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Scattered Musculoskeletal Venous Malformations in Children: A Rare Report on Clinical Evaluation and Sclerotherapy with Adjunctive Stasis of Efflux

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Learning Point of the Article:

Clinical presentation, evaluation and treatment planning in patients with musculoskeletal venous malformations.

Abstract

Introduction: Venous malformations are rare lesions of unknown etiology, with a reported incidence of 0.8–1%. Patients with inexorable growth and expansion of vascular malformations, or" have an unpredictable clinical course and a wide range of presenting symptoms. Often, they are erroneously diagnosed and inadequately treated due to their rarity and lack of expertise among clinicians. To author's information this is the first report of diffuse venous malformations with multiple phleboliths involving various compartments of the upper extremity in children.

Case Report: The uthors discuss the clinical presentation, evaluation, and treatment over 8 months of slow-flow venous malformations with phleboliths in an11-year-old girl presenting with multiple painful swellings throughout her right upper extremity. The right upper extremity had multiple swellings over the right hand, forearm, arm, and shoulder region involving multiple compartments. The digital swellings had bluish discoloration, indicating a vascular nature. Blood tests revealed a raised D-dimer level (2.42 mg/L). Radiographs, Ultrasound, Magnetic resonance imaging, and CT angiography suggested a slow-flow venous malformation. The excisional biopsy confirmed the diagnosis. Ultrasound-guided Sclerotherapy with the Sclerotherapy with Adjunctive Stasis of Efflux Technique was performed for other lesions. Sodium Tetradecyl Sulfate (60 mg/2 mL; 0.5mL) was used in each lesion. Post-intervention, at 6 months follow-up, cosmetic appearance improved drastically, with the hands benefitted most. Parents were satisfied with overall outcome. Sclerotherapy was stopped after 4 cycles.

Conclusion: Ultrasound-guided sclerotherapy is effective in treating venous malformations. The ideal result is seen after 4–5 sittings. Sclerotherapy must be performed in the operating theatre under sedation or appropriate anesthesia with resuscitation equipment at the ready disposal. Excision is reserved for bigger superficial lesions.

Keywords: Vein, Child, Sclero therapy with Adjunctive Stasis of Efflux, India, Scero therapy.

Introduction

Venous malformations are rare lesions of unknown etiology, with a reported incidence of 0.8–1% [1]. Believed to be "embryologically originated mistakes of vascular morphogenesis," they present as localized or diffuse structural abnormalities [2] of affected vascular territory.

Patients with inexorable growth and expansion of vascular malformations have an unpredictable clinical course and a wide range of presenting symptoms [3]. Often, they are erroneously diagnosed and inadequately treated due to their rarity and lack of expertise among clinicians [4].

Slow-flow venous malformations occasionally present with

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Figure 1: Initial clinical presentation.

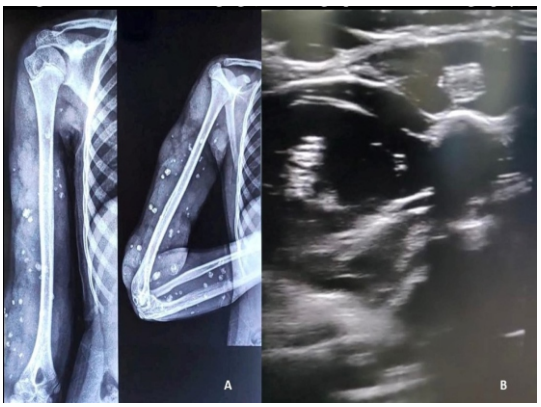

Figure 2: Non-Invasive Diagnostics: Radiograph (green); Ultrasonography (red).

"phleboliths," considered a complication [5]. These are intravascular calcifications forming within the malformation due to venous stasis. Various studies have reported phlebolith associated with venous malformations, but they are mostly restricted to oral and maxillofacial regions such as the buccal fat pad [5], maxillofacial region [6], submandibular region [7, 8], and salivary gland [9].

Malformations grow rapidly with advancing age; thus, it is crucial to evaluate them, establish a diagnosis, rule out malignancies, and treat them early, as their rapid growth may affect children's social acceptability and the movements of affected limbs and joints. Children present with pain and discomfort due to expanding blood collection and limb heaviness.

For the author's information, this is the first report of diffuse venous malformations with multiple phleboliths involving various compartments of the upper extremity in children. The authors discuss clinical presentation, evaluation, and treatment over 8 months in an 11-year-old girl presenting with multiple painful swellings throughout her right upper extremity and hand.

Case Report

An 11-year-old girl was brought to us by relatives with complaints of pain in the right upper extremity with restricted elbow and hand movements for 4 weeks. There was a history of injury on the right hand six years ago. The girl was normal-built for her age. The swelling had not been present since birth; the first swelling appeared in the dorsum of the hand. This was observed after the injury; later; the swelling increased in size and became painful; subsequently, multiple swellings appeared, initially painless. Gradually, all swellings grew to their present size, with the ones in the hands and arms being painful. She was not able to freely move her wrist or elbow due to the increased weight of her upper extremity.

The right upper extremity had multiple swellings over the right hand, forearm, arm, and shoulder region involving multiple compartments (Fig. 1). The digital swellings had a bluish discoloration. On examination, the hand and forearm swellings were compressible and non-tender, while those in the forearm, arm, and shoulder were firm and non-tender. On palpating the swellings in the forearm and arm, a single hard, pea-like mass could be felt within. There was no temperature rise. The skin over the swelling was normal, and the surface was smooth. There were no pulsations. The margins of the swelling were distinguishable from surrounding tissue, and the plane was superficial in the hand but deep in the muscles in the forearm and arm. The systemic examination was unremarkable. Clinically, the swelling appeared to be vascular, and not related

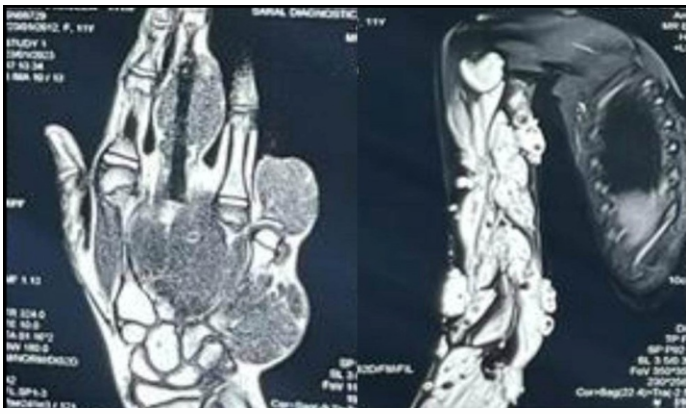

Figure 3: Non-invasive diagnostics: MRI.

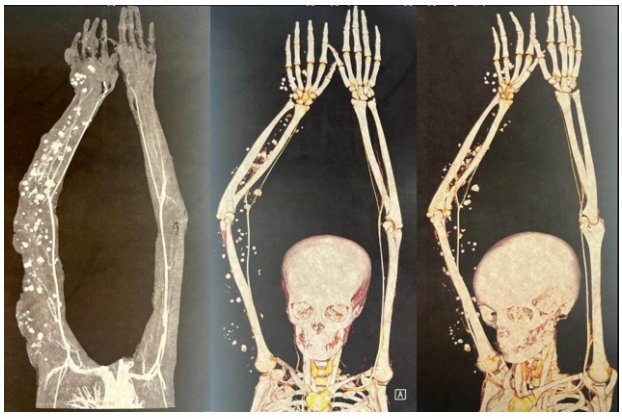

Figure 4: Non-invasive diagnostics: CT angiography.

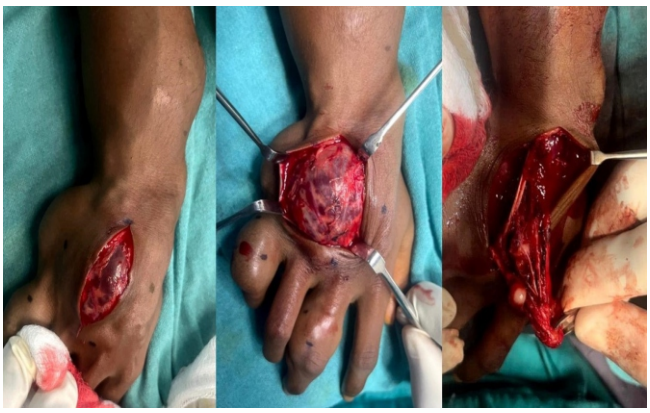

Figure 5: Intraoperative pictures: Biopsy.

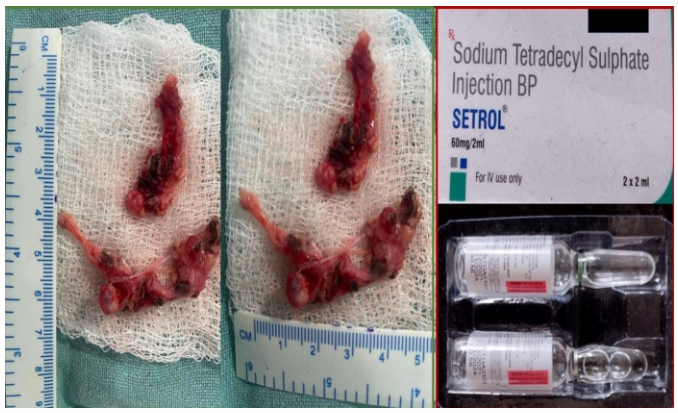

Figure 6: Histopathology Study Specimen (green); Sclerosant Ampoules (red).

to a single vessel.

Blood tests revealed raised D-dimer levels (2.42 mg/L) and Fibrin degradation products (0.16 mg/L); rest all parameters were within normal limits. Non-invasive diagnostics were performed before proceeding with biopsy and therapeutic procedures.

Serial radiographs of the upper extremities revealed multiple small circular calcified masses in the arm, forearm, and hand. Mases appeared to be superficial and contained in muscular compartments. Bones and Joints were spared (Fig. 2a).

Ultrasound revealed thin-walled fluid blood filled spaces, and a phlebolith-laden lesion showed an echogenic shadow, that was freely mobile within a hypoechoic vascular lesion; doppler ultrasound confirmed its venous nature (Fig. 2b).

Magnetic resonance imaging was performed to delineate the nature of the swelling, plane and rule out malignancy. The swellings were confined to muscle, tendon, and subcutaneous regions. No involvement of bones and joints was appreciated. The scan confirmed lesions to be well defined and scattered, involving all compartments of the arm, forearm, and hand. No malignant features were seen (Fig. 3).

CT Angiography revealed an innumerable multispacial heterogeneous density lesion involving all compartments of the right arm, forearm, and hand with intrinsic phleboliths. Dynamic post-contrast imaging revealed early nodular enhancement with slow progressive enhancement on venous delayed phase images. No arterial feeder, nidus, or draining vein was identified, thereby ruling out a high-flow vascular malformation (Fig. 4).

With this information, a therapeutic protocol was outlined. An excisional biopsy was planned for the lesion on the dorsum of the hand (Fig. 5). Ultrasound-guided Sclerotherapy with the Sclerotherapy with Adjunctive Stasis of Efflux (STASE) Technique was planned for the remaining lesions. Potential advantages and risks were explained to family members and documented. Procedures were performed in a standard operating room under general anesthesia. A high-quality ultrasound set at musculoskeletal or small-part settings with a sterilely prepped transducer was secured into the operating field.

A biopsy specimen (Fig. 6) revealed an admixture of malformed blood vessels (veins) with the presence of calcification in a few of them. Irregularly thickened wall of dilated vascular spaces the phlebolith section showed a fibro-collagenous lining of laminated concentric rings of calcification. No malignancy features were seen (Fig. 7).

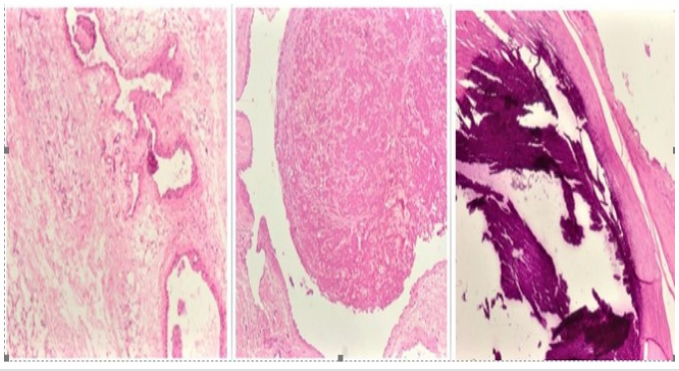

Figure 7: Histopathology microscopic pictures (H&E staining).

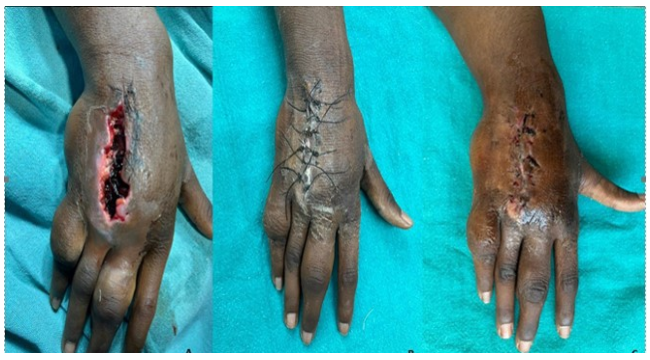

Figure 8: Clinical images: Wound dehiscence (a); Debridement and Re-closure (b); Healed suture line (c).

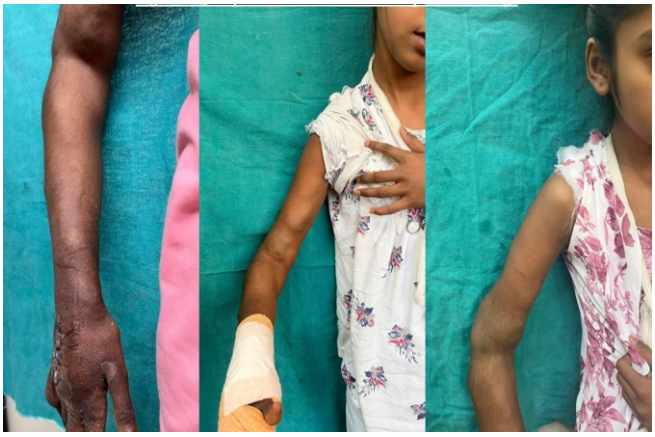

Figure 9 : Clinical pictures: 2-week review.

Figure 10: Clinical pictures: 8-week review.

Sclerotherapy was done by the STASE Technique [10] under Ultrasound Guidance. The sclerosant used was Sodium Tetradecyl sulfate (Fig. 6), 0.5 mL in each lesion on all sittings. In the digits, sclerosant was injected on the medial and lateral sides alternatively. The sclerosant was loaded in 2 cc syringes, lesions were punctured sequentially from distal to proximal (hand to shoulder) by a 4 cm or 7 cm, 21-gauge needle under USG guidance, blood was evacuated by manual compression, and immediately the sclerosant was pushed in and a compressive gamgee was applied, followed by a crepe compressive bandage from distal to proximal. Procedure done under standard antibiotic cover. Relook was done in 5 days. A repeat procedure was done at 3-weeks intervals. Four sclerosant therapy cycles were performed over three months. The post-therapy period was uneventful; no skin complications were encountered.

The biopsy site showed wound dehiscence over 10 days after the procedure; closure was repeated, and the wound healed over the next 2 weeks (Fig. 8). Follow-ups were otherwise uneventful, with a gradual reduction in the size of the swellings (Fig. 9).

At 6th and 7th month follow-up after the primary procedure, upper extremity cosmetic appearance improved drastically, and hands benefitted most (Fig. 10). Overall, the limb was thinner and lighter. The young girl could relate to her limb more now. Her self-propelled wrist and elbow movements reached functional range. Parents were satisfied with the clinical outcome and enhanced cosmesis; the sclerotherapy sessions were stopped after 4 cycles on their request .

Discussion

These cases are rare citing in orthopedics outdoor department, especially in the pediatric population. In our case, the patient was referred to us by Pediatric Department. On first look, the multiple swellings involved multiple compartments of the arm, elbow, forearm, wrist, and hand; thus, they could not be attributed to any particular dermatome, muscle, or joint. The lesion in the hand was superficial and tense with bluish discoloration, indicating blood within, but the ones in the forearm, arm, and shoulder were deep; only a bulge could be appreciated. Clinically, the lesions looked vascular; the patient's attendants were counseled to get a specialist opinion from a high-volume pediatric hospital, a vascular or plastic surgeon, first, but they were unable to get a clear picture. After a delay of 2 months from the initial presentation, parents requested the orthopedic team treat the child.

Clinicians are unaware of the treatment of musculoskeletal venous malformations due to their rarity. Although various reports from dental and maxillofacial specialties have discussed the treatment of oral venous malformations and phleboliths, their suggested options include observation, surgical excision, and laser therapy in a few patients. The musculoskeletal and global involvement of the upper extremity makes this case rare. In these rare conditions, no definite treatment algorithm has been suggested, and reports are limited to clinical presentation and diagnostics.

Phlebolith, or "Calcified Thrombii," as a characteristic feature of a vascular lesion was first reported in the splenic vein [7], with the submandibular region being the commonest site [8]. Changes in Blood flow dynamics or vascular stasis within the dilated vascular spaces are primarily responsible for phlebolith formation. Subsequently, thrombus, its organization, dystrophic calcification [6], and lamellar fibrosis follow [9]. Sano et al., through X-ray diffraction and Infrared spectrometry, demonstrated Calcium carbonate and Calcium Phosphate as major phlebolith stone components [8, 11]. On the cut section, phleboliths are laminated with a radiopaque center with concentric rings of calcification, giving them an

"onion skinning-like" appearance.

According to the ISSVA 2014 Guidelines, Venous Malformations are classified as "Simple Vascular Malformations" and "Slow Flow" considering blood flow characteristics [12]. They demonstrate a slow, steady growth pattern that is commensurate with the growth of the child, and further, they also never involute. Angiography is the best diagnostic modality to confirm the diagnosis and establish the extent of the disease.

While treating pure venous lesions, clinicians must evaluate the patient for localized intravascular coagulation (LIC) and coagulopathy. Sclerotherapy can sometimes aggravate factors favoring the LIC and Coagulopathy cascade [10]. Compression garments, low molecular-weight heparin or direct anti-Xa agents are helpful in these patients [13, 14]. These patients are also likely to develop painful phleboliths, thrombophlebitis, Thromboembolic events [15, 16], or a full-blown DIC. The sclerosant is a heavy molecule that, when injected, can be painful and make the patient uncomfortable. Therefore, sclerotherapy procedures must be performed in the operating theatre under sedation or appropriate anesthesia with resuscitation equipment at the ready disposal.

The history of injury and appearance of swelling in our patient appear to be coincidental. We believe the swelling must have been present since a younger age, but it was observed only after the trauma incident when people were more attentive and observant, especially in people living in rural areas. Our patient could now play with her friends like other children. She can now attend school and wear regular clothing without people bothering her about her clinical condition. Her overall social acceptability improved. We thank our patient for her condition, and the data will help clinicians treat their patients in a better way and thereby benefit society on a larger scale.

Conclusion

Ultrasound-guided sclerotherapy is effective in treating venous malformations. The ideal result is seen after 4–5 sittings. Sclerotherapy must be performed in the operating theatre under sedation or appropriate anesthesia with resuscitation equipment at the ready disposal. Excision is reserved for bigger superficial lesions.

Clinical Message

Patients with vascular (venous) malformations in the extremities do visit orthopedic clinics. Mostly children, they should be properly evaluated clinically and with diagnostics. A biopsy establishes a diagnosis and rules out malignancies. Sclerotherapy can be performed by Orthopedic surgeons. Better results are achieved if the procedure is done under ultrasound guidance. Cosmetic Appearance dictates the end point of treatment.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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