

Two Cases with Lower Limb Intraosseous Arteriovenous Malformation: Embolization Therapy Option

Alt Ekstremitede İntraosöz Arteriyovenöz Malformasyonlu İki Olgu: Embolizasyon Tedavisi Seçeneği

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ABSTRACT Arteriovenous malformations (AVMs) are high-flow vascular malformations which may remain asymptomatic for many years or become apparent with pain, swelling, ulceration, distal ischemia, and hemorrhage. Treatment is mandatory in symptomatic lesions. In English literature there are lots of studies about extremity AVMs signs and treatments. However intraosseous AVMs not including surrounding soft tissues are very rare and there are few case reports about its characteristics and treatment. In this study we present two cases with AVMs localized in the distal femur and involving the growth cartilage. The first patient was a 20-year-old man who presented with an intra articular haematoma following a minor trauma in his left knee and an AVM was detected with DSA in the femur. It was treated with multiple selective embolizations. The second patient was a 14-year-old boy who presented with left leg swelling and an AVM was detected in the femur. Because the intraosseous anomalous vessels were spreading into the growth cartilage, we decided to follow him clinically until a complication occurred. Under the light of these two cases, we reviewed clinical presentations, angiographic findings and embolization treatments of extremity AVMs and the relevant literature.

Key Words: Embolization, therapeutic; arteriovenous malformations; lower extremity

ÖZET Arteriyovenöz malformasyon (AVM)'ler yüksek akımlı vasküler malformasyonlar olup, yıllarca asemptomatik kalabilirler veya ağrı, şişlik, ülserasyon, distal iskemi ve kanama ile ortaya çıkabilirler. Semptomatik lezyonlarda tedavi gereklidir. İngilizce literatürde ekstremitte AVM'nin bulgularının ve tedavilerinin anlatıldığı birçok çalışma mevcuttur. Ancak komşu yumuşak dokuları kapsamayan, sadece intraosöz yerleşimli AVM'ler çok nadir olduğundan bunların karakteristiklerini ve tedavisini anlatan birkaç adet olgu bildirisi mevcuttur. Biz bu çalışmada, distal femurda yer alan ve büyüme kartilajına uzanan 2 AVM olgusunu sunuyoruz. İlk hasta 20 yaşında erkekti ve hafif bir travmadan sonra sol dizinde eklem içinde hematoma ile gelmişti ve DSA ile femurda AVM saptanmıştı. Lezyon multipl selektif embolizasyonlar ile tedavi edildi. İkinci hasta sol bacağına şişlik ile gelen 14 yaşında erkek çocukta ve femurunda AVM saptanmıştı. İntraosöz anormal damarlar büyüme kartilajına uzandığından bu olguda komplikasyon gelişene kadar takip kararı verildi. Bu iki olgunun ışığında ekstremitte AVM'lerinin klinik prezentasyonları, anjiyografik bulguları ve embolizasyon tedavileri literatürlerle birlikte tartışıldı.

Anahtar Kelimeler: Tedavi edici embolizasyon; arteriyovenöz malformasyon; alt ekstremitte

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Mulliken classifies vascular malformations into low flow and high-flow types.¹ Capillary, venous, lymphatic, and mixed malformations are low-flow type anomalies.¹ AVMs, which are typified

with hypertrophied inflow arteries, shunting through a primitive vascular nidus into tortuous dilated outflow veins, are high-flow congenital vascular malformations. The most common anatomic locations are the pelvis, extremities, and the intracranial circulation.^{2,3}

Primary intraosseous AVMs make up less than 1% of all vascular tumors of bone.⁴ Extremity AVMs can remain asymptomatic for many years or become manifested with pain, swelling, ulceration, distal ischemia, hemorrhage, and leg-length discrepancy. Treatment is mandatory in symptomatic lesions.

We present two cases of arteriovenous malformations localized in the bone without soft tissue involvement, both of which were involving the growth cartilage. Clinical presentations, angiographic findings of these rare cases and embolization treatments of extremity AVMs are reported and the relevant literature is reviewed.

CASE REPORT

CASE 1

The first patient was a 20-year-old man. Left knee intraarticular haematoma had developed after a minor trauma and an AVM was detected with DSA. The patient had been referred for embolization but he had refused treatment. After 2 years, he came to our clinic with knee pain and swelling due to intraarticular bleeding and decided to undergo therapy. After informed consent was obtained diagnostic angiography was performed to define the AVM's angioarchitecture and identify any anastomoses with normal structures, aneurysms within the lesion, venous drainage, and the potential routes of access for endovascular therapy. The AVM was quite large with multiple nidi. The AVM was feeding from deep femoral artery and popliteal artery's multiple muscular and genicular branches. Malformed vascular vessels were gathered in-and around the epiphysis. There were multiple arteriovenous shunts, intranidal, and venous side aneurysms. It was draining via the popliteal vein (Figure 1A, B). Embolization with glue [N-butyl-cyanoacrylate



FIGURE 1A: AP Knee radiography of patient 1 shows lytic lesions with fine borders. There is no cortical reaction or soft tissue mass.

(NBCA) Histoacryl] was planned. A 5F guiding catheter was placed into the popliteal artery with retrograde puncture. A 3-F micro catheter was advanced coaxially through a 5 F guiding catheter and advanced carefully as close as possible to the nidus. Papaverine was ready for severe vasospasm but it did not occur. Proper catheter placement was confirmed by contrast injections through the guiding catheter. After insertion of the micro catheter, glue and lipiodol mixture was injected. Embolization was done through deep femoral artery and popliteal artery branches. Glue-lipiodol ratio was 25%. Embolization was done in 3 repeated sessions with three months time interval. After the embolizations, superficial femoral artery was patent and minor fever and pain oc-

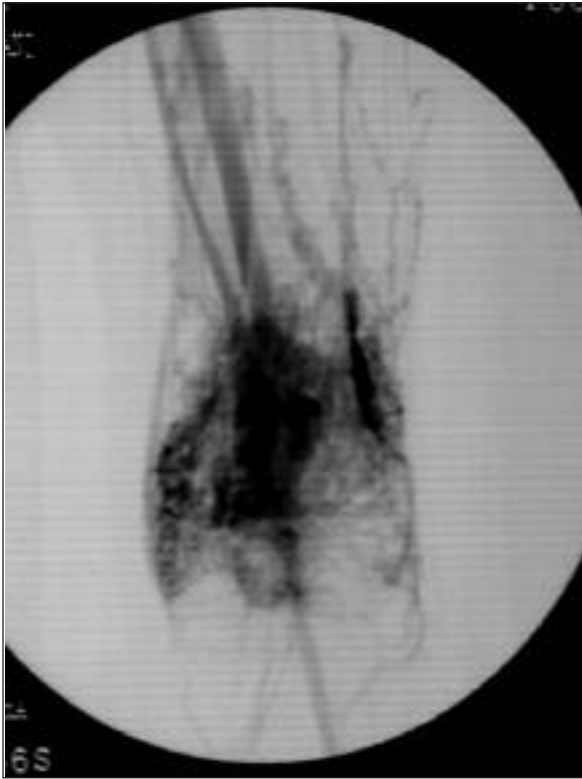


FIGURE 1B: Angiography by superficial femoral artery injections shows vascular malformation in the left knee. Vascular channels are gathering in-and around the femur epiphysis.

curred (Figure 1C, D). Fever lasted for a few days and pain was palliated with nonsteroid analgesics. No skin ulceration was detected. The patient was symptom free until to date (15 months).

CASE 2

The second patient was a 14-year-old boy with left leg swelling and multiple varicose veins (Figure 2A). Ascending venography showed multiple varicose veins with patent deep crural venous system. Because of localized dilution of contrast opacification in the popliteal vein, high flow communication (AV fistula draining vein entrance?) was suspected (Figure 2B, C). On diagnostic arteriography, he had AVM in the left knee at the same localization as case 1, which was feeding from medial and lateral geniculate arteries. The interosseous anomalous vessels were spreading into the growth cartilage (Figure 2D). After consultation with orthopaedics, clinical follow up was decided until a complication occurred (because of fear of iatrogenic ischemia of epiphyseal cartilage and frustration of the growth) and en-



FIGURE 1C: Postembolization angiography shows that there is no nidi and fistula opacification from popliteal artery. Also note that it seems there is short occlusion at the popliteal artery due to compact glue cast subtraction artifact (arrow).



FIGURE 1D: Postembolization angiography shows that there is no nidi and fistula opacification from deep femoral artery.

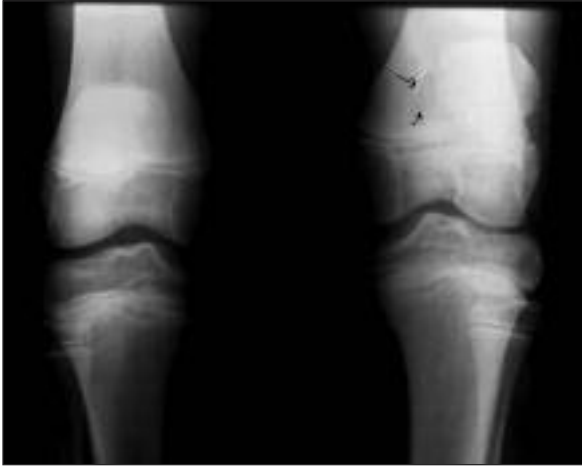


FIGURE 2A: Bilaterally AP knee radiography of patient 2 shows lytic lesion within the left femur with fine borders.

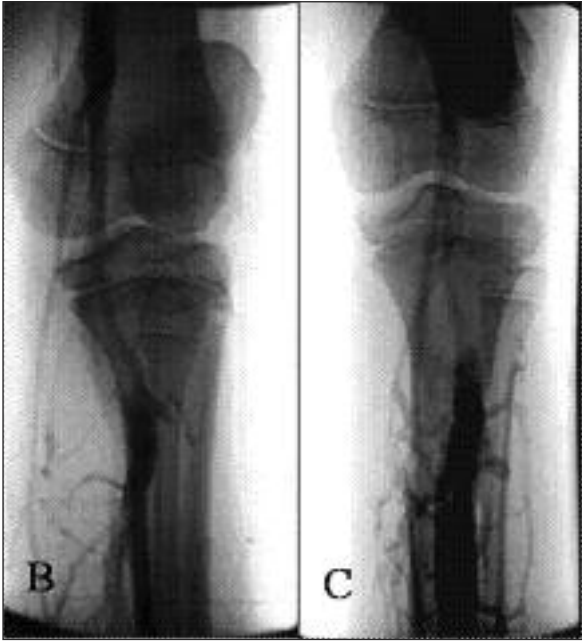


FIGURE 2B, C: B (Oblique position), C (AP position) Ascending venography shows multiple varicose veins with patent deep crural venous system and localized dilation of contrast opacification in the popliteal vein.

dovascular therapy was planned after closure of growth plate.

DISCUSSION

AVM's can result from penetrating trauma or can be congenital. Although posttraumatic lesions are

usually single, congenital lesions can be multiple and more diffuse. Congenital lesions can be established after delivery or can appear after trauma or during periods of hormonal stimulation, such as in menarche and pregnancy or any time during lifetime.³ Up to 50% of congenital AVM's are localized in the limbs and 2/3 of them are in the lower extremities.⁵

The signs and symptoms of AVM in extremities are related to the lesion size and location. Thrill, palpable mass, hypertrophy in the neighboring bones and soft tissues, atrophy and ulcers at the tip of the extremities, changes in the overlying skin pigment, varicose veins, tiredness of the leg due to chronic venous hypertension, pain due to nerve impressions and heart failure in large lesions can be seen. They can lead to leg-length discrepancy due to hypertrophy. Parkes-Weber syndrome (PWS) is characterized by arteriovenous malformations with bony and soft-tissue hypertrophy of one or more limbs.^{3,6} Although the diagnosis of extremity AVM's can usually be made by



FIGURE 2D: Diagnostic arteriography shows an AVM in the left knee which is feeding from medial and lateral geniculate arteries and the interosseous anomalous vessels are spreading into the growth cartilage. There is a venous lake seen in the lytic lesion.

history and physical examination, intraosseous component of a soft tissue AVM can not be excluded with clinical findings alone.

In English literature there is limited case reports describing arteriovenous malformations localized in the bone.^{4,7} Most of intraosseous AVMs are localized in the craniofacial bones and the bodies of vertebrae.⁸ Intraosseous AVMs can be detected as cortical and medullar multiple, smooth, lucent serpiginous lesions often with sclerotic borders and without periosteal reaction and soft tissue mass. The lesion can lead to expansion of the medullary canal and enlarging of the nutrient canal. Multiple phleboliths can be found in venous ectasias on the radiograph.^{4,9} Haemarthrosis with intra-articular extension can be seen. Intraosseous AVM's X-ray findings may not be differentiate from other lytic lesions and should be evaluated with other diagnostic modalities.

Computed tomography (CT) scanning and magnetic resonance imaging (MRI) can be used for confirmation. Bone expansion, periosteal reaction, lytic or sclerotic osseous changes, contrast enhancement, thrombus formation and calcification are easily detected with CT. CT provides less information about blood flow; MRI has replaced CT in the evaluation of vascular malformations. MRI is the most valuable modality in the classification of vascular malformations.¹⁰ The extent of the AVM (focal, multifocal or diffuse) and involved tissue layers could be determined with MRI. Slow-flow vascular malformations have high signal intensity on T2 weighted images. High-flow vascular malformations show signal voids.¹⁰ Low and high-flow vascular lesions can be distinguished with MRI. On occasion atypical low flow malformations may share features with angiosarcoma or fibrosarcoma. In such a case biopsy should be done.¹¹ Angiography is important for preoperative assessment and as a precursor to interventional procedures. Feedings arteries, draining veins and nidi are seen on angiography in high flow vascular malformations. In our patients, AVM had been diagnosed with clinical findings and they were referred to our clinic for DSA. After DSA we did not require MRI examination.

The management of all types of vascular malformations requires a multidisciplinary approach and treatment should, if possible, be conservative. Therapy indications for AVMs that are localized in the bone or soft tissues are the same. It is mandatory in the case of an enlarging lesion before significant overgrowth of the extremity or severe disfigurement of the patient develops. Bleeding of the lesion, infection and critical distal ischemia findings are other indications for treatment.¹² Angiography is necessary if treatment is planned.

The treatment options of intraosseous and soft tissue AVMs are similar. Total surgical resection is generally considered the optimal therapy because of the tendency of the AVMs to expand and recur with time. However, surgery is often associated with profuse bleeding, incomplete resection and local recurrence.¹³

Embolization is an important option in complex AVM treatment alongside surgery. It may be used in all AVMs whether they are localized in the bone or soft tissues. It is rarely curative but often palliative. It can also be used preoperatively to achieve effective devascularization, minimize intraoperative blood loss, facilitate subsequent surgical excision and permit a more limited amputation. The goal of embolization is to occlude at the level of the nidus. Permanent embolic materials are used for the treatment. Liquid adhesives such as onyx, cyanoacrylate, and polyvinyl alcohol particles (PVA), stainless steel coils and detachable balloons are all used materials. Because embolization coils and detachable balloons occlude large vessels, they generally have no role in the embolization of complex AVMs except that of the lung AVMs. PVA have been used, but determining the appropriate size to use is problematic, leading to the risk of paradoxical embolization to the lungs.¹⁴ The ability of a liquid agent to penetrate and occlude at the level of the nidus makes it critically important. Cyanoacrylate is the main liquid adhesive used in endovascular procedures. It is easy to deliver cyanoacrylates through microcatheters because they have low viscosity. However, careful attention and fluoroscopic control is mandatory during the treatment. Because reflux and

glue entering a branch distal to the catheter tip can occur and can lead to unwanted ischemic injury. Gluing of the catheter tip is another complication. As the volume of glue is usually quite small, pulmonary embolism is usually asymptomatic and can be ignored.¹⁵ The development of new collaterals can be managed with repeated embolization therapy when necessary. In our case cyanoacrylate embolization was achieved without major complications. Minor fever and pain occurred and nonsteroid analgesics were sufficient for palliation.

There are several studies defining long-term results of transcatheter embolization due to the AVM's time course prolong. White et al treated 20 patients, followed those 1-18 years, and reported their results in 2000. They noticed that lower extremity AVM's with diffuse involvement of all three trifurcation arteries ultimately required amputation because of recurrence of symptoms after technically and clinically successful embolotherapy. Cyanoacrylate embolotherapy alone or in combination with surgical resection of the AVM provided excellent long-term palliation in patients with upper extremity AVM's.¹⁶ Rockman et al treated 34 patients with predominant arteriovenous shunting or predominantly arterial AVM. They

used different embolization materials and said that transcatheter embolization should be the treatment of choice for symptomatic cases.³ Kromhout et al reported their experience on 81 patients. They treated them with a combination of surgical and transcatheter techniques. They felt that the more proximal lesions with arteriovenous shunts did better with embolization.¹⁷ Our patient was symptom free for 15 months. After embolization sclerosis in the intracortical or intramedullar lucent areas were seen.⁴

Prognosis in all types of AVMs depends on making a correct diagnosis before complications occurs and choosing suitable treatment options.

CONCLUSION

Because of the rarity, there are few reports in English literature published about characteristics and treatment options of introsseous AVMs. In this study we reported 2 cases with introsseous AVM along with their clinics, graphics and management. It is important to know that when an AVM has become symptomatic, curative complete surgical resection is rarely possible. In patients with large AVM's transcatheter arterial embolization is useful to eliminate or improve symptoms, although multiple procedures are often necessary.

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