CASE REPORT

Recurring facial low-grade fibromyxoid sarcoma in an elderly patient : A case report

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Abstract : Low-grade fibromyxoid sarcoma (LGFMS) is a rare type of sarcoma that is characterized by benign-appearing histologic features but a paradoxically aggressive clinical course. These tumors generally occur in young to middle-aged adults, sometimes in children, but rarely in high-aged adults. LGFMS typically affects the deep soft tissues of the trunk or lower extremities ; however, it is rarely seen in the face. We here describe a case of LGFMS on the right forehead of an 84-year-old woman. After resection with a 1-cm skin margin, recurrence occurred at 15 months postoperatively. Additional wide excision was subsequently performed with a 2-cm skin margin. Recurrence and metastasis have not been observed for 1 year after the second excision. A wide surgical margin should be considered in cases of LGFMS. J. Med. Invest. 59 : 266-269, August, 2012

Keywords **:** *low-grade fibromyxoid sarcoma, elderly patient, forehead, recurrence, wide excision*

INTRODUCTION

Malignant soft tissue tumors rarely occur in the head and neck, and surgeons find it difficult to determine the optimal method for excising these tumors, particularly those located in the face. Malignant soft tissue tumors are generally resected along with thick surgical barriers, such as ligaments or the periosteum. However, resection involving such barriers is difficult, due to the serious risks posed when the tumor to be resected is situated adjacent to important organs in the face. Moreover, accurate diagnosis of soft tissue tumors is often complicated even for well-trained pathologists due to the histologic characteristics of these tumors. Low-grade fibromyxoid sarcoma (LGFMS) is a rare type of sarcoma ; determination of the disease course based on oncologic and histologic features remains controversial (1-3). Moreover, the efficacy and advantages of adjuvant therapy for this neoplasm have not been elucidated. We describe here the case of an elderly woman with recurring facial LGFMS that occurred at a rare location and an unusual age.

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CASE REPORT

An 84-year-old woman noticed a subcutaneous, gradually enlarging tumor on her face. The tumor was firm and immobile, approximately 3 × 2 cm in size, and located on the right forehead (Fig. 1). Magnetic resonance imaging (MRI) revealed that the mass was connected to the deep temporal fascia, appearing as a low-intensity area on T1 images and a high-intensity area on T2 images (Fig. 2 a, b). No metastasis was noted on computed tomography. Based on MRI findings, the mass was found to resemble nodular fasciitis or a tumor such as a hemangioma or fibroma. We performed an excisional biopsy to confirm the diagnosis. Histologic examination of the biopsy specimen revealed fibrous and myxoid components (Fig. 3 a, b). The fibrous parts

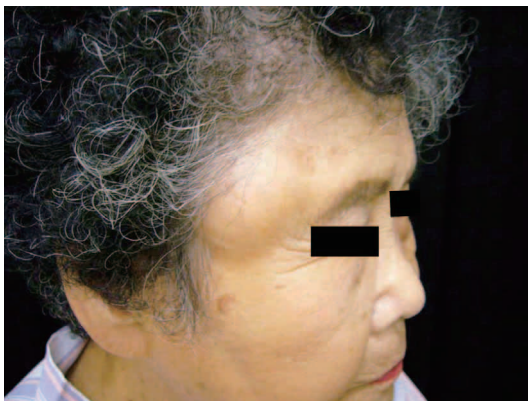
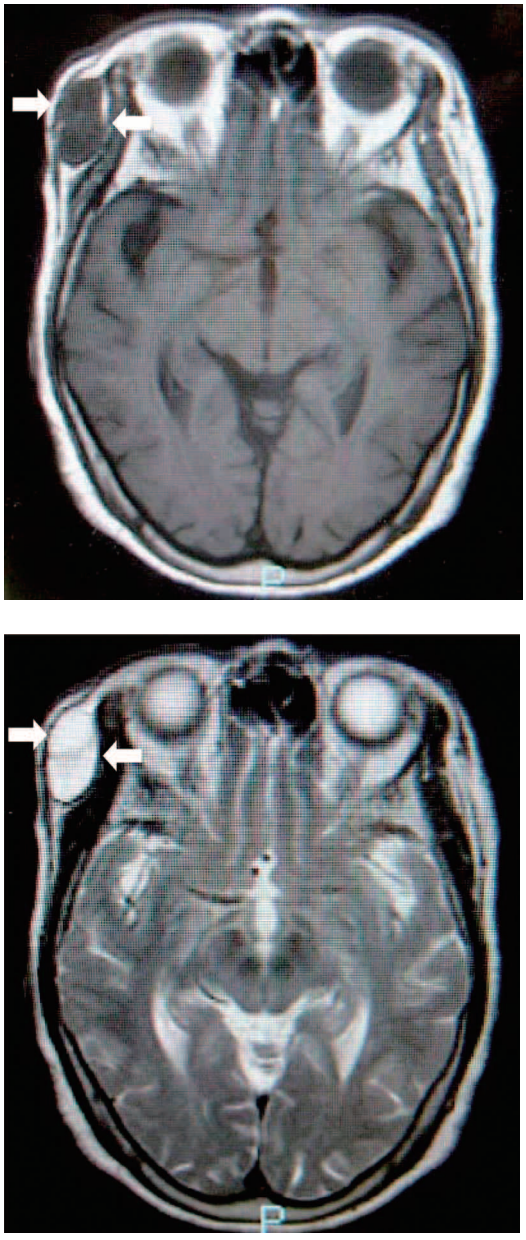


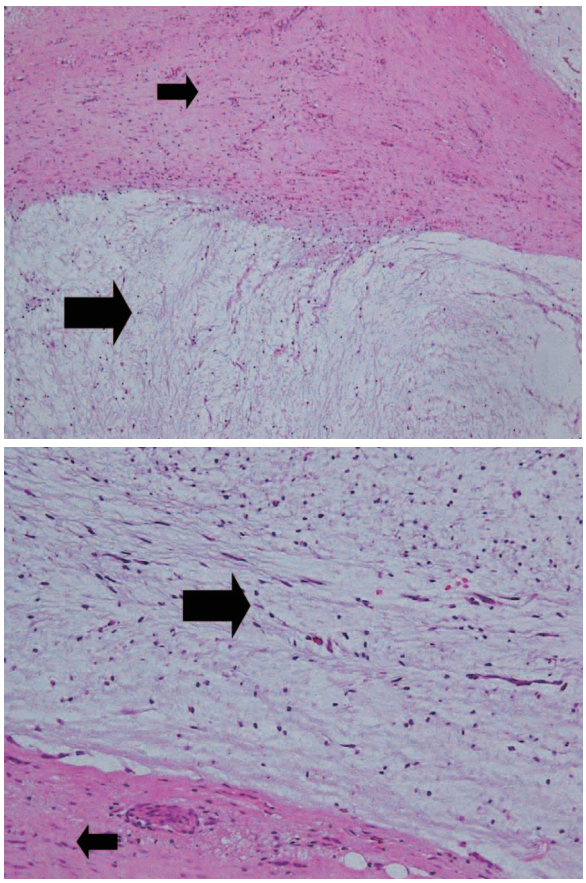
Figure 1 : Photograph showing subcutaneous tumor on the right forehead. It was approximately 3 × 2 cm in size.



(a)

(b)

Figure 2 : (a) MRI revealed the mass (arrows) in the subcutaneous tissue, appearing as a low-intensity area on T1 images. (b) The mass (arrows) was showed as a high-intensity area on T2 images.



(a)

(b)

Figure 3 : (a) The tumor composed of fibrous (small arrow) and myxoid (large arrow) components. (heamatoxylin and eosin stain, ×100) (b) The spindle-shaped cells (small arrow) were found in the fibrous parts, the spindle to stellate-shaped cells (large arrow) with an abundant intercellular matrix in the myxoid parts. (heamatoxylin and eosin stain, ×400)

comprised spindle-shaped cells in a linear arrangement showing whorled and swirling growth patterns, whereas the myxoid parts comprised spindle to stellate-shaped cells with an abundant intercellular matrix and, in some parts, relatively rich vascular networks. The tumor cells contained oval or short spindle-shaped nuclei without a high degree of atypism or pleomorphism. Immunohistochemical examination revealed diffuse positivity to anti-vimentin antibody, whereas tests for other antibodies were negative (Table 1). Based on the histologic findings, including the absence of nuclear atypism or pleomorphism, and the presence of predominant fibrous components, we diagnosed the tumor as LGFMS, not myxofibrosarcoma. Although we recommended an additional wide excision, the patient refused. One year later, the tumor recurred at the same location. We had previously resected the recurrent tumor with a 1-cm skin margin, including the temporal branch of the facial nerve and the temporal muscle, and reconstructed the defect with the remaining

Table 1. The results for immunohistochemical staining

Positive	Negative
Vimentin	Desmin, α-SMA, CD-34, CD-56, S-100 protein, cytokeratin

temporal muscle flap and a skin graft (Fig. 4). At that time, we also completely dissected the tumor postoperatively under a microscope. Despite this approach, the tumor recurred 2 cm away from the skin graft 15 months after the first surgery. We performed additional surgical excision with a 2-cm skin margin, including the periosteum, and covered the defect with a local skin flap (Fig. 5). At present, 1 year after the second excision, there is no evidence of recurrence or metastasis of the LGFMS.

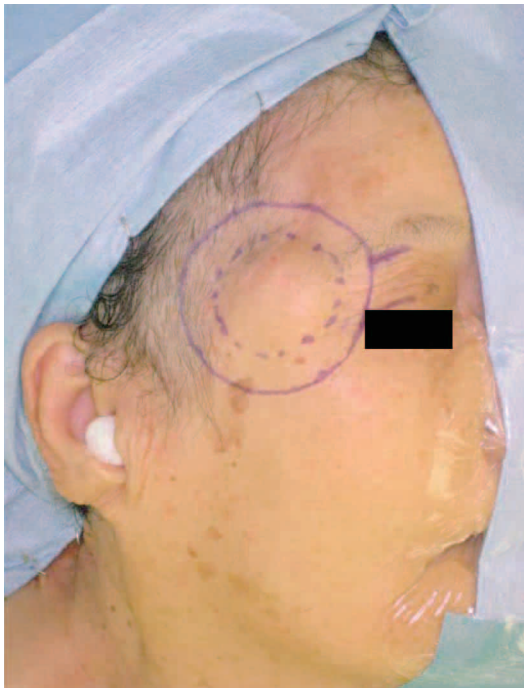


Figure 4 : One year later, LGFMS recurred at the same location. Surgical excision was performed with a 1-cm skin margin, and a skin graft was applied.

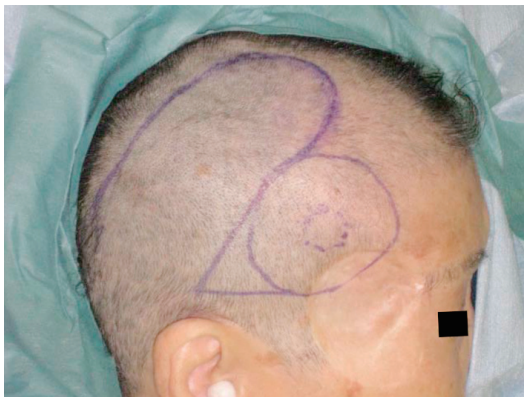


Figure 5 : Fifteen months later, re-excision was made with a 2-cm skin margin, and reconstruction with a local skin flap and a skin graft was performed.

DISCUSSION

LGFMS is a rare tumor type first described by Evans in 1987 (1). In the original description, LGFMS was histologically characterized as a bland-appearing soft tissue neoplasm, but the tumor was found to be associated with aggressive behavior and a high degree of local recurrence or distant metastasis. In the second paper, Evans reported that 9 of the 12 patients examined experienced local recurrence and that 7 who experienced distant metastasis died from LGFMS (2). However, Folpe *et al.* described that, of their 54 patients, local recurrence and distant metastasis were seen in 5 (9%) and 3 (3%) patients, respectively (3). In addition, the diagnosis of LGFMS or "sarcoma" was made at the initial presentation in 51 of these cases ; therefore, wide and adequate surgical excision could be performed. The authors also highlighted that early surgery was one of the reasons why the rates of local recurrence and distant metastasis in their series were much lower than those of previous reports. The most frequently reported location of LGFMS is the lower extremity, especially the thigh, followed by the trunk and groin, upper extremity, and buttock. The head and neck region is a very rare location for this sarcoma, as based on previous reports (1-5).

Histologically, LGFMS shows alternating fibrous and myxoid areas and a swirling and whorled growth pattern. The fibrous area contains deceptively benign-appearing fibroblastic cells, with low to moderate cellularity, uncommon mitotic figures, and absent or slight nuclear pleomorphism. The myxoid area comprises spindle to stellate-shaped cells with an abundant intercellular matrix. Furthermore, Evans described that some recurrent and metastatic tumors contain zones of increased cellularity and mitotic activity. In our case, we observed high cellularity in only some areas. However, further follow-up is necessary to determine whether the presence of these areas impairs the recurrence and metastatic rate of LGFMS. Immunohistochemically, most cells of this

sarcoma are strongly positive for vimentin antibody, but are generally negative for α-smooth muscle actin, desmin, S-100 protein, cytokeratin, CD34, and CD56 antibodies. In addition to these features, Zámečník and Michal described the presence of strong and diffuse bcl-2 reactivity (5), although we did not investigate bcl-2 reactivity in the present patient.

The differential diagnosis of LGFMS in terms of other malignant tumors chiefly includes malignant fibrous histiocytoma and myxofibrosarcoma. Typical malignant fibrous histiocytoma is considerably more cellular and has greater malignant potential than LGFMS, that is, it shows greater nuclear hyperchromatism, pleomorphism, and mitotic activity. Myxofibrosarcomas are classified into 4 grades, with grade 1 myxofibrosarcoma considered to resemble LGFMS. However, this tumor can be differentiated from LGFMS by the greater degree of nuclear atypia, predominant myxoid component, and absence of metastasis ; further myxofibrosarcoma generally occurs in older adults. Moreover, a t(7 ; 16)(q34 ; p11) translocation resulting in a *FUS/CREB3L2* fusion transcript has been reported in more than 95% of LGFMS cases when combinations of classic cytogenetic and molecular cytogenetic methods, such as reverse transcription polymerase chain reaction (RT-PCR) and fluorescence in situ hybridization (FISH), are employed. A rare *FUS/CREB3L1* variant has also recently been described in a small subset of LGFMS cases (6). Although we did not attempt to analyze these fusion genes, these methods may be valuable tools in the differential diagnosis (7).

Undoubtedly, adequate surgical excision of the tumor is necessary, because of the frequent recurrence of LGFMS. Although a vertical incision that includes barriers such as the thick fascia or periosteum is recommended, as based on the criteria for soft malignant tumors, potential advantages of a horizontal skin incision remain to be elucidated. In our patient, a 1-cm skin margin was not sufficient and therefore, during the second operation, an extended 2-cm skin margin was employed. At 1 year after the second surgery, there is no obvious evidence of recurrence ; nonetheless, a long follow-up period is important. Adjuvant radiotherapy or chemotherapy has not been recommended in previous reports ; however, these treatment strategies may be used in cases of multiple metastases or frequent recurrence.

DECLARATION OF INTERESTS

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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