

Arthropathy in Patients with Vascular Malformations- Long term follow-up Results of a Rare Condition

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Introduction

Vascular malformations (VM) represent a challenge in diagnosis and treatment of orphan diseases. If these VM cause decreased joint mobility and/or pain, this may be accompanied by destructive joint changes. This entity is called Hauert's Disease (HD) or angiodysplastic destructive arthropathy and is classified into 3 stages – from only synovial thickening in MRI (grade 1) to decreased cartilage in grade 2 and destructed bone in grade 3. With this long-term follow-up we describe the outcome of this condition.

Diagnostic and therapeutic algorithm of Hauert Disease

	Grade 1	Grade 2	Grade 3
Clinical Feature	synovial membrane	synovial membrane, cartilage	synovial membrane, cartilage, bone
Nativ Radiographic Findings	* unspecific, phlebolithis	irregularity of subchondral border lamella, demineralization	subchondral sclerosis, multilication
MRI	condensing of synovial membrane, vascular malformation	cartilage loss, vascular malformation	cartilage destruction, vascular malformation
Symptoms	limping, soft tissue swelling, no/mild pain, mild decrease of ROM	functional deficit with pronounced decrease of ROM, mild to severe pain	sometimes severe pain, severe restriction in ROM or contractures
Therapie	conservative, partial transarthroscopic synovectomy	transarthroscopic (open) debridement, with or without redressing brace	Transarthroscopic (open) debridement, with or without redressing brace, joint replacement

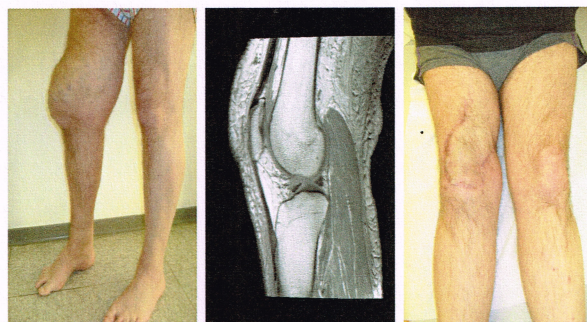
Methods

The files of patients treated for HD in our institution from 1983-2017 were collected in retrospective study. Medical data was entered in a MS Excel file. For evaluation of the joint damage, native x-ray, MRI and arthroscopy was conducted.

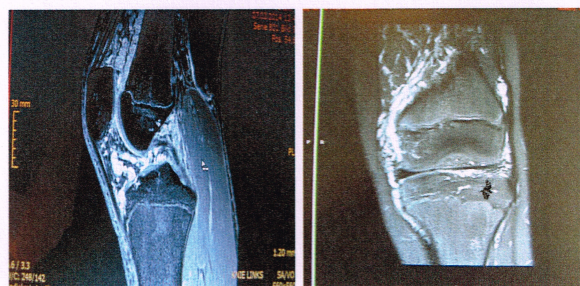
Follow up was taken out by examination of the patients. The patients were classified according to the extent of the VM, localization of the arthropathy and orthopedic procedures performed.

Results

50 patients (34 female, 16 male) with HD were diagnosed. The disease was isolated to one joint in 48 cases (2 shoulder, 3 hip, 41 knee, 2 ankle) and found in multiple joints in 2 cases (both knee + ankle). Out of the 48 single-joint-cases 14 were grade 1, 13 grade 2 and 21 grade 3. One of the poly-joint-case showed grade 3 in both joints, the other grade 3 in one and grade 1 in the other. Besides numerous vascular interventions, the patients were treated with arthroscopic debridement (n=29) or by joint replacement (n=10; all of them were grade 3). The other patients either did not require debridement (n=7) or due to excessive VM orthopedic treatment was not possible (n=4).



Grade 1: 42 year old male patient with monstrous extratuncular malformation (left fig.), intraarticular shows only little malformation compared to extraarticular (center fig.), treatment is only vascular surgery (right fig.).



Grade 2: 18 year old male patient with progressive restriction in the range of motion, parietal malformation. Treatment by arthroscopic synovectomy and cartilage-smoothing.



Grade 3: 18 year old female patient treated by core decompression (forage) by incomplected diagnosis of femoral head necrosis (left fig.), combined vascular surgery und cementless joint replacement (center & right fig.).

Conclusion

Angiodysplastic destructive arthritis is a rare disease. Orthopedic treatment is good option to improve the patients mobility with excellent long-term results. The classification suggested by Hauert seems reasonable to compare cases.

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Literature

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