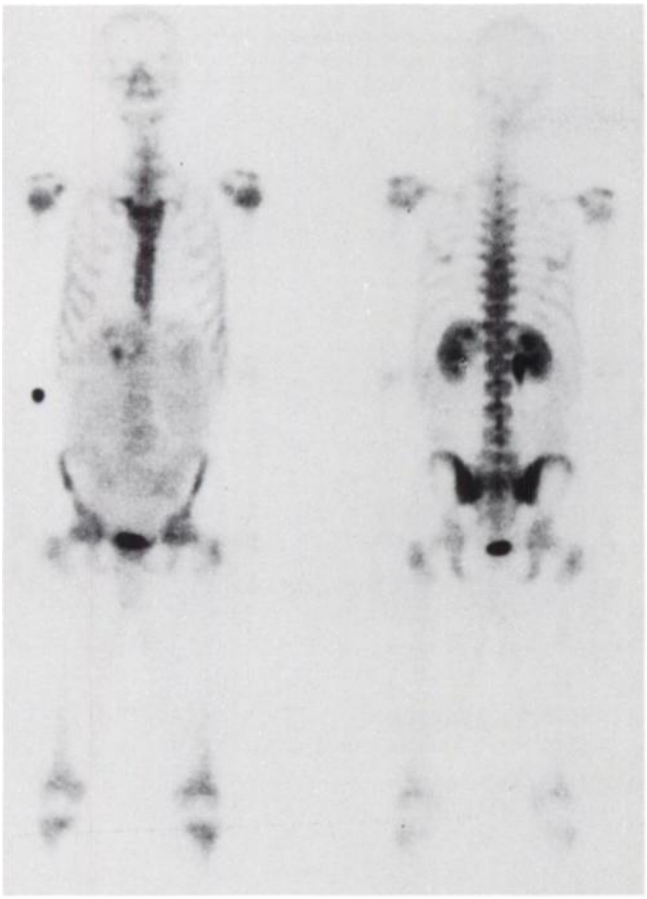

FIGURE 2. Whole-body-bone images performed 3 hr postinjection of 740 MBq ^{99m}Tc-HDP show diffuse abdominal uptake. The observed increased activity in the end of the long bones is thought to reflect bone marrow expansion secondary to prolonged anemia.

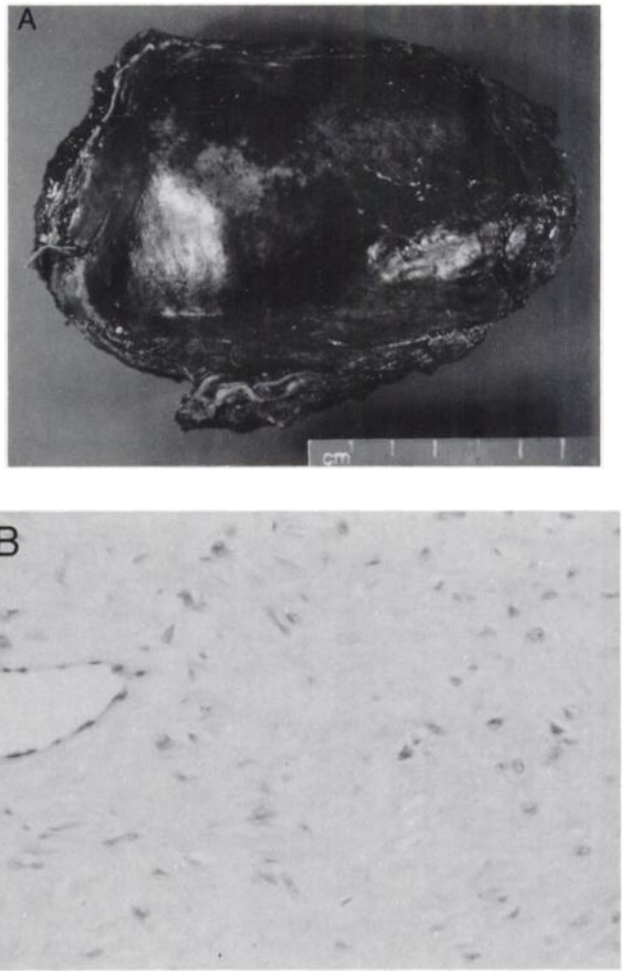

FIGURE 3. (A) Excised anterior abdominal wall desmoid. (B) High-power microscopy demonstrates cells with spindle-shaped nuclei and abundant collagen characteristic of desmoid tumors.

and define the extent of isolated desmoid tumors (*9,10,11,13,15*). Technetium(V)-99m-dimercaptosuccinic acid has been found to be superior to ^{67}Ga for the detection of desmoid tumors (*14*). Bone scintigraphy appears to be the most commonly used adjunct to other modalities. At Crawford Long Hospital in Atlanta, triple-phase skeletal scintigraphy is used since an early vascular blush helps further define the marginal extent of the tumor, while delayed images may demonstrate involvement of adjacent bone, and therefore, absence of a surgical plane. Fibrosarcoma, which tends to be more hypervascular, may also be excluded. The mechanisms of localization in these tumors have not been established.

In this patient, skeletal scintigraphy excluded extra-abdominal skeletal lesions and/or desmoids. Although benign, this patient will undoubtedly experience progressive morbidity and eventual mortality from mesenteric fibromatosis. At discharge he was started on tamoxifen which, along with a variety of other agents, including progestinal agents, radiotherapy and even indomethacin, has resulted in objective tumor regression (*12*).

REFERENCES

1. Devic A, Bussy MM. Un cas de polypose adenomateuse generalisée autour de l'intestine. *Arch Mal Appar Dig* 1912;6:278–299.
2. Gardner EJ, Plenk HP. Hereditary patterns for multiple osteomas in a family group. *Am J Hum Genet* 1952;4:31–36.
3. Gardner EJ, Richards RC. Multiple cutaneous and subcutaneous lesions occurring simultaneously with hereditary polyposis and osteomatosis. *Am J Hum Genet* 1953;5:139–147.
4. Gardner EJ. Follow-up study of a family group exhibiting dominant inheritance for a syndrome including intestinal polyps, osteomas, fibromas and epithelial cysts. *Am J Hum Genet* 1962;14:376–390.
5. Pierce ER, Weisbord T, McKusick VA. Gardner's syndrome: formal genetics and statistical analysis of a large canadian kindred. *Clin Genet* 1970; 1:65–80.
6. Cohen MM. Syndromology: an updated conceptual overview. II. Syndrome classifications. *Int J Oral Maxillofac Surg* 1989;18:223–228.
7. Halling F, Merten HA, Lepsien G, Honig JF. Clinical and radiological findings in Gardner's syndrome: a case report and follow-up study. *Dentomaxillofac Radiol* 1992;21:93–98.
8. Harned RK, Buck JL, Olmsted WW, Moser RP, Ros PR. Extracolonic manifestations of the familial adenomatous polyposis syndromes. *Am J Roentgenol* 1991;156:481–485.
9. Chew FS, Hudson TM, Enneking WF. Radionuclide imaging of soft-tissue neoplasms. *Semin Nucl Med* 1981;11:266–276.
10. Lessig HJ, Devenney JE. Localization of bone-seeking agent within a desmoid tumor. *Clin Nucl Med* 1979;4:164–165.

11. Sty JR, Starshak RJ, Oechler H. Extra-osseous uptake of ^{99m}Tc-MDP in congenital fibromatosis. *Clin Nucl Med* 1981;6:123.
12. Hardoff R, Ben Dov D, Font A. Gallium-67 scintigraphy in the evaluation of Gardner's syndrome. *Cancer* 1988;61:2353–2358.
13. Hitoya Ohta, Keigo Endo, Jungi Konishi, et al. Scintigraphic evaluation of aggressive fibromatosis. *J Nucl Med* 1990;31:1632–1634.
14. Hitoya Ohta, Keigo Endo, Toru Fujita, Tetsuo Nakajima, et al. Imaging of soft-tissue tumors with technetium(V)-99m dimercaptosuccinic acid: a new tumor seeking agent. *Clin Nucl Med* 1984;10:568–573.
15. Hudson TM, Vandergriend RA, Springfield DS, Hawkins IF Jr, Spanier SS, Enneking WF, Hamlin DJ. Aggressive fibromatosis: evaluation by CT and angiography. *Radiology* 1984;150:568–573.

(continued from page 7A)

FIRST IMPRESSIONS:
A GRADE I STRESS FRACTURE IN THE RIGHT FEMUR

PURPOSE
A 34-yr-old man who plays soccer was recently referred for evaluation of pain in the right thigh. A three-phase bone scan showed normal anterior flow (Fig. 1A) and blood-pool activity (Fig. 1B) in the right thigh. Delayed images of the anterior pelvis and femora apparently showed irregular, linear increased tracer uptake in the medial cortex of the right midfemur (Fig. 2, arrows). This uptake, however, was not seen in the posterior view; therefore, a right lateral view was obtained which showed that the abnormal activity was actually confined to the soft tissues of the right thigh (Fig. 3, arrow) and is consistent with myostitis ossificans. A plain film radiograph (Fig. 4) showed a similar pattern of calcification in the soft tissues.

TRACER
Technetium-99m-methylene diphosphonate, 740 MBq

ROUTE OF ADMINISTRATION
Intravenous

TIME AFTER INJECTION
3 hours

INSTRUMENTATION
Vision 1024 RZ, Summit Nuclear

CONTRIBUTORS
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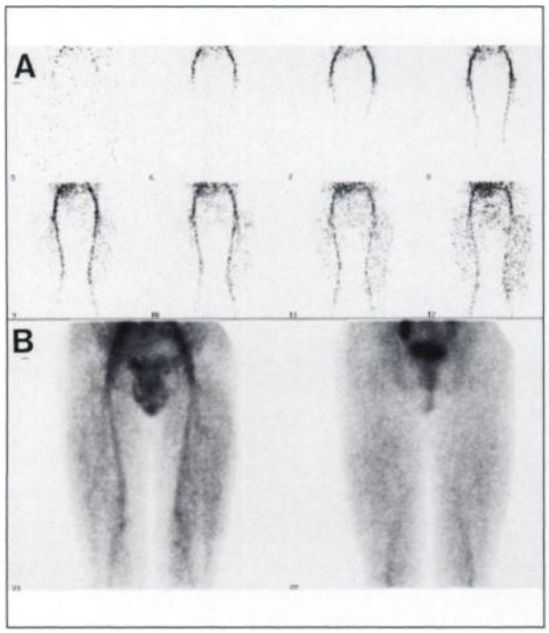

FIGURE 1.

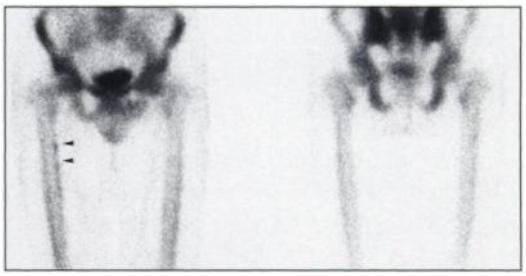
FIGURE 2.


FIGURE 3.

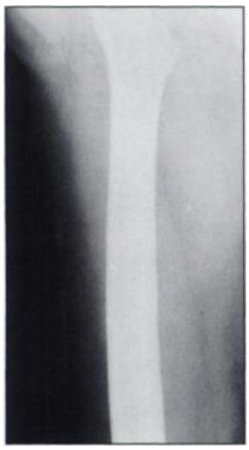
FIGURE 4.