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Case Report

Desmoplastic fibroma of the scapula with fluorodeoxyglucose uptake on positron emission tomography: a case report and literature review

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Abstract: We present a case of desmoplastic fibroma (DF) arising from the right scapula that was incidentally identified by fluorodeoxyglucose-positron emission tomography (FDG-PET) imaging performed to evaluate the presence of metastasis due to a history of surgical treatment for endometrioid adenocarcinoma. A 65-year-old woman was admitted to our hospital for consultation about a bone lesion in the right scapula although she was asymptomatic. FDG-PET revealed moderate focal ^{18}F-FDG uptake in the right scapula with a maximal standardized uptake value of 3.2. The lower angle of the scapula was unclear on plain radiology. Needle biopsy was performed to make a differential diagnosis between primary bone and metastatic tumor. Pathologically, the tumor was composed of a relatively sparse proliferation of spindle-shaped fibroblastic/myofibroblastic cells in a dense collagenous background. Therefore, the diagnosis was a primary fibrous bone tumor. Wide excision was performed, because of the possibility of malignant tumors such as low-grade fibrosarcoma in light of the FDG-PET uptake. Pathologically, the resected tumor was composed of a proliferation of less atypical spindle cells in the collagenous stroma with focally myxoid change; no mitotic figures were observed. Immunohistochemically, β-catenin nuclear/cytoplasmic staining was not observed, and no β-catenin genetic mutations were detected. Therefore, the tumor was diagnosed as DF. DF is a tumor that exhibits FDG-PET uptake. There were no signs of recurrence 6 months after surgery.

Keywords: Desmoplastic fibroma, scapula, β-catenin, positron emission tomography (PET)

Introduction

Desmoplastic fibroma (DF) is a rare, locally aggressive, solitary tumor microscopically composed of well-differentiated myofibroblasts with abundant dense collagen deposition [1]. The incidences of DF among all primary and benign bone tumors are 0.06–0.11% and 0.3%, respectively [2, 3]. The common sites are the long tubular bones (56%), mandible (26%), and pelvis (14%) [4, 5]. DF has been reported in rarer locations as well. However, DF arising from the scapula is extremely rare; to our knowledge, only 4 cases have been published [6-9].

The radiological characteristics of DF on magnetic resonance imaging (MRI) and computed tomography (CT) are well described in many published cases [3, 6, 10, 11]. Typical radiological features include osteolytic lesion with destruction of the cortical bone, pseudotrabeculation, and marginal sclerosis. However, these findings are indistinct from those of other bone tumors such as fibrous dysplasia, hemangioma, eosinophilic granuloma, central low-grade osteosarcoma, and metastatic tumors. The radiological diagnosis of DF is sometimes difficult because of its rarity and nonspecific radiographic findings. On the other hand, positron emission tomography (PET) is useful for locating recurrence and metastasis of malignant tumors in addition to detecting primary malignant tumors. PET and PET/CT imaging have been used for musculoskeletal tumors [12-16]. However, the fluorodeoxyglucose (FDG)-PET

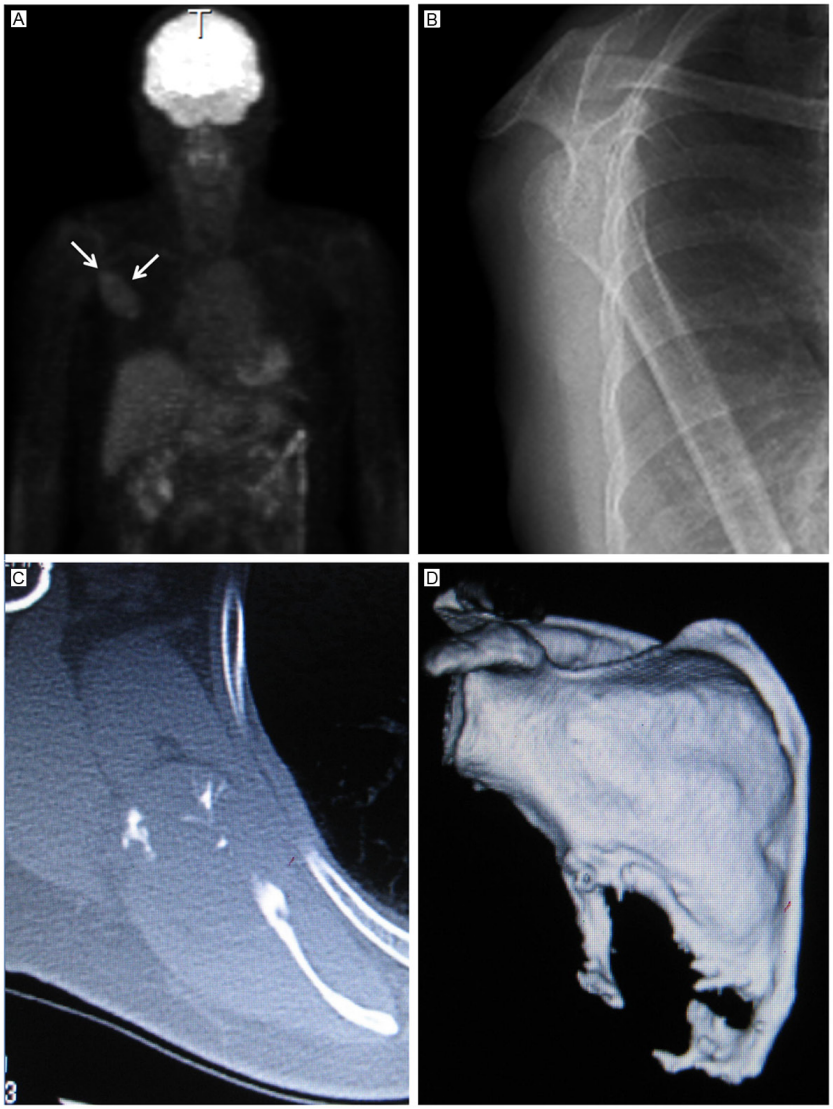

Figure 1. A: Positron emission tomography showing moderate focal ^{18}F-fluorodeoxyglucose uptake in the right scapula with a maximal standardized uptake value of 3.2; B: Plain radiology showing the unclear lower angle of the scapula; C, D: Computed tomography (C) and 3-dimensional computed tomography (D) showing a 61 × 42 × 27-mm osteolytic lesion with destruction of the cortical bone and a partially disappeared scapular rim.

findings of DF are not precisely described in the literature. Therefore, we present a case of DF arising from the scapula that was incidentally identified by FDG-PET imaging performed to evaluate the presence of metastasis due to a history of endometrioid adenocarcinoma.

Case report

A 65-year-old woman with a history of endometrial cancer was admitted to the Department of Orthopaedic Surgery of Juntendo University Hospital for consultation about a bone lesion in the right scapula although she was asymptomatic. She underwent surgical treatment for endometrioid adenocarcinoma 8 years earlier. PET taken to detect possible metastasis revealed moderate focal ^{18}F-FDG uptake in the right scapula with a maximal standardized uptake value (SUV) of 3.2 (**Figure 1A**); the lower angle of the scapula was unclear on plain radiology (**Figure 1B**). CT and 3-dimensional CT revealed a 61 × 42 × 27-mm osteolytic lesion with destruction of the cortical bone and that the rim of the scapula had partially disappeared (**Figure 1C**, **1D**). MRI demonstrated the presence of a mass with low and partially high intensities on T1- and T2-weighted images, respectively (**Figure 2A**, **2B**). The mass intensities on short tau inversion recovery did not change (**Figure 2C**, **2D**), and the tumor was partially enhanced by gadolinium (**Figure 2E**, **2F**). Needle biopsy was performed to make a differential diagnosis between primary bone and metastatic tumor. Pathologically, the tumor was composed of a relatively sparse proliferation of spindle-shaped fibroblastic/myofibroblastic cells in a dense collagenous background with focal myxoid change, leading to a diagnosis of a primary fibrous bone tumor suggestive of DF (**Figure 3A**, **3B**). Wide excision was performed considering the possibility of low-grade fibrosarcoma. On the cut surface of the resected tumor, the tumor clearly destructed the cortex of the scap-

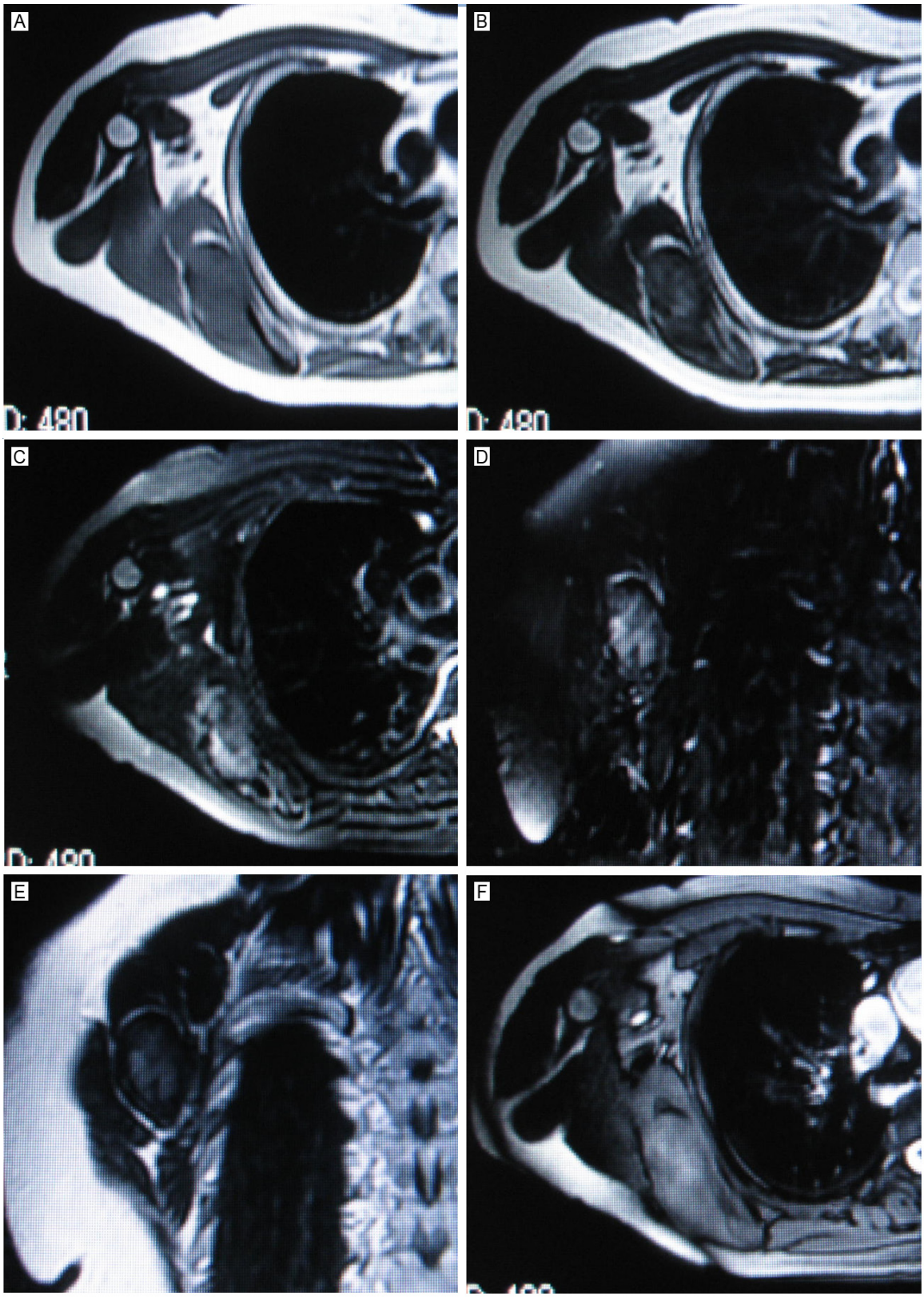

Figure 2. A, B: Magnetic resonance imaging showing a mass with low and partially high intensities on T1- and T2-weighted images, respectively; C, D: Mass intensities did not change on short tau inversion recovery; E, F: Partial enhancement of the tumor by gadolinium.

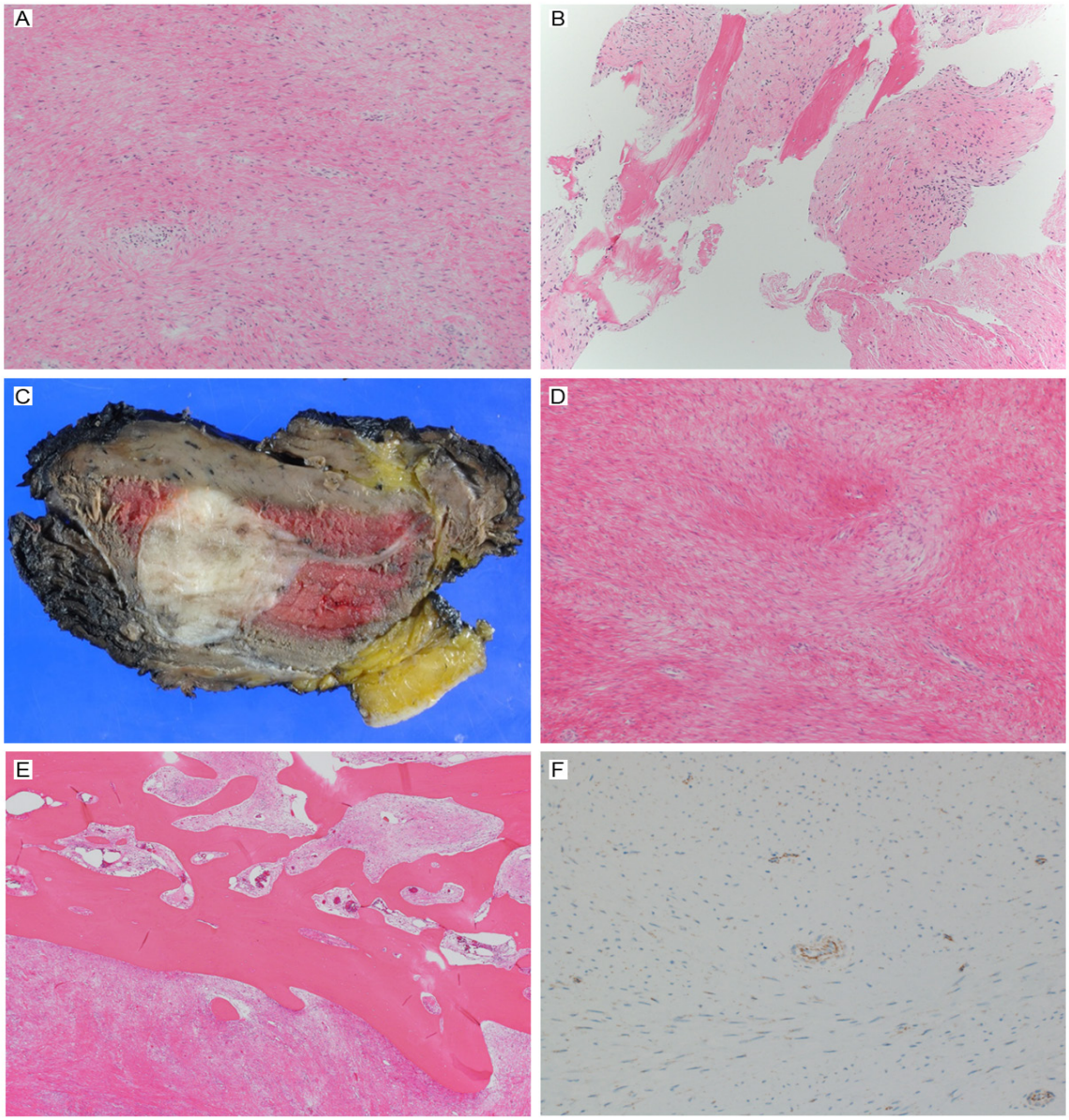

Figure 3. A: Biopsy specimen revealed that the tumor was composed of a relatively sparse proliferation of spindle-shaped cells in a dense collagenous background with focal myxoid change; B: Trabeculae of the lamellar bone engulfed within the collagenous background; C: Cut surface of the resected tumor clearly showing destructed scapular cortex and tumor expansion into the surrounding soft tissue; D: Tumor composed of a proliferation of less atypical spindle cells in the collagenous stroma. Mitotic figures are rare; E: Involvement of the tumor cells in the bone cortex; F: Tumor cells failing to show β-catenin nuclear/cytoplasmic stainin.

ula and expanded into the surrounding soft tissue (**Figure 3C**). Pathologically, the tumor was composed of a proliferation of less atypical spindle cells in the collagenous stroma, and no mitotic figures were observed (**Figure 3D**, **3E**). Immunohistochemically, β-catenin nuclear/cytoplasmic staining was not observed (**Figure 3F**), and no β-catenin genetic mutations were detected. A diagnosis of DF was made on the basis of these pathological and clinical findings. There were no signs of recurrence at the 6-month follow-up.

Discussion

DF is a rare primary bone tumor first described by Jaffe in 1958 [7]. Although any bone can be affected, DF arising from the scapula is extremely rare. To our knowledge, only 5 cases of DF affecting the scapula including the pres-

Table 1. Summary of reported cases of desmoplastic fibroma in Scapla

author	age	sex	clinical symptoms	size (mm)	Radiographic findings	treatment	Follow-up/outcome
Bertoni et al. [6]	43	F	swelling	-	-	curettage	NED/23 years
Nilsonne et al. [7]	16	M	pain, dysmotility of shoulder	-	osteolytic, destruction	curettage	recurrence after 10 years/18 years
Nilsonne et al. [7]	71	F	pain	-	trabeculated cysts	total excision	NED/4 years
Inwards et al. [22]	-	-	-	-	-	-	-
Present case	65	F	none	61 x 42 x 27	osteolytic, destruction	wide excision	NED/6 months

ent case have been published in the literature (**Table 1**) [6-9]. The chief complaints were pain in 2 cases, and swelling and dysmotility of the shoulder in 1 case each. Tumor size was not described in any of the previously reported cases. Major radiographic findings were osteolytic and destruction of cortical bone. Wide excision was performed in 1 case. Among the 2 cases treated with curettage, recurrence occurred in 1 case 10 years after surgery. The rate of local recurrence in cases treated with curettage or intralesional resection is at least 40% [3, 8, 10, 17]. Some authors regard this tumor's aggressive behavior as ''borderline'' or ''semimalignant'' rather than benign [2, 3, 17]. There is a consensus that the best treatment for DF is total excision [3, 8, 17]. There was no sign of local recurrence in the present case 6 months after wide excision.

Radiologically, the major features of DF include osteolytic lesion with destruction of the cortical bone, marginal sclerosis, and invasion into the surrounding soft tissue without periosteal reaction. CT and MRI are effective for evaluating the degree of invasion and bone destruction and useful for planning surgery. However, these findings are indistinct from those of other bone tumors such as fibrous dysplasia, hemangioma, eosinophilic granuloma, central low-grade osteosarcoma, and metastatic tumors. Recent literature supports the role of PET/CT for assessing disease response to treatment and detecting disease recurrence in a range of solid tumors [18-20]; PET and PET/CT are commonly used to detect recurrence or metastasis as well as search for unknown primary cancer. PET and PET/CT imaging has been used in musculoskeletal oncology to evaluate tumors [12-16]. Although ^{18}F-FDG PET is widely used to evaluate various tumors, recent reports suggest it cannot be used as a screening method for the differential diagnosis between benign and malignant musculoskeletal lesions. High ^{18}F-FDG accumulation can be observed in histiocytic, fibroblastic, and some neurogenic lesions regardless of malignancy. Thus, the use of ^{18}F-fluoro-α-methyltyrosine PET in combination with ^{18}F-FDG or ^{11}C-choline PET may be useful for distinguishing benign lesions from malignant tumors as well as preoperative planning in patients with musculoskeletal tumors [12]. Desmoid tumors, which are considered soft-tissue counterparts of DF, exhibit focal intensive uptake in ^{18}F-FDG PET [12, 15]. The average SUV in the 5 published cases is 3.04 [12, 15]. However, the ^{18}F-FDG PET findings in DF have not been documented so far, although DF is predicted to exhibit some uptake because of the histological similarity to desmoid tumors as observed in the present case. Despite the histological features revealed by the biopsy sample, the uptake still made us consider the possibility of a malignant tumor.

In particular, the important differential diagnosis in the present case involved low-grade fibrosarcoma of the bone and bony invasion of desmoid tumor [1]. Typical fibrosarcoma is more cellular and exhibits a herringbone pattern with more pleomorphism and higher mitotic activity [21]. However, determining malignancy by biopsy using conventional microscopic findings alone is not always easy.

Immunohistochemistry for β-catenin is useful for differentiating DF from desmoid tumors. Of 14 reported cases of DF, 7 were immunohistochemically positive for β-catenin in more than 10% of cytoplasm or nucleus [10, 22]. Although this appears to be concordant with desmoid-type fibromatosis, desmoid tumors generally exhibit diffuse β-catenin nuclear accumulation

[23, 24]. Furthermore, β-catenin (CTNNB1) mutations are reported very frequently [25-28], although they are absent in DF [22]. These findings indicate that β-catenin plays a rather important role in the tumorigenesis of desmoid tumors. Furthermore, the detection of β-catenin mutations may be a specific diagnostic tool for the diagnosis of this tumor [27, 28]. β-catenin (CTNNB1) mutations and β-catenin accumula tion were not detected in the present case, further corroborating the diagnosis of DF.

In summary, we experienced a highly unusual case of DF arising from the scapula. DF should be considered a benign bone tumor that exhibits ^{18}F-FDG uptake on PET.

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Disclosure of conflict of interest

The authors have no conflicts of interest related to this article to declare.

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