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Orthopedic issues in vascular anomalies

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ABSTRACT

Vascular malformations impact the musculoskeletal system depending on the tissue involved (skin, subcutis, muscle, cartilage, or bone), the extent of involvement, and the type of anomalous vessels (arteries, capillaries, veins, or lymphatics). These malformations can cause a multitude of musculoskeletal problems for the patient and their Orthopedic Surgeon to manage. Leg-length discrepancy, intra-articular involvement, muscular lesions, and primary or secondary scoliosis are just to name a few. All of these problems can cause pain, deformity, and a range of functional limitations. Surgical and nonsurgical treatment plans both have a role in the care of these patients. Patients with vascular malformations may also suffer from life-threatening cardiovascular and hematologic abnormalities. For those patients who undergo surgery, thromboembolic risk is elevated, wound breakdown and infection are much more common, and bleeding risk continues well into the postoperative course. Because of the complex nature of these disorders, the clinician must have a full understanding of the types of lesions, their natural history, appropriate diagnostic studies, associated medical problems, indications for treatment, and all the treatment options. For severe malformations, especially syndromes such as CLOVES and Klippel–Trenaunay syndrome, interdisciplinary team management is essential for the best outcomes.

Introduction

Vascular malformations impact the musculoskeletal system dependent on the tissue involved (skin, subcutis, muscle, cartilage, or bone), the extent of involvement, and the type of malformation (arteries, capillaries, veins, or lymphatics). These malformations can cause a multitude of musculoskeletal problems for the patient and their Orthopedic Surgeon to manage. Leg-length discrepancy, intra-articular involvement, muscular lesions, and primary or secondary scoliosis are common issues. These problems can cause pain, deformity, and a range of functional limitations. These can be treated with interventional and non-interventional supportive means. Non-operative management includes shoe lifts to balance leg-length discrepancy, physical therapy to stretch joint contractures, and bracing for scoliosis. Surgical treatment ranges from minor procedures such as guided growth/epiphysiodesis for leg-length discrepancy or percutaneous Achilles lengthening for equinus contracture to major endeavors such as spinal fusion for scoliosis, joint replacement, or amputation.

Patients with vascular malformations may also suffer from life-threatening cardiovascular and hematologic abnormalities. Postoperatively, thromboembolic risk is elevated, wound breakdown and infection are much more common, and bleeding risk continues well into the postoperative course. Due to the complex nature of these disorders, the clinician must have a full understanding of the types of lesions, their natural history, appropriate diagnostic studies, associated medical issues, indications for treatment, and all the treatment options. For severe malformations, especially syndromes such as CLOVES and Klippel-Trenaunay (capillary lymphatico-venous malformation), interdisciplinary team management is essential for the best overall outcomes.

Intra-articular lesions

Intra-articular lesions are generally venous malformations or lymphatico-venous malformations that invade the joint space. Patients with intra-articular vascular malformations frequently present with joint pain, swelling, stiffness, contractures, weakness, and hemarthrosis.[1–3] Intrinsic joint symptoms are primarily due to intra-articular vascular malformations. Intra-articular vascular malformations can affect joint function by the formation of an intra-articular mass or arthritis. The arthritis may be due to generalized synovial inflammation or rapid cartilage degeneration similar to that seen in hemophiliac induced arthritis. Arthropathy can be a serious complication of repeated joint bleeding in patients

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Fig. 1. Lateral x-ray of a knee of a patient with an intra-articular vascular malformation showing the presence of an intra-articular mass.

with hemophilia and vascular malformations and can result in chronic pain, deformity, and functional impairment. Although the pathogenesis of this type of arthropathy is not fully understood, it appears to have similarities with the degenerative joint damage that occurs in osteoarthritis and the inflammatory processes associated with rheumatoid arthritis. The mechanism begins with an intra-articular bleed, which results in an enzymatic cascade that seems to affect multiple sites within the joint. In the end, it results in fibrosis of the synovial lining and disintegration of the hyaline cartilage. This may occur after one or two bleeds in predisposed patients or may take multiple bleeds in those that have some unknown form of protection.[4–8]

Findings on plain x-ray may show soft tissue phlebitis, signs of articular joint expansion either due to an effusion or a soft tissue mass. Late findings would include symmetric joint space narrowing, sclerosis, subchondral cysts, and other findings consistent with that of arthritic changes (Figure 1). MRI demonstrates an intra-articular vascular lesion (Figure 2). This lesion may be focal or diffuse. A bloody effusion is usually seen with varying degrees of articular surface damage.[9,10]

Fig. 2. Sagittal MRI of a knee of a patient with an intra-articular vascular malformation demonstrates the presence of an intra-articular mass and cartilaginous changes.

Treatment

Non-operative treatment includes physical therapy to improve motion and strength. This is particularly important in patients with extra-articular cause of dysfunction such as a quadriceps or hamstring contracture. Compression garments are also important to control swelling. These modalities along with immobilization are also essential in treating patients with acute bleeds. Sclerotherapy is often used as the primary treatment for soft tissue vascular malformations. It has been shown to be effective in reducing pain and reducing the size of the mass thereby increasing the patient's function.[8] However, its use in the treatment of intra-articular lesions is somewhat limited. Due to the numerous feeding vessels, it is difficult to treat the entire intra-articular component with sclerotherapy alone. However, sclerotherapy can be used as an adjunct to surgery and has been used to reduce surgical blood loss.[11–13]

Surgical treatments include synovectomy and joint arthroplasty. Before embarking on any surgical treatment of vascular malformations, one must have a full understanding of its natural history as well as the appropriate diagnostic studies, associated medical issues, and indications. Since many of these patients have coagulopathies, tourniquet use may or may not be contraindicated.[14] If a tourniquet is not to be used, bleeding may be difficult to control, and the patient may experience high blood loss. If a patient has involvement of the skin and subcutis, wound healing problems may occur resulting in an increased risk of infection. However, with collaborative, multidisciplinary care, these procedures can be performed safely and effectively on even high-risk vascular malformation patients.

Joint synovectomy is the mainstay of treatment for intra-articular involvement to reduce spontaneous bleeding episodes, articular cartilage damage, and the associated poor patient outcome. It may be performed in an open or arthroscopic fashion.[15] An open procedure is generally preferred to ensure a complete synovectomy and allow easier control of intraoperative and post-operative blood loss. For patients with localized disease as seen in nodular pigmented villonodular synovitis, excision of the lesion is usually curative. Patients with diffuse disease tend to have a worse prognosis. These patients tend to have hemosiderin-stained tissue and varying degrees of cartilage damage can be seen at the time of surgery (Figure 3). Although synovectomy is a commonly used

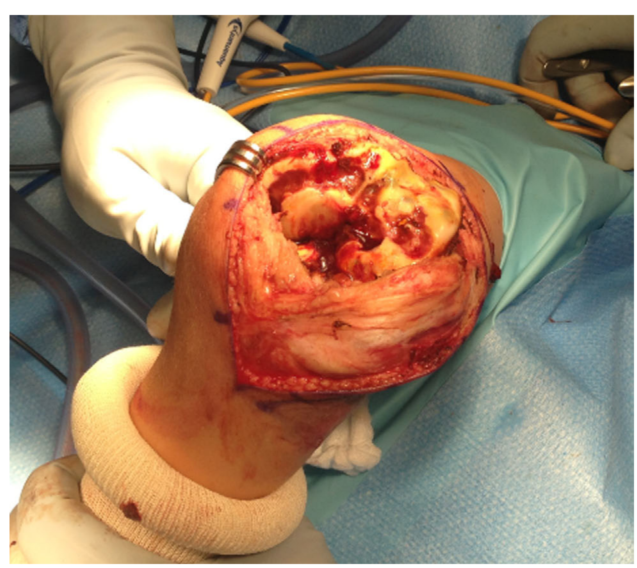

Fig. 3. Intraoperative photograph of a knee showing cartilage defects. (Color version of figure is available online.)