CASE REPORT

Synovial Arteriovenous Malformation Masquerading as Arthritis

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Background: Synovial arteriovenous malformation is rare. **Case characteristics**: We present three children with recurrent monoarthritis secondary to synovial arteriovenous malformation. **Outcome**: Two children underwent excision of arteriovenous malformation. Another child had diffuse arteriovenous malformation, which was inoperable. **Message:** Synovial arteriovenous malformations should be considered in the differential diagnosis of monoarthritis, especially of the knee.

Keywords: *Arthralgia, Misdiagnosis, Vascular malformation.*

Arteriovenous (AV) malformations are congenital vascular lesions composed of a complex tangle of arteries and veins connected by one or more fistulae. They are commonly seen in the brain and the spine and rarely in joints. Patients with AV malformations involving the joints may present with symptoms of joint pain, swelling (hemarthrosis) and limitation of movement [1]. These lesions can lead to chronic synovitis and progressive cartilage denudation leading to secondary osteoarthritis. We present three children who presented with recurrent knee joint swelling, and were diagnosed to have synovial AV malformations.

CASE REPORT

Case 1

A 4-year and 6-month old boy presented with recurrent swelling of the right knee joint since the age of 2 years. The patient on examination was noticed to have a purplish compressible swelling in the sole of the right foot at the base of the great toe. There was no history of similar complaints in the family members. Complete blood count, coagulation studies (prothrombin time and activated partial thromboplastin time) and erythrocyte sedimentation rate (ESR) were normal. Magnetic resonance imaging (MRI) of the affected limb revealed the AV malformation involving the whole right lower limb (***Fig.* 1**). The knee swelling was as a result of recurrent hemarthrosis secondary to synovial AV malformation. As the AV malformation was involving the whole lower limb (diffuse variety), it was not amenable to treatment with either surgical excision or sclerotherapy/

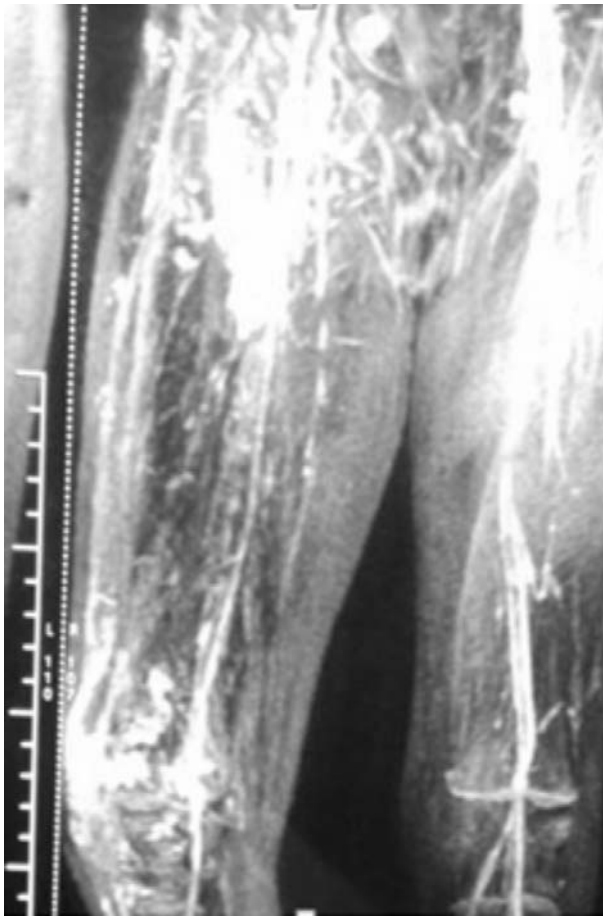

FIG.1 *MRI showing hypertrophy of the right lower limb with underlying AV malformation (dilated vessels infiltrating the subcutaneous fat,muscles, patella and knee joint).*

embolization. The patient continues to be under observation, and persists to have recurrent knee joint swelling. He continues to have periods of immobilization and school absenteeism because of recurrent hemarthrosis.

Case 2

A 5-year-old boy presented with recurrent right knee joint pain and swelling from infancy, which was brought about by trivial trauma. On examination, he had swelling and tenderness of the right knee joint. Complete blood count, coagulation studies and ESR were normal. *X*-ray of the right knee joint showed early erosive changes. MRI of the right knee joint revealed a synovial AV malformation. He underwent excision of the AV malformation. Histopathological examination was suggestive of hemangioma. He had complete resolution of the symptoms for the first 6 months post-surgery. Subsequently he was lost to follow-up.

Case 3

A 3-year and 5-month old boy presented with recurrent episodes of left knee pain and swelling that had been present for the last 2 years. He had complaints of early morning stiffness of the left knee joint. On examination, he had swelling and tenderness of the left knee joint. He was diagnosed as oligoarticular Juvenile idiopathic arthritis and was treated with methotrexate and folic acid. He had a persistent microcytic, hypochromic anemia, which did not improve significantly with iron supplementation. In view of recurrent monoarthritis with lack of response to methotrexate and persistent anemia, he was subjected to MRI, which revealed heterogeneously hyper-intense lesion on T2-weighted image in the anterolateral aspect of the left knee joint with post-contrast non-homogenous enhancement with small branches of popliteal artery supplying the lesion (***Fig.* 2**). The child underwent open synovectomy with *en-bloc* excision of the AV malformation. Biopsy was suggestive of hemangioma. Four months post-surgery, he was doing well.

DISCUSSION

Synovial AV malformations are rare in children [2]. They are usually congenital malformations but are often diagnosed later in life. They can be localized or diffuse in nature. Intra-articular AV malformations can arise from any synovial surface. Possibility of synovial AV malformations should be considered in patients having recurrent painful hemarthoses with normal coagulation parameters [3].

Synovial AV malformations commonly tend to involve the knee joint, but have also been reported in the elbow, wrist, tempo-mandibular joint and ankle [4]. The knee joint was involved in all our patients. The usual symptoms brought about by these synovial AV malformations include recurrent joint pain, joint swelling – either because of the AV malformation or because of the recurrent hemarthrosis caused by minimal trauma – and limitation of movement. Kassabach Merrit syndrome is a rare association with this type of AV malformation [5]. Synovial AV malformations cause premature destructive arthropathy secondary to repeated hemarthrosis. The differential diagnoses that need to be considered are pigmented villonodular synovitis, tuberculous monoarthritis, synovial sarcoma, other arthritis/ arthropathies (Juvenile idiopathic arthritis, hemophilic arthropathy, synovial osteochondromatosis) – usually distinguished clinically or on MRI [6].

Radiographs usually reveal a soft tissue swelling of the joint. Rarely phleboliths are seen in the joint. MRI is the investigation of choice for diagnosis and planning of the management strategies in this condition. MRI typically shows isointense or hypointense lesion in T1-weighted images and hyperintense in T2-weighted images [7]. The diagnosis was unmarked by MRI in all our patients. Whole body MRI to rule out AV malformations in other organs could not be done in view of cost-constraints.

Early surgical excision is indicated to prevent joint damage. Arthroscopic excision is done when the hemangioma is well circumscribed and pedunculated. Open excision is the treatment modality of choice for diffuse hemangioma, but carries a higher risk of recurrence

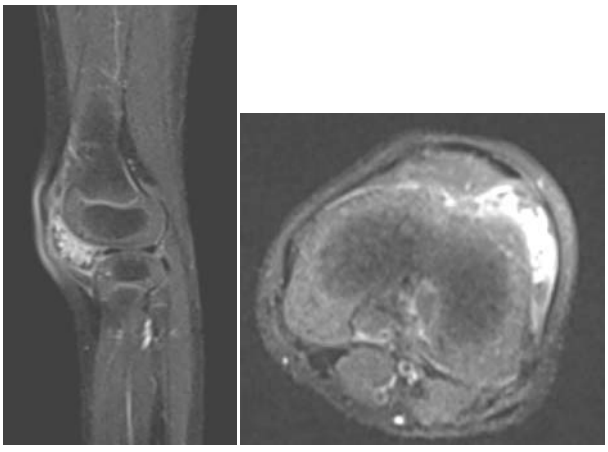

FIG. 2 *Sagittal and coronal T2-weighted MR image showing a heterogenous mass hyper intense lesion in the anterolateral aspect of the left knee joint with post contrast non-homogenous enhancement.*

[8]. Angiography defines the size, location of the lesion and can identify the feeder vessels associated with the AV malformation. High flow lesions require trans-arterial embolization whereas percutaneous sclerotherapy is typically used for low-flow lesions [8]. These are usually performed in cases of associated cutaneous hemangioma or abnormal varicosity because these findings are indicative of a more general vascular abnormality [9].

We conclude that pediatricians should consider synovial AV malformations in the differential diagnosis of recurrent joint swelling affecting a single joint.

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