Low-grade myofibroblastic sarcoma of the proximal femur
A case report and literature review
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Abstract
Rational: Low-grade myofibroblastic sarcoma (LGMS) is an atypical type of tumor composed of myofibroblasts. LGMS in the femoral head neck junction is extremely rare and no case treated by hip arthroscopy was reported.

Patient concerns: We reported a case of LGMS in the femoral head neck junction treated by hip arthroscopy. A 30-year-old female was admitted to our hospital with discomfort and pain after left hip sprained one year prior. Physical examination revealed swelling of the left hip and magnetic resonance images showed a soft tissue mass in the femoral head neck junction.

Diagnosis: Via microscopy of pathological specimens, spindle cell proliferative lesions, atypia of some cells, and mitotic figures/pathological mitotic figures of some cells were observed. Immunohistochemistry revealed positive for smooth muscle actin, focally positive for CD34 and CD68, while negative for S-100, desmin, and anaplastic lymphoma kinase. The imaging, histomorphological and immunohistochemical features suggested a final diagnosis of LGMS of the proximal femur.

Interventions: This patient underwent hip arthroscopy for excision of the soft tissue mass.

Outcomes: The clinical and imaging follow-up at 6 months postoperatively showed that surgery had achieved good clinical outcomes.

Lessons: To the best of our knowledge, this is the first case report of LGMS in the femoral head neck junction treated by hip arthroscopy. Beyond the present case, other 120 cases from 58 literatures (1998–2022) are reviewed and discussed. The age of LGMS patients ranged from 11 months to 77 years and the male-to-female ratio was approximately 1.28:1. The location distribution of previously reported LGMS cases and the present case was as follows: Head&neck (45.90%), trunk (30.33%), and extremity (23.77%). Hip arthroscopic excision of LGMS may achieve relatively good clinical outcomes.

Abbreviations: CT = computed tomography, LGMS = low-grade myofibroblastic sarcoma, MRI = magnetic resonance images, SMA = smooth muscle actin.

Keywords: Low-grade myofibroblastic sarcoma, proximal femur, hip, arthroscopy

1. Introduction
Low-grade myofibroblastic sarcoma (LGMS) was firstly described by Mentzel et al,[1] which represents an atypical and extremely rare type of tumor composed of myofibroblasts. It was first classified as a new group of soft tissue and bone tumors by the WHO in 2002, and this classification was maintained in 2020.[2] LGMS has been reported to occur in deep soft tissues with predilection for the head and neck.[3] However, according to previous study by Kim et al,[4] the incidence of LGMS in the extremities or trunk may be higher than expected. Based on the rarity of this condition, this study aims to introduce a case report of LGMS in proximal femur and to perform a review of relevant literature.
observed. Computed tomography (CT) showed ossification in adjacent acetabulum (Fig. 1C and D).

This patient underwent hip arthroscopy for excision of the mass. Outside-in approach was used because of anterior ossification. The soft tissue mass, adjacent bone tissue, hyperplastic synovium, and ossification in the acetabulum were debrided. The range of motion returned normal after surgery, just like the contralateral hip. Two pathological specimens were taken during the operation, which were acetabulum ossification and soft tissue mass. The bony tissue of the left acetabulum contained part of dense connective tissue, in which a few proliferative spindle cells could be seen and the shape was mild. In the left hip soft tissue mass, spindle cell proliferative lesions were observed. Some cells showed atypia. Some cells had mitotic figures, and occasionally pathological mitotic figures. Tumor borders were difficult to identify. The soft tissue mass was sent for immunohistochemistry, which revealed positive for smooth muscle actin (SMA), focally positive for CD34 and CD68, while negative for S-100, Desmin, P53, nuclear β-catenin, ALK1, SOX10, MUC4, and STAT6.

The imaging, histomorphological, and immunohistochemical features suggested a final diagnosis of LGMS of the proximal femur. Postoperative CT and MRI were conducted 1 day after surgery and revealed that the tumor was completely removed (Fig. 1E–H). Adjuvant chemotherapy or radiotherapy were not used for this patient. The patient underwent MRI follow-up 6 months after surgery and MRI showed no local recurrence (Fig. 2). Modified Harris Hip Score and visual analog scale for pain improved from 60 and 5 preoperatively to 78 and 1 postoperatively 6 months after surgery.

The patient had provided informed consent for publication of the case, and the study protocol was approved by Medical Ethics Committee of Peking University Third Hospital.

3. Discussion

Two electronic databases were searched: PubMed (1966-), Web of Science (1900-), with the key words “low-grade myofibroblastic sarcoma” and its synonyms. Non-English literature
## Table 1
Details of included articles: information and study sample characteristics.

| Authors                     | No. of cases | Age mean | Sex (M/F) | Site                | Treatment                  | Clinical outcome |
|-----------------------------|--------------|----------|-----------|---------------------|----------------------------|------------------|
| Mentzel et al.[1]            | 18           | 42       | 11/7      | Head&neck           | S 12                       | NR 8             |
|                             |              |          |           |                     | Trunk 6                    | S, RT 2          |
|                             |              |          |           |                     | Extremity 7                | S, CTX 1         |
|                             |              |          |           |                     | S, RT, CTX 1               | NA 2             |
|                             |              |          |           | Head&neck           | S 7                        | R 5              |
|                             |              |          |           |                     | Trunk 3                    | S, RT 2          |
|                             |              |          |           |                     | Extremity 3                | NA 1             |
|                             |              |          |           |                     | Trunk 1                    | S 1              |
|                             |              |          |           |                     | Trunk 1                    | S 1              |
|                             |              |          |           |                     | Extremity 1                | S 1              |
|                             |              |          |           |                     | S, RT, CTX 1               | NA 1             |
|                             |              |          |           | Head&neck           | R 1                        |                 |
|                             |              |          |           |                     | Extremity 3                | S 1              |
|                             |              |          |           |                     | S, RT, CTX 1               | NA 1             |
|                             |              |          |           | Head&neck           | S 1                        | R 1              |
|                             |              |          |           |                     | S, RT, CTX 1               | NA 1             |
|                             |              |          |           | Head&neck           | RT, CTX 1                  | DOD 1            |
|                             |              |          |           |                     | Head&neck 6                | S 8              |
|                             |              |          |           |                     | Trunk 5                    | S, CTX 4         |
|                             |              |          |           |                     | Extremity 3                | S, RT 2          |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 3                | S, RT 3          |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | S, RT, CTX 1               | NA 1             |
|                             |              |          |           | Extremity 1         | S 1                        |
|                             |              |          |           |                     | S 1                        |
|                             |              |          |           |                     | S, RT, CTX 1               | NA 1             |
|                             |              |          |           | Head&neck 2         | S 2                        |
|                             |              |          |           |                     | Trunk 1                    | S, CTX 1         |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S, RT 1          |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S, RT, CTX 1     |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
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|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |
|                             |              |          |           |                     | Head&neck 1                | S 1              |

(Continued)
and inaccessible literature were excluded. We analyzed 120 cases from 58 literatures. With this present case, we have a basic understanding for the information and characteristics of the LGMS patient population (Table 1). Among the 121 reported LGMS patients, the age of them ranged from 11 months to 77 years (mean, 42.43; median, 44). The male-to-female ratio was approximately 1.28:1 (68 males versus 53 females). The detailed age and gender distribution are shown in Figure 3.

We classified locations of LGMS into 3 groups: Head&neck (56 cases, 45.90%), Trunk (37 cases, 30.33%), and Extremity (29 cases, 23.77%). It should be noted that one of the patients had LGMS which occurred in multiple organs, including the diaphragmatic pleura and head and neck region. The case was counted into Head&neck group and Trunk group, respectively. In addition, several new primary sites of LGMS have been reported in recent years, including limbus,[33] orbit,[38] and multiorgan.[64] LGMS was reported to be prone to local recurrence rather than distant metastasis.[4] And the data we collected support this conclusion. Notably, 2 cases of cardiac metastasis of LGMS were reported, both of which resulted in the eventual death of the patient.[17,33] Among the 121 patients with LGMS, the number of No Recurrence cases during the follow-up was 76 (62.81%), which suggests that LGMS patients after proper treatment generally have good prognosis. And Chan et al[60] reported that the 5-year overall survival rate of LGMS was 71.6% and the disease-specific survival rate was 76.3%, which is consistent with our conclusion.

The most common treatment for LGMS was surgical excision/wide excision. Some clinicians used chemotherapy or radiotherapy as adjuvant treatment strategies, while most were more concerned about whether the tumor is completely removed. Based on the data we collected, once diagnosed properly and excised by wide enough, LGMS is less likely to recur. A quantitative study by Xu et al[61] supports our observations. They reported that age greater than 60 years, positive nodal status, and no surgical treatment were independent prognostic factors for patients with LGMS, whereas chemotherapy and radiation treatment were not. However, some authors reported that LGMS occurring in the Head&neck may not obtain adequate resection margins due to surgical limitations.[21,32] Recently, Lin et al[69] reported that Apatinib functioned as an effective treatment of LGMS via potential VEGFR-PISK/Akt signaling pathway.

LGMS is an atypical type of tumor composed of myofibroblasts, which results in the strong expression of Vimentin.[62] Therefore, immunohistochemistry can be used for diagnosis and differential identification. Gonçalves et al[67] reported that in their literature review consisting of 30 studies, 26 and 10 of them used α-SMA (alpha-SMA) and muscle-specific actin as myogenic marker, respectively. In our present case, immunohistochemistry revealed positive for SMA, suggesting the tumor cells were derived from myofibroblasts. Also, it’s well known that S-100 protein is a specific biomarker for schwannoma and malignant peripheral nerve sheath tumor.[11] S-100 should be negative in LGMS, which is consistent with our present case. In addition, immunohistochemistry revealed negative for anaplastic lymphoma kinase, helping to differentiate LGMS from inflammatory myofibroblastic tumor.[63] Inflammatory myofibroblastic tumor is a type of tumor similar to LGMS because of their morphologic similarity and the overlapping immunophenotype.[46]

4. Conclusion

To the best of our knowledge, this is the first case report of LGMS in the femoral head neck junction treated by hip arthroscopy. Beyond the present case, other 120 cases from 58 literatures (1998–2022) are reviewed and discussed. The age of LGMS patients ranged from 11 months to 77 years and the male-to-female ratio was approximately 1.28:1. The location distribution of previously reported LGMS cases and the present case was as follows: head&neck (45.90%), trunk (30.33%), and extremity (23.77%). Wide excision with clear margins may achieve relatively good clinical outcomes.

Author contributions

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