Low-grade myofibroblastic sarcoma (LGMS) is a rare sarcoma with myofibroblastic differentiation. LGMS has a propensity for local recurrence and is associated with a low risk of metastatic spread. A 26-year-old man presented with a 12-month history of a slow growing palpable hard mass in the right parietal scalp. Enhanced CT scan of head showed a 3×4 cm sized well-defined and heterogeneously enhancing scalp mass. The patient underwent excision of the tumor with macroscopically clear margins. The mass was relatively well-circumscribed, firm, and grayish with fibrous. Histological examination revealed a spindle cell tumor with interspersed collagen bundles. The spindle cells show moderate degree of nuclear pleomorphism (Fig. 2A). A mitotic count was 1 to 2 per 10 high power fields (HPFs). There was no necrosis or multinucleated giant cells in the numerous sections. The spindle cells were found to be positive for α-smooth muscle actin (Fig. 2B) and negative for desmin, S-100 protein, epithelial membrane antigen and CD34 on immunohistochemistry. Ki-67 labeling index was at 10%. Ultra-thin section shows oval and spindle cells having scant cytoplasm. The tumor cells showing scant organelles except dilated rough endoplasmic reticulum. There are primitive intercellular junctions with cytoplasmic dense bodies (Fig. 3). All of features were consistent with a LGMS. After confirmed a diagnosis, we performed a brain magnetic resonance image (MRI) for finding a possible remnant tumor. T1 weighted image (T1WI) axial brain MRI showed iso-signal intensity remnant mass in right parietal scalp.
scalp (Fig. 4A). Gadolinium enhancement T1WI axial brain MRI showed the heterogeneously enhancing mass, which was measured 0.3×1 cm (Fig. 4B). We performed re-operation for remnant tumor removal. Postoperative course was uneventful. No adjuvant radiotherapy and chemotherapy should be applied. Regular follow-up showed no sign of recurrence. After 14 months of surgery, follow up brain MRI showed no evidence of recurrent lesion in scalp (Fig. 5). FDG-PET CT scan demonstrated no abnormal hot uptake in the body. The patient was well-being state.

DISCUSSION

LGMS is a recently defined tumor that can be considered a part of the spectrum of malignant mesenchymal tumors, showing features of myofibroblastic differentiation. Myofibroblasts have been recognized for over three decades, but the existence of sarcomas composed of myofibroblasts has become generally accepted recently. Myofibroblastic sarcoma must be distinguished from other neoplasms, which include nodular fasciitis, proliferative myositis and fibromatosis, and also other types of low-grade spindle-cell sarcomas, leiomyosarcoma and fibrosarcoma. Mentzel et al. characterized myofibrosarcoma or myofibroblastic sarcoma as a spindle cell sarcoma composed of myofibroblasts. They summarized clinical findings of LGMS patients, which data show that the age of the patients ranged between 19 and 72 years (median : 42 years). Male to female ratio is 1.57. In most cases, patient complained of a painless swelling or an enlarging mass.

The diagnosis of LGMS is made on the basis of ultrastructural findings as determined by light microscopy and immunocytochemistry.
Low Grade Myofibroblastic Sarcoma Occurred in the Scalp

SR Han and GT Yee

LGMS is composed of slender spindle cells arranged in interlacing fascicles. LGMS have a mild to moderate amount of eosinophilic or amphophilic cytoplasm and fusiform nuclei, and usually characterized with mild nuclear pleomorphism and low mitotic rate (1–6/10 HPFs). Our case shows 1 to 2 per 10 HPFs of mitotic rate and moderate pleomorphism of spindle cells.

LGMS tumors cells express strong positive immunoreactivity for at least one of following myogenic markers that muscle-specific actin (HHF35), α-smooth muscle actin, rarely desmin. LGMS may also express focal expression of CD34 and CD99. The tumors do not stain for S-100 protein, epithelial markers such as laminin and h-caldesmon. Our case is negative for CD34 and desmin but positive for α-smooth muscle actin.

LGMS usually affects deep soft tissue sites and is more often poorly circumscribed with fascicles and individual cells infiltrating between muscle fibers, although focal circumscription is not unusual.

The treatment of this rare tumor should include a simple local excision with wide surgical margins and adjuvant therapies such as chemotherapy or radiotherapy have also been used in some cases.

Local recurrences are common in LGMS, but metastases occur only rarely. Follow-up information is limited in the literature, but multiple soft tissues, bone, and pulmonary metastases from a chest wall mass were reported. Therefore, LGMS are needed years of clinical follow-up evaluation including scans.

CONCLUSION

We report of the case of LGMS occurred in the scalp. Because of the rarity of this sarcoma types, its biological behavior and proper treatment are still unclear. However, LGMS can be best managed with wide surgical resection and regular follow-up.

References

1. Arora R, Gupta R, Sharma A, Danda AK : A rare case of low-grade myofibroblastic sarcoma of the femur in a 38-year-old woman : a case report. J Med Case Rep 4 : 121, 2010
2. Demarosi F, Bay A, Monighini L, Carrassi A : Low-grade myofibroblastic sarcoma of the oral cavity. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 108 : 248-254, 2009
3. Diaz-Cascajo C, Borghi S, Weyers W, Metze D : Fibroblastic/myofibroblastic sarcoma of the skin : a report of five cases. J Cutan Pathol 30 :
4. Fisher C : Myofibroblastic malignancies. Adv Anat Pathol 11 : 190-201, 2004
5. Jay A, Piper K, Furlingham PM, Carter J, Diwakar A : Low-grade myofibroblastic sarcoma of the tongue. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 104 : e52-e58, 2007
6. Majno G : The story of the myofibroblasts. Am J Surg Pathol 3 : 535-542, 1979
7. Mentzel T, Dry S, Katenkamp D, Fletcher CD : Low-grade myofibroblastic sarcoma : analysis of 18 cases in the spectrum of myofibroblastic tumors. Am J Surg Pathol 22 : 1228-1238, 1998
8. Nagata Y, Matsuno T, Hamada N, Shimose S, Arihiro K, Ochi M : Low-grade myofibroblastic sarcoma of the palm. Scand J Plast Reconstr Surg Hand Surg 42 : 164-167, 2008
9. Niedzielska I, Janic T, Mrowiec B : Low-grade myofibroblastic sarcoma of the mandible : a case report. J Med Case Rep 3 : 8458, 2009
10. Qiu JY, Liu P, Shi C, Han B : Low-grade myofibroblastic sarcomas of the maxilla. Oncol Lett 9 : 619-625, 2015
11. San Miguel P, Fernández G, Ortiz-Rey JA, Larrauri P : Low-grade myofibroblastic sarcoma of the distal phalanx. J Hand Surg Am 29 : 1160-1163, 2004
12. Takácsi-Nagy Z, Murzakízy G, Pogány P, Fedor J, Orosz Z : Myofibroblastic sarcoma of the base of tongue. Case report and review of the literature. Strahlenther Onkol 185 : 198-201, 2009