Pseudoaneurysm Requiring Differential Diagnosis from Malignant Soft Tissue Tumor —A Case Report—

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悪性軟部腫瘍との鑑別を要した大腿部仮性動脈瘤の一例

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Arteriovenous malformation (AVM) is classified as a type of congenital venous malformations. It is sometimes difficult to distinguish AVM from malignant soft tissue tumor based on the clinical findings. The authors report a case of AVM that brought about pseudoaneurysm without any events.

A 49-year-old female who recognized a tumor on her left thigh from one year ago complained of an increase in the size of the tumor and pain. Physical examination showed a large tumor from the medial to posterior part of the left thigh. The size of the tumor was about 31×27 cm, an elastic hard mass by palpation, and resembled arterial murmur by stethoscope.

A malignant soft tissue tumor was suspected based on the rapid increase in size and physical findings at the first visit to our outpatient clinic. Ultimately, diagnosis was made as AVM with pseudoaneurysm using enhanced CT and angiography. Embolization procedures were required three times to prevent blood flow into the pseudoaneurysm, occluding the branches of superficial femoral artery and superficial iliac artery, which fed the AVM. Pseudoaneurysm was occluded finally, but all feeders were not occluded because of broad AVM exposure. Continuous follow-up is necessary due to the risk of rebleeding from AVM.

(はじめに) 臨床上、悪性軟部腫瘍との鑑別を要した大腿部仮性動脈瘤の一例を経験したので報告する、（症例）49歳女性。既往歴：先天性極端部脱臼に対して16歳時に骨切術を受けた。現病歴：平成20年6月より左下肢に違和感を、同年秋頃から大腿部の硬結、腫脹を自覚していた。平成21年初めから腫脹が徐々に増大し、同年5月より膝関節の伸展障害も出現したため当科受診となった。左大腿部内側より後面にかけて26×15×20cmの弾性硬の腫瘤で血管雑音を聴取した。圧痛、熱感は伴っていなかった。造影CTにて動静脈奇形（AVM）とAVM周囲に仮性動脈瘤をみとめ、持続的な出血をみとめた。その後、出血の塞栓術を2回に分けて施行した。貧血の進行が無いことを確認後、外来での経過観察とし現在も観察中である。（考察）急性に増大する腫瘤と腫瘤内に著明な血管雑音をみとめた場合、本疾患も鑑別として考慮すべきである。

Key words: arteriovenous malformation (AVM) (動静脈奇形), pseudoaneurysm (仮性動脈瘤), malignant soft tissue tumor (軟部組織恶性腫瘍), embolization (塞栓術)

Introduction

Although the head, neck and distal part of the extremities are more commonly affected by AVM, the occurrence of AVM on the proximal part of the thigh, which includes the pelvis, is rare.

The authors report a case of AVM with

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pseudoaneurysm on the thigh, which was misdiagnosed as malignant soft tissue tumor during the first examination.

Case

A 49-year-old female was referred to our hospital for the management of a mass lesion on her left thigh. She had an approximate one-year history of gradual tumor growth and pain. The patient was not concerned about it because it did not affect her daily life. She recognized the condition to be abnormal after about six months when poor knee extension and walking developed. She denied any recent trauma to the leg or recent MRI examination, which could influence residual internal fixators.

The subject had a past surgical history of osteotomy on the left hip due to the worsening of congenital dysplasia.

Physical examination revealed firm consistency, located on the medial to posterior aspect of the left thigh. The size of soft tissue mass was 31×27 cm; bruit was audible, but thrill was not palpable (Fig. 1).

Blood examination showed slightly inflammatory reaction and anemia (white blood cell level was 5900/μl, C-reactive protein was 2.45 mg/dl, and hemoglobin level was 7.3 g/dl). X-ray showed soft tissue mass shadow and bone atrophy at the proximal part of the left femur, where the internal fixation devices used in the last osteotomy were observed (Fig. 2).

Enhanced CT scan showed the pseudoaneurysm, for which the cavity was filled with contrast medium and hematoma around it (Fig. 3).

Transarterial embolization for pseudoaneurysm and AVM was performed by radiologists in our hospital. Feeding arteries derived from the branches of internal iliac artery, lateral femoral circumflex artery, and deep femoral artery were confirmed. Transarterial embolization was per-

Fig. 1 Patient’s thigh at the time of first examination
Soft tissue-mass was located from the medial to lateral side of the left thigh. The size of the mass measured about 31×27 cm.

Fig. 2 X-ray findings
X-rays showed an increase in the size of soft tissue in the thigh. Internal fixation devices were observed. Atrophic bone change was advancing in comparison with the right femur. Due to the free foreign metal body derived from prior surgery, it was difficult to perform MRI examination.
Fig. 3  Enhanced CT scan showing the condition in the thigh
An enhanced CT scan revealed enhanced pseudoaneurysm surrounded by hematoma. Huge hematoma compressed the normal adjacent tissue. The condition can lead to several symptoms.

Fig. 4  Embolization procedures were performed several times
Pseudoaneurysm (arrow: •••) was almost occluded, but the lesion of AVM (arrow: →) was not occluded completely because of its broad range. Even though embolizations were attempted three times, residual AVM lesions leave the risk of severe complications.

formed to occlude feeding arteries, and percutaneous embolization was added to occlude the huge pseudoaneurysm cavity by injecting of NBCA (N-butyl-2-cyanoacrylate) + Lipiodol. Cessation of embolization was decided before occluding all feeding arteries because the range of
AVM was too broad to occlude all feeding arteries at one time. Two weeks later, a second embolization was performed. Confirming no new pseudoaneurysm formation and extravasation of contrast medium, the series of procedures was finished. The patient was discharged from our hospital and visited the outpatient clinic once per month. Three months after discharge, extravasation into the pseudoaneurysm was confirmed with enhanced CT scan again. A third embolization was performed (Fig. 4). Periodic examination of the patient is ongoing.

Discussion

Parry et al. reported that pseudoaneurysm formation in the femoral vessels is rare and represents only 3-7% of all pseudoaneurysms\(^{12}\).

Several factors can be the pathogenesis of pseudoaneurysm. In this case, the patient had a surgical history of osteotomy for congenital dislocation of the hip joint. Surgery was suspected as one of pathogeneses of AVM. The most recent surgery, which was performed 33 years ago, may have influenced the pseudoaneurysm formation.

Because the authors were unable to find literature reporting surgery that influenced the development of AVM after over three decades, the most recent surgery could not be attributed as a pathogenesis of AVM in this case. Nevertheless, Chatt et al. reported one case which showed the deterioration of pseudoaneurysm 54 years after injury for an explosion accident on the proximal left thigh, and reported a residual mortar fragment\(^6\).

This report advocates the possibility that the residual foreign body may become the cause of the pseudoaneurysm development.

This case was diagnosed as malignant soft tissue tumor because the soft tissue mass was very large and showed rapid increase in size. Keller pointed out that physical examination findings in pseudoaneurysm are relatively straightforward and include swelling, audible bruit, pulsatile mass, and pain\(^5\).

The presentation of a pseudoaneurysm mimicking a musculoskeletal tumor is very rare. AVM can be diagnosed with adequate radiological findings; for example, enhanced CT scan, MRI, and angiography\(^{13}\).

Diagnosis by clinical findings was difficult, but radiological findings were useful. X-ray examinations show a nonspecific soft-tissue mass, and rim calcification\(^4\).

Erler et al. reported that MRI allows for differential diagnosis between hematoma and tumoral conditions, and angiography is the most sensitive radiological method for the diagnosis and detection of the arterial origin of a false aneurysm\(^5\).

This case was diagnosed as AVM with pseudoaneurysm by enhanced CT scan. Enhanced MRI was not performed in this case due to the risk of migration of metal foreign bodies in the muscle. AVM seems to have relation with rapid increase of pseudoaneurysm. AVM behave aggressively like malignant tumors because the majority of AVM belong to the residual remnants of a developmental arrest in the early stage of embryonic life. They have a tendency to progress with a more destructive potency. The disposition of AVM makes the diagnosis difficult among tumoral lesions\(^6\).

Several authors advocate treatments including surgical resection, transarterial percutaneous embolization, sclerotherapy, and the combination of these therapies. The definite treatment is not still clear because AVM, which originates from the mesenchymal cells at an early stage of embryogenesis, can grow rapidly. Its explosive growth makes the selection of treatment difficult\(^5\).

Tan et al. suggested the indications for embolization therapy included (1) hemorrhage, (2) disabling pain requiring regular use of analgesia,
(3) functional impairment secondary swelling or pain, (4) high-output heart failure, and (5) ulceration.

Olcott et al. classified AVM in three types for the indication of treatment. His classification pointed out the indication of the treatment, which depends on the size and the location of the lesion. Inadequate treatment often stimulates dormant AVM to grow rapidly, making the condition worse. The authors judged embolization therapy was adequate for the case reported because the lesion was too large to resect, and incomplete resection could lead to uncontrolled recurrence. Therefore, the first purpose of the treatment was to interrupt the blood flow into the cavity of pseudoaneurysm; the second purpose was to occlude the feeding arteries.

Several authors reported the repeated recurrence of AVM can bring about severe complications. Kim et al. reported three cases that were selected for surgical resection but ultimately reached amputation procedure. Onishi et al. reported a case experiencing several embolization treatments over 15 years where several recurrences led to the disarticulation of the hip.

The primary effect of AVM and/or pseudoaneurysm on the surrounding tissues is the direct compression by the lesion itself and ischemic erosion. Secondary hemodynamic effects include a potential arterial steal phenomenon, which is widely known where arterial blood is shunted through the low-resistance arteriovenous shunts of the AVM away from the normal tissue adjacent to and distant from the AVM. The heart can be affected by high-output cardiac failure. Peripheral tissues can be affected in a wide range of changes from distal ischemia to gangrene, venous stasis dermatitis, and ulcer or gangrene caused by venous hypertension. Finally, only amputation procedure was left as the possible treatment.

Inadequate treatment can stimulate residual lesions and cause the recurrence of AVM. Its evolitional potential to grow can lead to more severe complications. Because AVM residual lesion was present in this case, it is necessary to pay attention to the progress of complications.

Conclusions

1. The authors experienced one case of AVM with pseudoaneurysm that was first suspected to be a malignant soft tissue tumor.
2. Enhanced CT was useful to differentiate AVM from malignant soft tissue tumors.
3. Because this case has AVM residual lesion, careful long-term follow-up of the patient is required.

References


