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Case report

Pancreatic metastasis of dermatofibrosarcoma protuberans

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Dermatofibrosarcoma protuberans (DFSP) is a relatively rare skin tumor that is considered to have intermediate malignancy; it demonstrates frequent local recurrence, but systemic metastasis is rare. We report a 49-year-old woman with pancreatic metastasis of DFSP who underwent total pancreatectomy with partial resection of the portal vein. Except for our patient, only two other cases of pancreatic metastasis of DFSP have been reported in the literature, to our knowledge. Radical resection may be considered for pancreatic metastasis of DFSP when there are no other metastatic lesions.

Key words: dermatofibrosarcoma protuberans, pancreas, metastasis, total pancreatectomy

Introduction

Dermatofibrosarcoma protuberans (DFSP), first described by Darier and Ferrand in 1924,[1] is a relatively rare skin tumor. DFSP is considered to have intermediate malignancy; it frequently exhibits local recurrence and, rarely, systemic metastasis. In this report, we present a case of pancreatic metastasis of DFSP in a patient who was treated by total pancreatectomy (TP) with partial portal vein resection. Pancreatic metastasis of DFSP is extremely rare. The only two other reported cases of pancreatic metastasis of DFSP are reviewed in this report.

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Case report

A 49-year-old woman had undergone excision of a giant DFSP tumor in the left lower abdomen, and the defect site had been reconstructed with a myocutaneous flap from the thigh. After the operation, adjuvant radiation therapy (total dose, 50 Gy) was performed. Ten months later, she was admitted to our department because of DFSP metastatic lesions in the pancreas and the left gluteus maximus muscle, detected on computed tomography (CT) (Fig. 1A,B).

The patient underwent resection of the metastatic lesion in the left gluteus maximus muscle, and total pancreatectomy (TP) was performed. Because the tumor had directly invaded the pancreas, a 3-cm segment of the portal vein was resected and reconstructed by end-to-end anastomosis. Macroscopically, the tumor was elastic hard and $10 \times 5 \times 5$ cm in size (Fig. 2A). The cut surface of the tumor was nonencapsulated and grayish. Microscopically, the tumor was composed of spindle cells arranged in a storiform and herringbone pattern (Fig. 2B). The tumor cells had invaded the pancreatic parenchyma and the portal vein. She recovered uneventfully, and after 7 months of follow-up, no recurrence or metastasis was detected.

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a soft-tissue neoplasm of intermediate malignancy and is considered to be a low-grade sarcoma. Most DFSPs occur during middle age (20–50 years).[2] Although DFSP lesions have low metastatic potential, they have a pronounced tendency to recur, and there are occasional reports of metastases and death. Metastases have been reported to occur in 1% to 6% of patients, and are a sign of poor prognosis, most often resulting in death within 2 years.[3] Multiple and repeatedly recurrent tumors

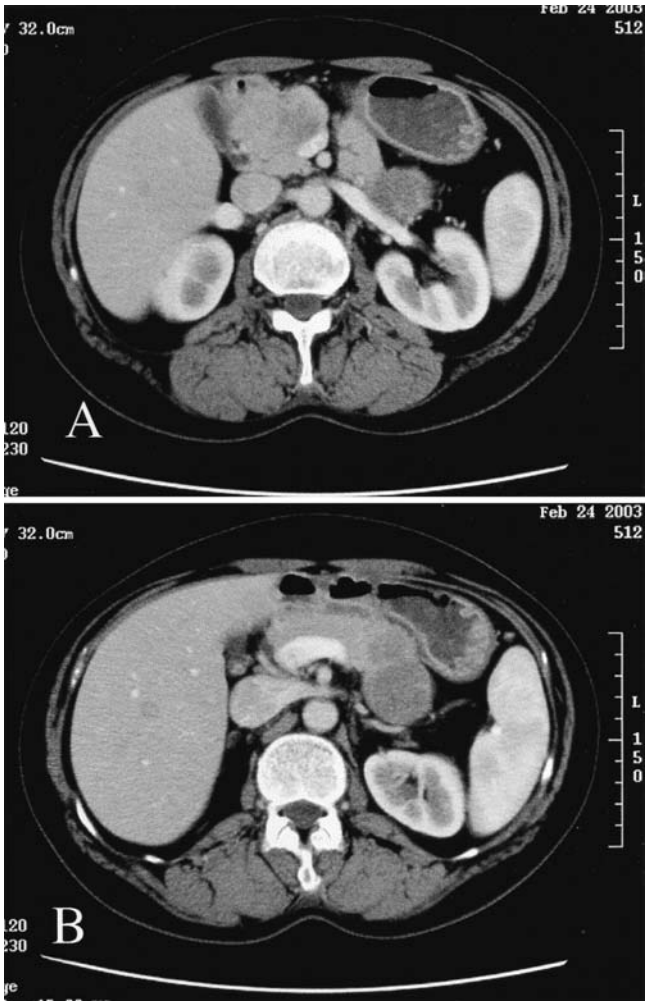

Fig. 1A,B. Computed tomography (CT) shows tumors in **A** the pancreatic head and **B** pancreatic body

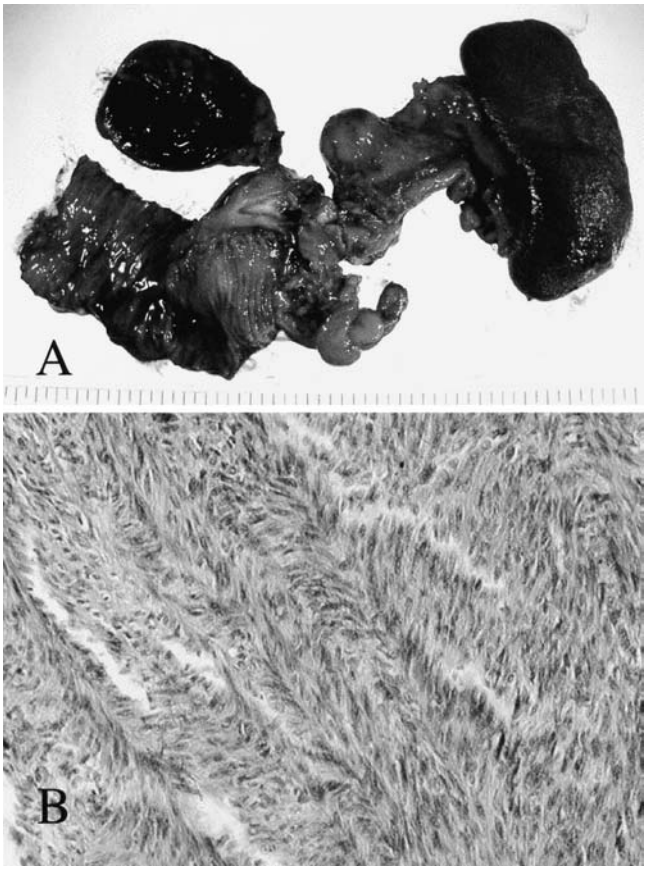

Fig. 2. A shows the macroscopic appearance of the specimen. **B** Microscopically, the tumor is composed of spindle cells arranged in a storiform and herringbone pattern. H&E, ×200

appear to be the greatest risk factors for metastatic disease, and the lung is the most common site of metastases (75%), via hematogenous spread.[4,5] In the recent literature,[6–8] eight patients with pulmonary metastasis were reported. Although one patient underwent lung resection, he died of this disease.[8] Among the various histological subtypes, it has been suggested that DFSPs with fibrosarcomatous (DFSP-FS) areas may have an increased likelihood of adverse outcome. DFSP-FS tends to have higher proliferative activity than DFSP without FS area.[9] In the present patient, the tumor had an FS area and was presumed to have high proliferative activity. The histologic characteristics of the tumor in the present patient are summarized in Table 1. All resected lesions; i.e., the primary lesion, and the metastatic lesions in the pancreas and the gluteus maximus, had a fibrosarcomatous area, herringbone pattern, and a high number of mitoses (25–30 mitoses per ten high-power fields). Immunohistochemically, the fibrosarcomatous cells showed expression of Ki-67 and loss of CD34.

Pancreatic metastasis of DFSP is extremely rare; only two cases have already been reported in the English-language literature, to our knowledge. In one case, autopsy revealed pancreatic metastasis after the patient died of brain metastasis.[10] The other patient was diagnosed as having a mass in the head of the pancreas and had undergone pancreaticoduodenectomy.[11] The three patients' details, including those of the present patient, are summarized in Table 2.

Regarding therapy, DFSP should be completely excised with tumor-free margins to avoid local recurrence. To achieve this goal, removal of a 2- to 3-cm-wide margin of visibly uninvolved tissue, including underlying fascia, is recommended for subjects aged more than 5 years.[12] Because there are patients who have survived after the resection of distant metastatic lesions of this disease,[13] radical resection may be considered for pancreatic metastasis of DFSP when there are no other metastatic lesions. DFSP has been proposed to be a relatively radiosensitive tumor, and radiation at doses of 50–60 Gy should be considered as an adjuvant to

Table 1. Histologic characteristics of the tumor in the present patient

		Metastastatic lesion	
	Primary	Pancreas	GM
Fibrosarcomatous area	+	+	+
Herringbone pattern	+	+	+
Number of mitoses (per ten HPFs)	25	30	25

GM, gluteus maximus; HPFs, high-power fields

Table 2. Reported patients with pancreatic metastasis of dermatofibrosarcoma protuberans

Patient no.	Year	Age (years)	Sex	Site	Size[a]	Treatment
1	1990[10]	36	M	NA	NA	Autopsy
2	2003[11]	29	F	Ph	6.0	PD
3	Present patient	49	F	Phbt	10.0	TP

NA, not analyzed; Ph, pancreas head; Phbt, pancreas head, body, and tail; PD, pancreaticoduodenectomy; TP, total pancreatectomy
[a] Maximum diameter (cm)

resection if the margins are positive.[2,3,14,15] However, the long-term prognosis of patients who have received radiation therapy is unclear.

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