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Metastatic Bednar tumor (pigmented dermatofibrosarcoma protuberans) with fibrosarcomatous change: a case report

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Abstract Bednar tumor (pigmented dermatofibrosarcoma protuberans) is a variant of dermatofibrosarcoma protuberans (DFSP) that constitutes 5% of all DFSP and has a very low rate of distant metastases. We encountered a rare case of Bednar tumor with multiple different distant metastases. A 51-year-old man, who had had a history of mass resection in his left shoulder 4 years previously, was referred to our institution, complaining of a recurrence of the shoulder mass. The histological diagnosis of primary tumor was Bednar tumor, and he underwent resectional surgery for the recurrent lesion. A second local relapse, lung metastasis, retroperitoneal metastasis, and metastasis to the sigmoid colon have occurred. The recurrence lesion and all metastatic lesions were resected surgically. The histological features of all specimens showed fibrosarcomatous change. Seven months after the last surgical resection, the tumor recurred and the patient died of multiple abdominal metastases 10 years after the first surgical treatment.

Key words Bednar tumor · Pigmented dermatofibrosarcoma protuberans · Fibrosarcomatous change · Multiple metastasis

Introduction

Bednar tumor is a rare pigmented neoplasm, featuring locally aggressive tumor. In 1957, Bednar first described nine cases of cutaneous neoplasm showing a histological pattern similar to dermatofibrosarcoma protuberans (DFSP) containing melanin pigmentation.[1] Because he termed this tumor storiform neurofibroma, it has not been considered as a variant of DFSP associated with the production of melanin pigment for a certain period. Clinically, both DFSP and Bednar tumor are considered to be of the same category and of intermediate malignancy because of their tendency for frequent local recurrence and lack of distant metastasis.[9] Moreover, their clinical and gross features are similar except for melanin pigmentation. However, only a few reports have described distant metastasis of Bednar tumor.[2,4,5,6,8] We encountered a rare case of Bednar tumor that was exhibiting distant metastasis and fibrosarcomatous change.

Case report

In 1998, a 51-year-old man who had recurrent swelling on his left shoulder was referred to our institution. He had a previous history of simple excision of a cutaneous lesion on the left shoulder at a local hospital in 1994, and the pathological diagnosis at that time was dermatofibrosarcoma protuberans (DFSP). On physical examination, a 4-cm protruded mass on the left shoulder was observed (Fig. 1a). A magnetic resonance imaging (MRI) scan showed a 8-cm subcutaneous mass (Fig. 1b). When primary material that had been resected at a local hospital was reviewed, the specimen showed a storiform pattern consisting of melanin pigment cells, and iron stain was negative (Fig. 2a,b). On the basis of these findings, the diagnosis of Bednar tumor was made.

The patient was treated with surgical resection with a wide margin. The lesion involved the subcutaneous tissue. On pathological examination, this tumor was composed of spindle-shaped cells with oval and elongated nuclei. Although some storiform pattern was observed, cellularity was increased and a herringbone pattern was also seen (Fig. 3a). However, melanin pigmentation was not observed, and mitotic activity revealed an average of 7.1 mitotic figures/10 high-power field (HPF). On immunohistochemical examination, positive reactions for vimentin and CD34 were consistently evident in the tumor cells of the recurrence lesion (Fig. 3b). Other markers such as S-100 protein, desmin,

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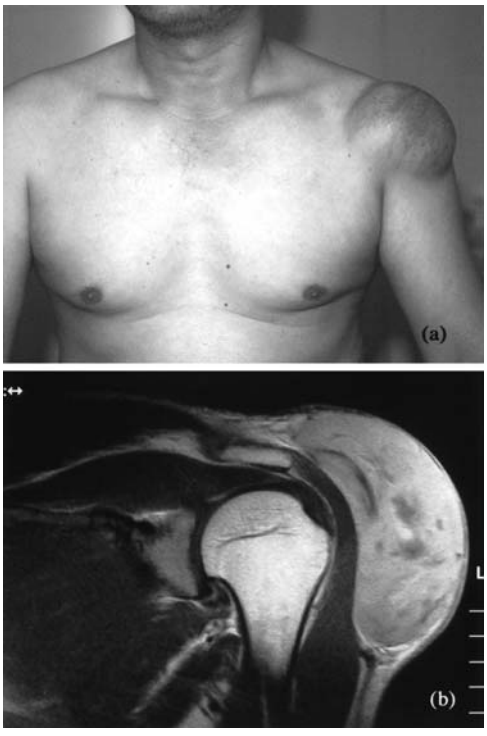

Fig. 1. **a** In the first local recurrence, the patient's shoulder mass measured about 4 cm. **b** Coronal magnetic resonance imaging (*MRI*) scans of the left shoulder. On T_1-weighted images with Gd enhancement, MRI scans revealed a homogeneous high-intensity mass over the deltoid muscle

SMA, and CD117 showed negative reactions. The pathological feature revealed was that of DFSP with fibrosarcomatous change. Although it did not show any pigmentation, it was quite possible that this is a local recurrence of Bednar tumor.

The patient did well until February 2002, when he noticed a subcutaneous mass on his left axillary fossa. In addition, a chest X-ray showed a coin lesion in the upper lobe of the left lung. A CT scan revealed 3-cm and 5-mm nodules extending into S3–S4 and S6, respectively, in the left lung (Fig. 4a,b). We regarded the mass of the left axillary fossa as a recurrence lesion and the nodules in the left lung as metastasis lesions. Subsequently, the patient received two courses of chemotherapy with ifosfamide, carboplatin, and etoposide that resulted in no change of the tumor size. The tumor of the axillary fossa was consequently resected with a wide margin. The skin defect after the resection was covered with a skin flap and a subsequent skin graft taken from the left thigh. For treatment of the lung metastasis, segmentectomy of the metastatic lesions in the left lung was performed. Pathological examination showed features that were similar to fibrosarcoma, and the diagnosis of the lesions was recurrence and metastatic Bednar tumor, respectively (Fig. 5a,b).

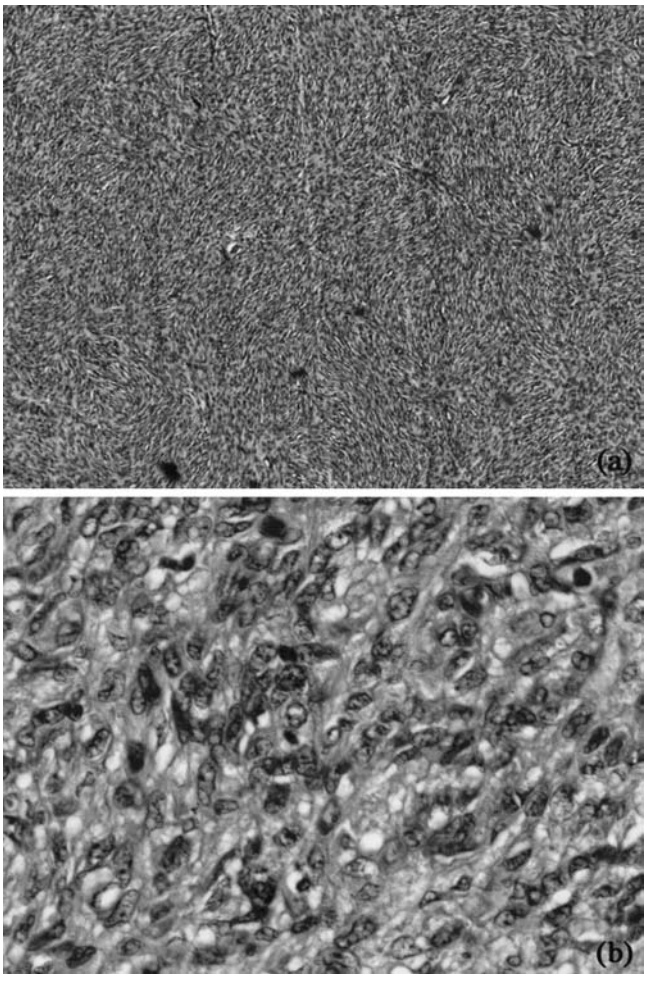

Fig. 2. **a** On histological section of the primary surgical material, striform patterns were observed frequently. H&E, ×100. **b** On high-power view of the primary surgical material, pigmented cells were observed. H&E, ×400

After surgical resection, the patient underwent radiotherapy (50 Gy) and had no symptoms for a while. In August 2002, when he was again admitted to our institution, he had acute abdominal pain and presented with shock status. In an abdominal echo, we recognized a mass and hemorrhage of the pancreas head. A CT scan revealed a retroperitoneal mass in the pancreas head and a large hematoma due to a hemorrhage from the pancreas (Fig. 4c). He was treated with pancreatoduodenectomy; the pathological diagnosis of the surgical specimen, which showed fibrosarcomatous change, was metastasis of the Bednar tumor. In December 2002, he was recognized with hematochezia, and a CT scan revealed a 5-cm mass that extended into the sigmoid colon (Fig. 4d). We regarded the mass as a metastatic Bednar tumor to the sigmoid colon. Hence, sigmoidectomy was performed and the pathological diagnosis of metastasis was confirmed. After 7 months, the

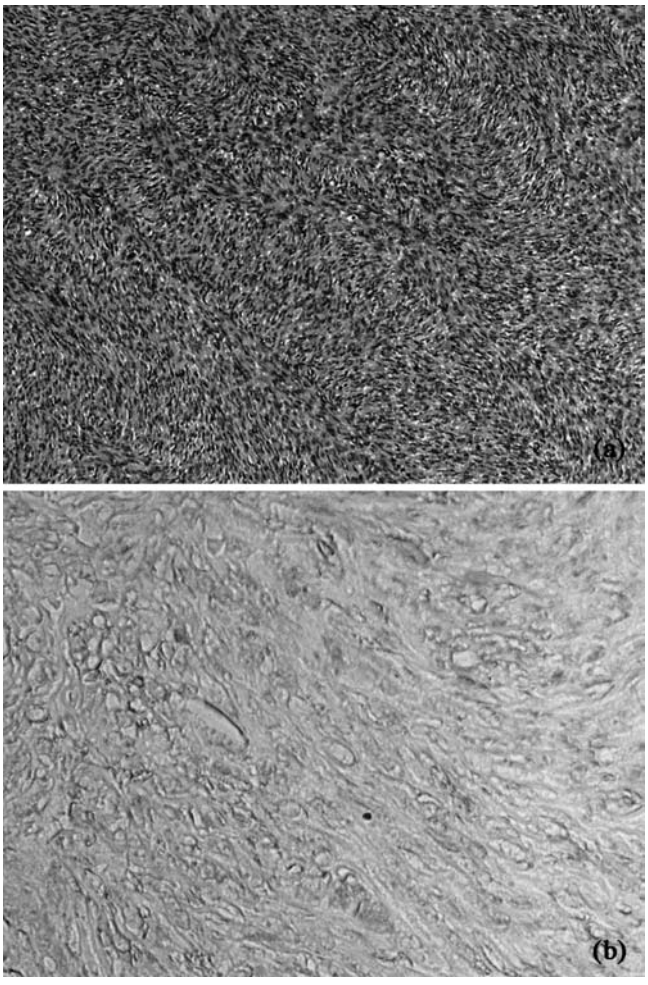

Fig. 3. **a** Histological section of the first recurrence surgical material shows a herringbone pattern. H&E, ×100. **b** CD34 immunoreactivity within the first recurrence surgical material. Diffuse staining is noted in most neoplastic cells. CD34, ×300

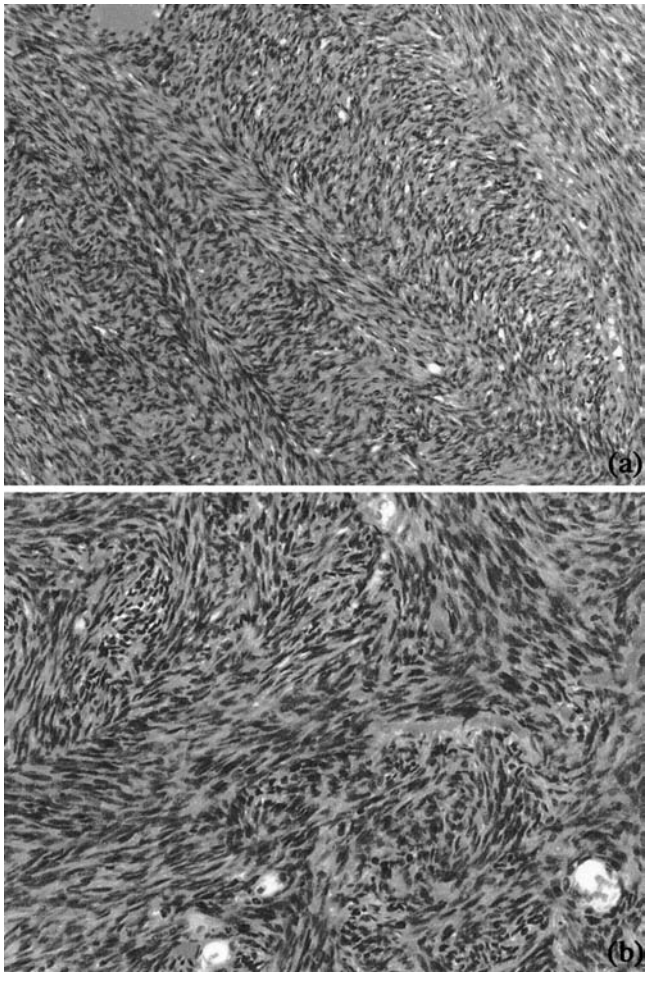

Fig. 5. **a** A second recurrence tumor on the left axillary fossa showed high cellularity, and the cells appeared more mature and exhibited a herringbone pattern consisting of spindle cells. H&E, ×125. **b** The left lung metastasis tumors showed hypercellular and fibrosarcomatous features that exhibited atypical cells. ×200

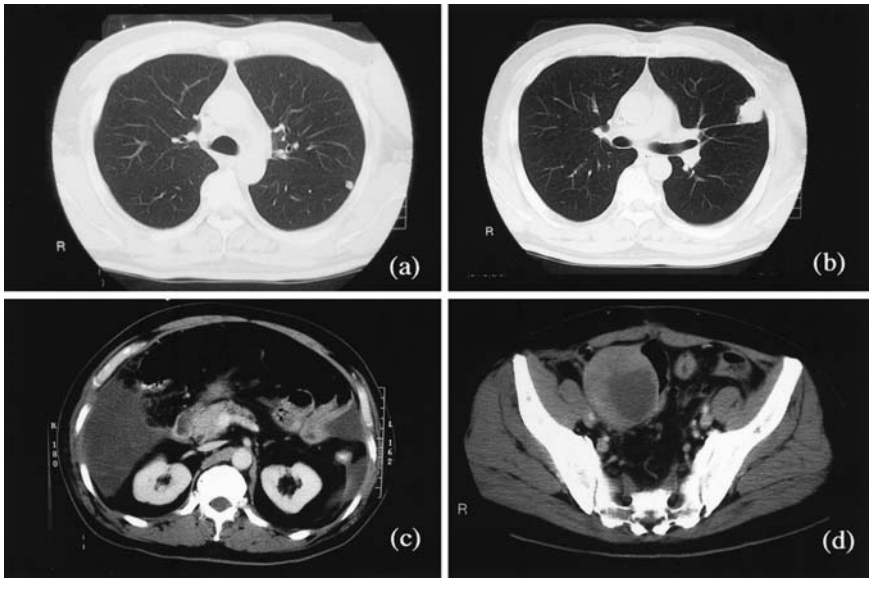

Fig. 4. **a** Computed tomography (*CT*) scan of the chest revealed a 5-mm nodule extended into S6. **b** CT scan of the chest shows a 3-cm nodule extended into S3–S4. **c** CT scans of the abdomen reveal a retroperitoneum mass involving the pancreas head and a large hematoma. **d** CT scan of the pelvic region shows a large mass extending into the sigmoid colon

tumor recurred, and the patient died of multiple abdominal metastases 10 years after the first surgical treatment.

Discussion

The incidence of Bednar tumor is quite low, accounting for less than 5% of all DFSP.[4,5,9] Bednar tumor usually grows as a nodular cutaneous tumor characterized by a prominent storiform pattern and melanin pigmentation. It typically presents during middle adult life as a nodular cutaneous mass, in males and females equally. The most frequent sites are the trunk and proximal extremities. The shape of the tumor tends to be well demarcated, with gray-white, protuberant masses involving the dermis and subcutaneous layers. In some cases lesions appeared gray or black owing to the amount of melanin present.[4,5,9]

Regarding the local recurrence rate and distant metastasis rate of Bednar tumor, Dupree et al. reported, in 20 cases of Bednar tumor, that local recurrence occurred in 11% and distant metastasis in none of the cases.[5] Ding et al. described in 45 cases that the local recurrence rate was 13%, with no cases of distant metastasis.[4] Moreover, the low incidence of local recurrence from Bednar tumor has been compared with the incidence from conventional DFSP (20%–50%), and Bednar tumor has been described as infrequently metastatic.[3,7,9]

There have been a few reports on metastatic Bednar tumor.[2,6,8] These papers described the histological features of metastatic Bednar tumor, which showed less storiform pattern and prominent fibrosarcomatous features with a herringbone or interlacing arrangement. All cases reported were associated with fibrosarcomatous change.

In our case, the primary tumor had some pigmented cells, which were that of typical Bednar tumor features, but the recurrence and metastatic tumors lost the pigmented cells. Although part of the first recurrence tumor on the patient's shoulder showed the typical features of conventional DFSP, most parts of the first recurrence tumor, the second recurrence tumor, and metastatic tumors did not show the storiform pattern. Instead, they acquired fibrosarcomatous features with a herringbone or interlacing arrangement. We concluded that these local recurrence and metastatic tumors were exhibiting fibrosarcomatous change.

Up to now, only four cases, including our case, of pigmented dermatofibrosarcoma protuberans with distant metastasis have been reported in the English literature and one case in Italian.[2,6,8] Those four patients were 45, 46, 51, and 75 years old, three men and one woman. The predominant locations of primary tumor were the shoulder, the upper arm, the foot, and the thigh. The locations of distant metastasis were lung (three), retroperitoneal space (one), sigmoid colon (one), brain (one), bone (one), soft tissue (one), and skin (one). The prognoses were as follows: three cases died (2 years, 7 years, and 10 years after first treatment) and one case was not available. On histological examination, the primary tumor features were of typical Bednar tumor (three) and fibrosarcomatous Bednar tumor (one). In the three cases of typical Bednar tumor, all metastatic materials showed fibrosarcomatous change and only one example presented melanin pigmentation; two were not observed. In addition, we recognized that the metastatic Bednar tumor has a tendency to present fibrosarcomatous change, and speculate that a Bednar tumor with fibrosarcomatous change exhibits more malignant behavior than a typical Bednar tumor.

In summary, we encountered a rare case of metastatic Bednar tumor with fibrosarcomatous change. All reported cases that showed distant metastasis also consistently showed fibrosarcomatous features. It seemed that a Bednar tumor with fibrosarcomatous change tends to a more aggressive clinical course than typical Bednar tumor, same as fibrosarcomatous change in DFSP.

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