

## Multicentric extra-abdominal desmoid tumors arising in bilateral lower limbs

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### Abstract

Extra-abdominal desmoid tumors preferentially affect the shoulders, arms, backs, buttocks, and thighs of young adults. Multicentric occurrence is rather rare but seems to be another distinctive feature of extra-abdominal desmoid tumors. In this article we report a rare case of multicentric extra-abdominal desmoid tumors arising in bilateral lower limbs.

### Introduction

Desmoid tumors are benign neoplasms that display local aggressiveness but no metastasis. They are classified according to their location; namely, whether they are located at extra-abdominal, abdominal wall, or intra-abdominal sites. This type of tumor is relatively rare, only occurring in 2-3/10<sup>6</sup> people.<sup>1,2</sup> Its extra-abdominal form occurs in the shoulder, upper arm, and thigh, and more rarely in the fascia of the lower arm, pelvis, or knee.<sup>3</sup> All previously reported cases of multicentric extra-abdominal desmoid tumors were present in the same extremity.<sup>2,4,6</sup> We report here on multicentric extra-abdominal desmoid tumors arising from the popliteal fossa of bilateral lower limbs and the appropriate treatment for patients presenting with such tumors.

### Case Report

A 64-year old female was referred to our hospital in December 1991. She had noticed an elastic hard mass in her right popliteal fossa for a period of three years, but recently it had grown in size. She had a history of hypothyroidism, but there was no particular incidence of trauma or any relevant medical history. Magnetic resonance imaging (MRI) identified the mass at the distal extent of the gastrocnemius fascia in the right popliteal fossa (Figure 1). A photomicrograph of a biopsy sample showed elongated spindle-shaped cells of uni-

form appearance and abundant collagen fibers showing a wave-like arrangement. The mass was diagnosed as an extra-abdominal desmoid tumor (Figure 2A). A surgical procedure to remove the tumor was performed. Post-operative radiotherapy was carried out with a total dose of 50 Gy to prevent recurrence.

Three years later, a new mass was seen in the left popliteal fossa (Figure 3A and B). To diagnose the tumor, open biopsy was performed again. The tumor was characterized by a proliferation of spindle-shaped cells without nuclear pleomorphism or hyperchromasia, the same as the desmoid tumor in the opposite limb (Figure 2B). This tumor was excised, followed by no additional therapy. However, next year when the patient was 68-years old, a recurrence developed with an extension surrounding the peroneal nerve in the popliteal fossa (Figure 3C and D). The tumor was partially excised because the peroneal nerve traversed the lateral part of the tumor. In addi-

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tion, the patient had a left femoral neck fracture and underwent internal fixation using an intramedullary nail at the age of 75 years (Figure 4A). One-and-a-half years after this

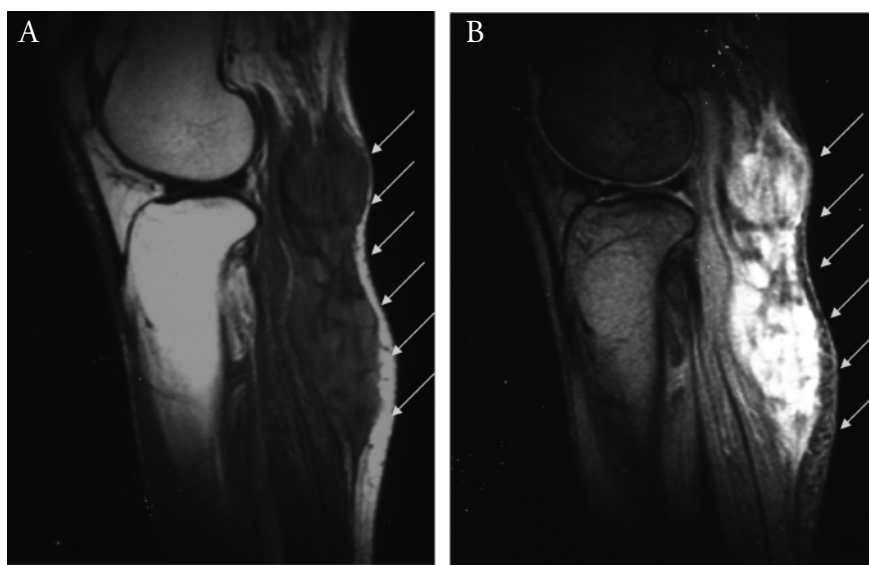


Figure 1. First preoperative magnetic resonance imaging (MRI) scan of a tumor in the right gastrocnemius fascia. MRI demonstrated that the tumor (arrows) had low signal intensity on a T1-weighted image (A) and low and high signal intensity on a fat-suppressed T2-weighted image (B).

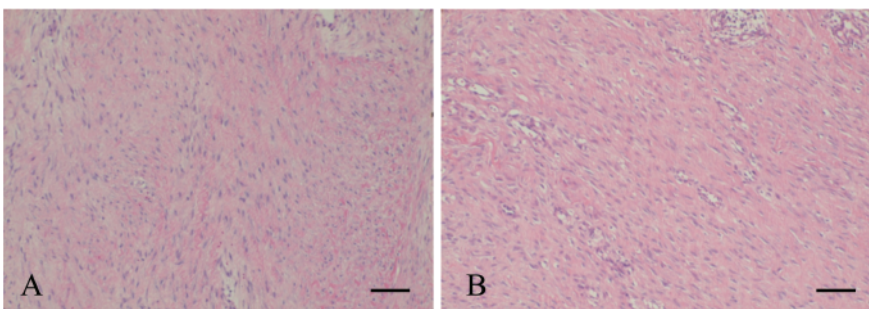


Figure 2. Characteristic uniform spindle-shaped cells and collagen fibers typical of the histological picture of the desmoid tumor. The histological findings of tumors harvested from the right (A) and the left (B) popliteal fossae indicated desmoid tumors. (Hematoxylin and eosin stain; bar, 100  $\mu$ m).

trauma, a new tumor appeared on the posterior aspect of her left thigh (Figure 4B and C). Histological examination revealed a desmoid tumor, and this tumor was excised with the normal tissue containing a 1- to 2-cm-thick layer of muscle. A recurrent tumor developed and surrounded the sciatic nerve one year later. This tumor was also excised intra-lesionally. However, because the desmoid tumor on the posterior aspect of the left buttock redeveloped soon thereafter, radiation with a total dose of 30 Gy was given to suppress pain and tumor growth. After that, the size of the tumor was stable, and the intensity of pain was decreased; to date, no other tumor-like lesions have been seen. The patient and her family provided written informed consent that the data from the case could be submitted for publication.

### Discussion

Ten to twenty multicentric extra-abdominal desmoid tumors have been reported previously.<sup>2,4-11</sup> They are observed mostly in connection with Gardner's syndrome implying familial occurrence, intestinal polyposis, and multiple mesodermal tumors such as in fibromas, desmoids, and leiomyomas.<sup>12,13</sup> The desmoid tumors usually affect the same sites and the same extremities but do not grow simultaneously; second growths appear years later and develop in close proximity to the primary lesion.<sup>14</sup> Therefore, it was considered that multicentric development was a result of extension from the primary site. In this case, both the popliteal lesions consisted of elongated spindle-shaped cells of uniform appearance and abundant collagen fibers, showing a typical histological picture of the desmoid tumor; consequently, we concluded that these tumors were multicentric extra-abdominal desmoid tumors arising in bilateral lower limbs. The case described in the present report is extremely rare (Figure 5), and to our knowledge there are no similar reports even in familial cases of desmoid fibromatosis.

The cause of the sporadic incidence in both limbs in this case may not be regional spread of the tumors, but rather mutation of proto-oncogenes in the developing limb bud.<sup>4</sup> With regard to endocrine agents, desmoids are associated with an increased number of estrogen receptors and anti-estrogen binding sites.<sup>15,16</sup> Serpell *et al.* reported that estrogen receptors have been identified in 22-33% of desmoid tumors,<sup>17</sup> but Sorenson *et al.* found no estrogen receptor defects among 72 patients.<sup>18</sup> In our case, estrogen receptor antibody immunohistochemistry was also negative. Furthermore, the expression of thyrotropic hormones was examined owing to a history of hypothyroidism,

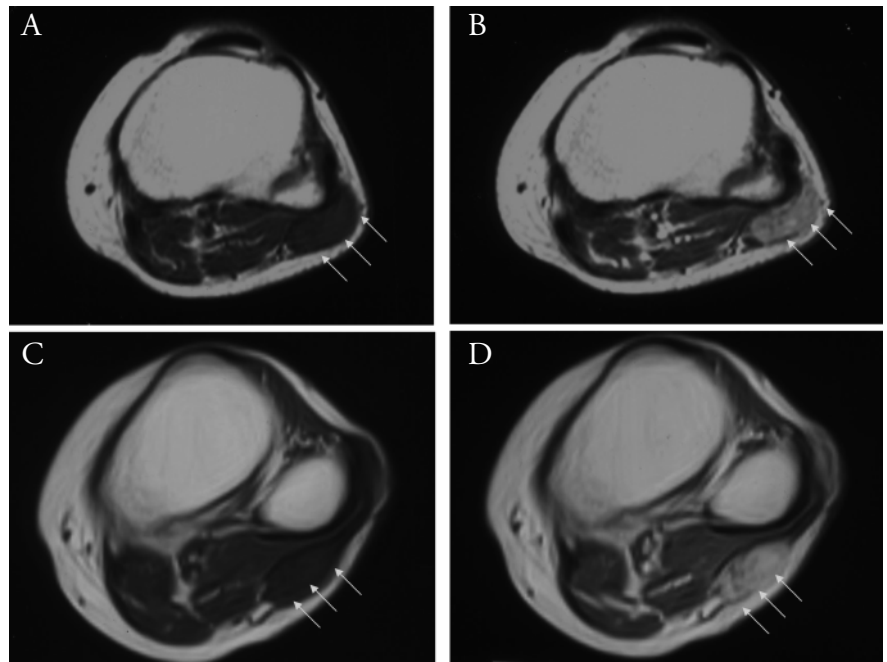


Figure 3. MRI scan of the tumor in the left popliteal fossa lesion. The tumor (arrows) revealed low signal intensity on T1-weighted image (A) and enhancement by gadolinium (B). The recurrent tumor (arrows) was attached to the left gastrocnemius fascia and surrounded the peroneal nerve on a T1-weighted image (C) and enhancement by gadolinium (D).

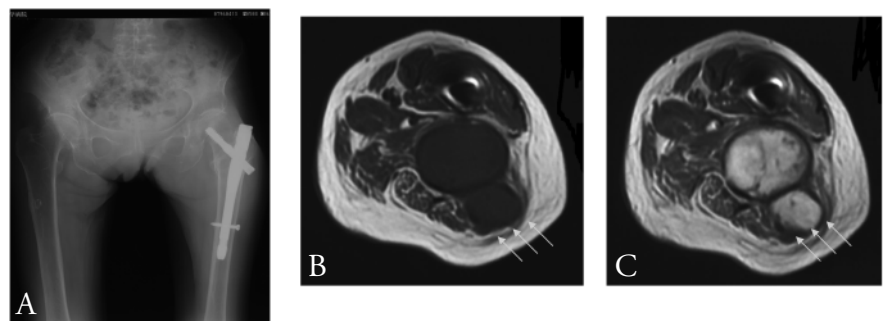


Figure 4. Postoperative radiograph of the left trochanteric femoral fracture (A). MRI scan of the new tumor in the posterior left thigh. The lesion (arrows) demonstrated low signal intensity on a T1-weighted image (B) and enhancement by gadolinium (C).

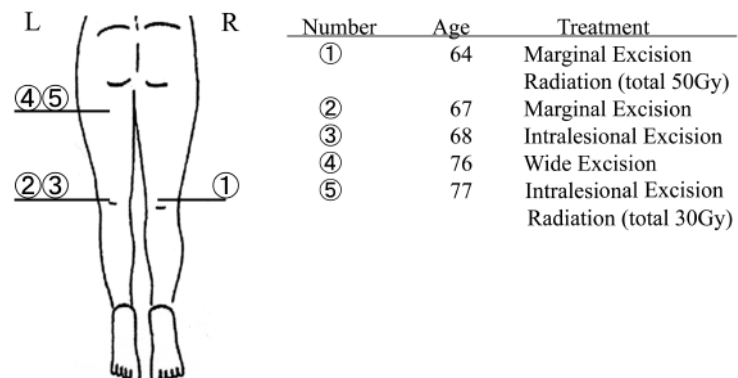


Figure 5. Scheme of the tumor distribution in our case.

but the counts for TSH, FT3, and FT4 were normal. This type of tumor is not known to metastasize but has a high recurrence rate after surgical excision. The risk of local recurrence appears to be most closely related to the extent of the surgical excision, with fewer recurrences in margin-negative than in margin-positive patients.<sup>2,18,19</sup> However, Merchant *et al.* and Gronchi *et al.* found that positive margins were not prognostically significant.<sup>20,21</sup> Concerning re-resection for the treatment of recurrent tumors, Reitamo *et al.* showed that the recurrence rate with resection after a second or third recurrence was equal to or lower than the recurrence rate after primary resection.<sup>15</sup> Posner *et al.* reported on 33 patients who underwent re-resection after local recurrence, and 28 of these patients (85%) remained free of disease at a median follow-up time of 70 months.<sup>22</sup> In our patient, therapeutic methods varied because the doctor in charge of the patient changed during the long period of treatment. Therefore, the surgical margins in each surgery varied and radiation therapy was performed postoperatively after the tumor resection in the right lower extremity but was not done after the resection in the left lower extremity. As a result, the recurrence in both lower limbs was controlled by surgical treatment and adjuvant radiation even though intralesional excision was performed. Thus, the surgical margin cannot be considered the only risk factor for local recurrence. Concerning radiation therapy, high disease-free survival rates have been demonstrated especially in cases where doses of 50-60 Gy external radiation therapy were administered after surgery.<sup>19,23</sup> In our case, the desmoid tumor in the right popliteal fossa that underwent postoperative radiation had no recurrence, and the growth of the desmoid tumor in the left buttock was also suppressed after radiation. On the other hand, radiation-induced side effects such as edema, fibrosis, ulcers, pathological fractures, and secondary malignancies have been reported previously.<sup>24</sup> Radiation therapy has its difficulties from the viewpoint that desmoid tumors are benign. An unusual feature of desmoid tumors is their ability to regress or cease growing naturally. Rock *et al.* indicated the efficacy of observation to control desmoid tumors.<sup>2</sup> The role of surgical and adjuvant treatment and their effects on local control for long-term care need to be evaluated.

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