

Joint related Complaints in Vascular Malformations (e.g. Hauer Disease) Management and Pitfalls

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PURPOSE

Patients with congenital vascular malformations often suffer from arthralgia, especially of the lower limbs. This orthopaedic disease pattern is defined as destructive, angiodysplastic arthritis or "Hauer Disease" (Hauer et al 2009). Because the motion pattern is developed during infancy, the reduced activity is often misinterpreted and leads to under-, over- or rather false treatment. To prevent the very early substantial joint destruction in infancy it is necessary to review the early use of orthopaedic surgical techniques and vascular surgery in order to improve quality of life and long follow up result.

METHODS

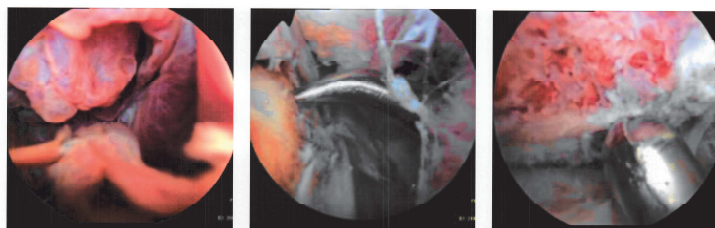
From our multidisciplinary team database of patients with vascular malformations affecting the limbs and the joints, we examined clinically all patients after combined orthopaedic and vascular surgery diagnosed as destructive, angiodysplastic arthritis. Control MRIs, radiographs and or control arthroscopies were performed and evaluated.

RESULTS

The 31 examined cases included 10 males and 21 females, ranging in age from 5 years to 68 years (mean 29,2 years). The average period of time between the first appearance of joint complains and the final diagnosis destructive, angiodysplastic arthritis was 10,7 years. 3 patients were managed conservatively (9,7%), 3 patients were treated with resective vascular surgery only (9,7%) and 25 patients received arthroscopic surgery in combination with vascular surgery (80,6%). In 8 cases the replacement of either the hip joint (3 patients, 9,7%) or the knee joint (5 patients, 16,1%) was necessary. All patients report a significant improvement of their complaints after treatment. These documentations correlated directly with the results of the imaging.

Diagnostic and therapeutic algorithm of Hauer Disease

	Stage I	Stage II	Stage III
n	6	11	14
Clinical Feature	synovial membrane	synovial membrane, cartilage	synovial membrane, cartilage, bone
Nativ Radiographic Findings	unspecific, phleboliths	irregularity of the subchondral border lamella, demineralization (regarding bilateral comparison)	subchondral sclerosis, mutilation
MRI	condensing of the synovial membrane, vascular malformations	cartilage loss, vascular malformations	cartilage destruction, vascular malformations
Symptoms	limping, soft tissue swelling, no/mild pain, mild decrease of ROM	functional deficit with pronounced decrease in ROM, mild to severe pain	sometimes severe pain, severe restriction in ROM or contractures
Therapy	conservative, diagnostic arthroscopy, partial transarthroscopic synovectomy	transarthroscopic (open) debridement, with or without redressing brace	transarthroscopic (open) debridement, with or without redressing brace, joint replacement



Transarthroscopic aspect: Intraarticular overgrowing malformation (Fig.1), Stage 2, chondropathy Grade 4 and „micro-fracturing“ (Fig.2), Stage 3, severe joint destruction (Fig.3)

Table of misdiagnoses of Hauer Disease in our cases (treated elsewhere) with the conducted non-indexed approaches:

•Rheumatism

- Juvenile idiopathic arthritis
- Seronegative rheumatism
- Pigmented villonodular synovitis (PVS)

•Pediatric (orthopedic) diseases

- Atraumatic haemarthrosis
- Haemophilia
- Thrombophilia
- Osteochondrosis dissecans
- Pott's disease
- Coxitis fugax

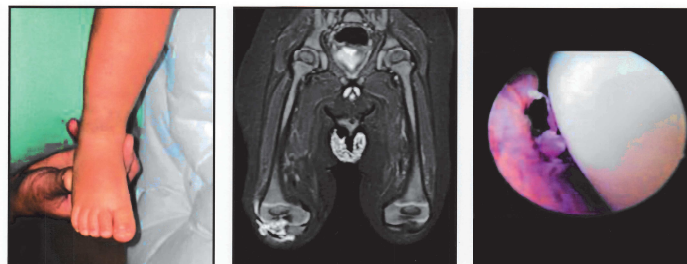
•Pathogen

- Borreliosis
- Arthritis in case of viral infection
- Empyema of the joint

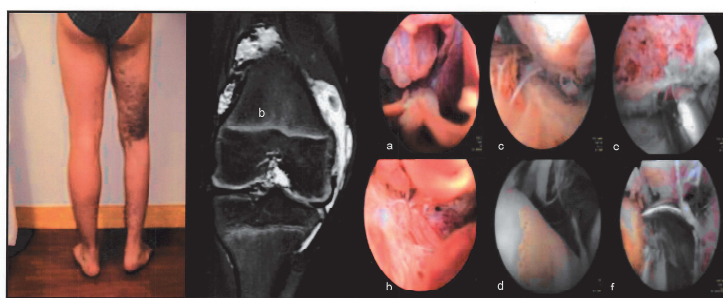
•Pain caused by malformation

•Degenerative arthrosis

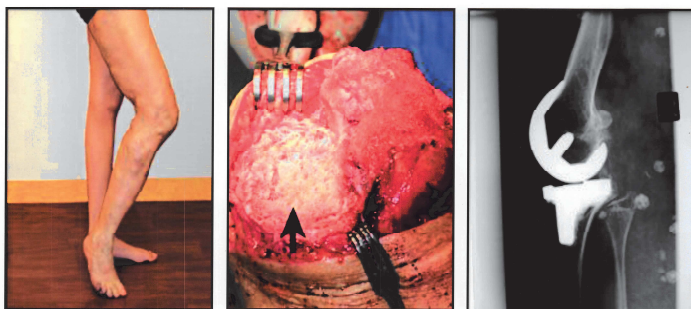
CASE REPORTS



Stage 1: 18 month old, male, clinical signs: reduced activity, recurrent knee joint swelling, movement restriction (ROM: E/F 0/10/140°) in combination with cutaneous malformation at the right ankle (Fig.1). Suspected diagnosis: haemarthrosis in case of haemophilia. Evidence of intraarticular malformation via MRI (Fig.2). Therapeutic approach with transarthroscopic resection of synovial membrane (Fig.3). Dramatical reduction of pain, extension of ROM.



Stage II: 19 year old young male complaining of intermittent gonalgia and progressive restriction in the range of motion of the right knee (fig. 1). ROM: 0/20/90°. Radiologic: enormous periarticular malformation (fig. 2, compared to the left knee), Arthroscopic: extensive intraarticular malformation (fig. 3 a,b), accumulation of hemosiderin in bone (fig. 3 c) and cartilage (fig. 3 d), bare bone under rudimentarily formed cartilage (fig. 3 e) or cartilage erosion (fig. 3 f). Significant improvement in joint mobility (ROM: 0/10/100°) and less pain after arthroscopic synovectomy and cartilage-smoothing.



Stage 3: 16-year-old female, predominantly venous malformation (Fig.1), advanced joint contracture of 60° despite long lasting therapy with debridement, joint mobilisation and physiotherapy. Combined with severe bone destruction operative approach with joint replacement and vascular surgery. Aspect with femoral bone necrosis and tibial intraspongiosa malformation (arrow) (Fig.2). 13 months post-op: considerable reduction of pain and enhanced ROM 0/5/100 (Fig.3).

DISCUSSION

Diagnostic and therapeutic algorithms emphasize the destructive, angiodysplastic arthritis as a possible differential diagnosis in order to minimize the risk of incorrect diagnosis which might lead to under-, over-, or even incorrect treatment. Thus pitfalls can be avoided. Accurate radiologic diagnosis, i.e. MRI, should be performed to ensure early intervention. A minimal invasive transarthroscopic therapy in combination with different vascular surgical tactics and techniques treating different species and forms of vascular malformation stages can lead to significant improvement of symptoms and prevention of progressive joint destruction in the early stages.

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