Pigmented Villonodular Synovitis Around the Elbow Joint That Required Upper Arm Amputation

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Abstract

Pigmented villonodular synovitis (PVNS) is a relatively rare benign proliferative disorder of the synovium, and it commonly occurs in the knee joint. A unique case of a huge PVNS originating around the elbow joint that required upper arm amputation is presented. A 74-year-old woman had a 5-month history of right elbow pain and noticed a mass measuring 8 × 5 cm on the olecranon side. She had chronic renal failure, with a shunt on the right forearm. On magnetic resonance imaging, the tumor around the elbow joint showed low signal intensity on T1-weighted images and low to high intensity on T2-weighted images. It was thought that it was difficult to preserve the limb because of the size of the tumor and the shunt blood vessel. Therefore, upper arm amputation was performed. Pathological examination showed invasion proliferation to the synovial membrane, osteoclast-like giant cells, proliferation of round cells, and hemosiderin accumulation between cells (Fig. 1). In addition, lesions were continuous from the periosteum and proliferated invasively in the dermis. The histological diagnosis was PVNS. There was no recurrence of PVNS postoperatively.

Keywords: Pigmented villonodular synovitis; Upper arm amputation; Tumor of soft tissue

Introduction

Pigmented villonodular synovitis (PVNS) is a relatively rare benign proliferative disorder of the synovium, and it commonly occurs in the joints, with the knee joint being involved most frequently, followed by the hip, ankle, and shoulder joints [1]. Elbow joint involvement is uncommon in PVNS [2]. A unique case of a huge PVNS originating around the elbow joint that required upper arm amputation is presented, along with a review of the literature.

Case Report

A 74-year-old woman presented at our outpatient clinic due to slight pain of the right elbow persisting for 5 months. She had a past history of undergoing tumor resection and flap surgery 9 years earlier, and the mass was diagnosed as a giant cell tumor. On pathological examination, the margin of the resected tumor was diagnosed as intralesional resection. After administration of gadolinium contrast, it was enhanced irregularly. A needle biopsy was performed, and histopathologic examination showed a giant cell tumor resembling the pathological findings 9 years earlier, with no malignancy.

The tumor gradually grew larger and formed ulcers. The tumor was present almost over the entire circumference of the upper arm and invaded the humerus, with the shunt blood vessel in the right forearm. Therefore, it seemed that it was difficult to preserve the limb, and upper arm amputation was performed. Pathological examination showed invasive proliferation to the synovial membrane, osteoclast-like giant cells, proliferation of round cells, and hemosiderin accumulation between cells (Fig. 1). In addition, lesions were continuous from the periosteum and proliferated invasively in the dermis. The histological diagnosis was PVNS. The patient’s postoperative course was uneventful. There was no recurrence of PVNS at the most recent follow-up 2 years after surgery.

Discussion

PVNS is a benign proliferative disorder of unknown origin forming nodular or villous changes in joints, synovial capsules,
and tendon sheaths [3]. PVNS is one of the benign tumors, so-called fibrohistiocytic tumors, and it is classified as diffuse type in tenosynovial giant cell tumor [4]. It generally has an intra-articular origin, and it shows extensive involvement of the synovial membrane and capsule [5]. PVNS with extra-articular extension is rare [6], and most have been reported to occur in the knee and hip joints [7, 8]. PVNS in the elbow is uncommon, with only 24 cases reported in the literature [9, 10]. Although a few cases of extra-articular giant cell tumor of the tendon sheath occurred at the elbow, there have been no reports of PVNS with extra-articular extension affecting the elbow joint [5].

If it is left untreated, the hypertrophic synovium and multiple soft-tissue masses may lead to continued pain, decreased range of motion, joint destruction, and osteoarthritis [11]. Therefore, operative management, such as open excision or arthroscopic excision, is often required. The recurrence rate after surgical treatment has been reported to be 31-33%, and it is important to completely resect the synovial membrane [12, 13]. However, complete resection is difficult if it occurs within complex joints such as the elbow joint [14]. To minimize the risk of recurrence, several studies have demonstrated a positive response with radiation therapy or adjuvant therapy [13, 15]. However, their effects are still controversial [16]. There are no cases of PVNS that required amputation of a limb. PVNS must be carefully treated because of its invasiveness, though it is not a malignant tumor.

In conclusion, a rare case of PVNS around the elbow joint that required upper arm amputation was presented. In the treatment of PVNS, initial surgery with complete resection of the synovial membrane and careful postoperative follow-up are considered important.

Acknowledgments

None to declare.

Financial Disclosure

The authors declare that there is no funding regarding the publication of this article.

Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Informed Consent

The patient and/or her family were informed that data from the case would be submitted for publication and gave their consent.

Author Contributions

YO and HT wrote the paper; NM, H. Nagasawa, H. Nanjo, and YS reviewed the paper; all authors gave the final approval.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

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