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Low-Grade Fibromyxoid Sarcoma of the Malar Area

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A 34-year-old female patient presented with a mass on the left malar area that had been palpable for 1 year. The mass had enlarged slowly. The patient complained of discomfort and had aesthetic concerns because of the increased size of the mass. She had no specific medical history. However, the patient had a history of trauma on the ipsilateral oral cavity at puberty. A physical examination revealed that there was a movable soft mass with a diameter of 2 cm and

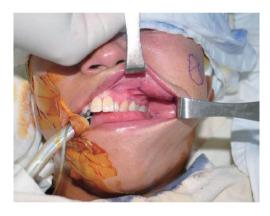
normal-appearing skin. There was no discoloration, erythema, and inflammation. On computed tomography (CT), a slightly diffuse enhancing lowdensity mass was seen on the subcutaneous layer adjacent to the zygomaticus muscle (Fig. 1). Removal of the mass was performed under general anesthesia by using an intraoral approach and a pull-through technique with a gingivobuccal incision. The mass was not easily separated from the surrounding tissues and zygomaticus muscles. The margin of the mass was intermingled with the surrounding fibrous tissue (Fig. 2). The lesion was completely excised with no remarkable occurrences (Fig. 3). Grossly, the mass was round, with a diameter of 1.3 cm. The gross findings of the mass included a well-circumscribed, oval-to-round mass with a thin, fibrous pseudocapsule. The cut surface showed a whorled, whitegray, firm, and fibrous consistency with a homogenous appearance. A histological examination revealed a well-circumscribed mass with no capsulation (Fig. 4). The tumor primarily had a myxoid appearance and was hyper-cellular with bland spindle cells; it had minimal cytologic atypia, generally low mitotic activity and no necrosis, but there were abrupt transitions to collagen rosettes, which was a heavily collagenized area with sparse cellularity. The vasculature in the tumor showed an arcade appearance with hyalinization (Fig. 5). An immunohistochemical study showed that the tumor cells were focally positive for smooth muscle actin but negative for S-100, desmin, and CD34. The



A computed tomography image of the mass: a slightly diffuse enhancing low-density mass on the left malar area on the coronal view.

diagnosis of low-grade fibromyxoid sarcoma (LGFMS) was made on the basis of the collected results. There was no evidence of recurrence at the 8-month follow-up period.

Malignant soft-tissue tumors rarely occur in the head and neck, and surgeons are challenged in determining the optimal method for tumor excision, particularly in the case of tumors located in the face [1]. Moreover, an accurate diagnosis of soft-tissue tumors is often complicated even for well-trained pathologists because of the histologic characteristics of these tumors. LGFMS is a rare type of sarcoma, and the determination of the disease course based on oncologic and histologic features remains controversial [1]. 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, associated with aggressive behavior and a high degree of local recurrence or distant metastasis [2]. LGFMS is a rare

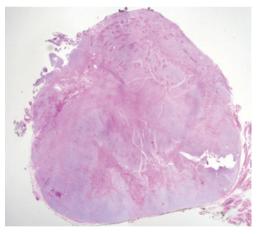


An intraoperative finding: the intraoral approach and the shape of the mass.



An intraoperative finding: the appearance of the exposed specimen.

neoplasm that commonly occurs in young to middleaged adults. Males are more commonly affected than females [3]. The most frequently reported location of LGFMS is the lower extremity, particularly the thigh, followed by the trunk and groin, upper extremities, and buttocks [1,2,4]. The head and neck region is a very rare location for this type of sarcoma. LGFMS is frequently surrounded by skeletal muscle and typically presents as a painless deep soft-tissue mass [3]. CT and magnetic resonance imaging (MRI) are widely used, while ultrasonography (USG) is occasionally used for diagnosing the disease. Immunohistochemically, most cells of this sarcoma are strongly positive for vimentin but are generally negative for α-smooth muscle actin, desmin, S-100 protein, cytokeratin, CD34, and CD56 [1]. The microscopic appearance of LGFMS exhibits bland fibroblastic spindle cells with a whorled or linear arrangement in the fibrous and myxoid areas [3]. The diagnosis of LGFMS is still difficult because of its characteristic bland-looking histologic features that can be confused with other benign or low-grade fibromyxoid lesions [3]. Although LGFMS shows a benign histologic appearance, local recurrence and late metastases have frequently been reported. Evans



A microscopic examination: the tumor is wellcircumscribed and has a myxoid feature (bluish background) (H&E, \times 20).

A microscopic examination: the tumor is hypercellular with spindle cells and contains small vessels with an arcade appearance (black arrows) $(H\&E, \times 100).$

noted that some recurrent and metastatic tumors contain zones of increased cellularity and mitotic activity [2]. In this type of tumor, adequate surgical excision of the tumor is necessary because of the frequent recurrence of LGFMS [1]. The young patient age and superficial location likely contribute to a benign clinical diagnosis in many cases of superficial LGFMS. This clinical impression could potentially result in a delayed biopsy or excision and potentially worsen the patient outcome. The recommended treatment for LGFMS includes radical surgery, wide en bloc surgical resection, complete resection, compartmental excision, and local excision. Adjuvant radiotherapy or chemotherapy has not been recommended in previous reports. As these tumors are prone to metastasis after a long interval, sometimes after as many as 45 years, a thorough clinical follow-up is recommended. However, no study to date has recommended any protocol for the follow-up [5]. Thus, we described a rare case of facial LGFMS in a woman. She had no evidence of local recurrence or metastases at the postoperative 8-month follow-up.

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Squamous Cell Carcinoma Arising from an Epidermal **Inclusion Cyst**

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Epidermal inclusion cysts are commonly encountered benign cystic lesions that can occur anywhere in the body. However, malignant changes are rare, and only a few cases of malignant change have previously been reported [1,2].

Anton-Badiola et al. [2], based on a total of 13 reported cases, found that the mean age at presentation of squamous cell carcinoma arising from an epidermal inclusion cyst was 43.2 years. They also observed that it occurred more frequently in men, that the head and neck area was the most commonly



A 62-year-old male who presented with a 2 x 2-cm soft bulging mass on the left cheek.