Multiple giant venous malformations

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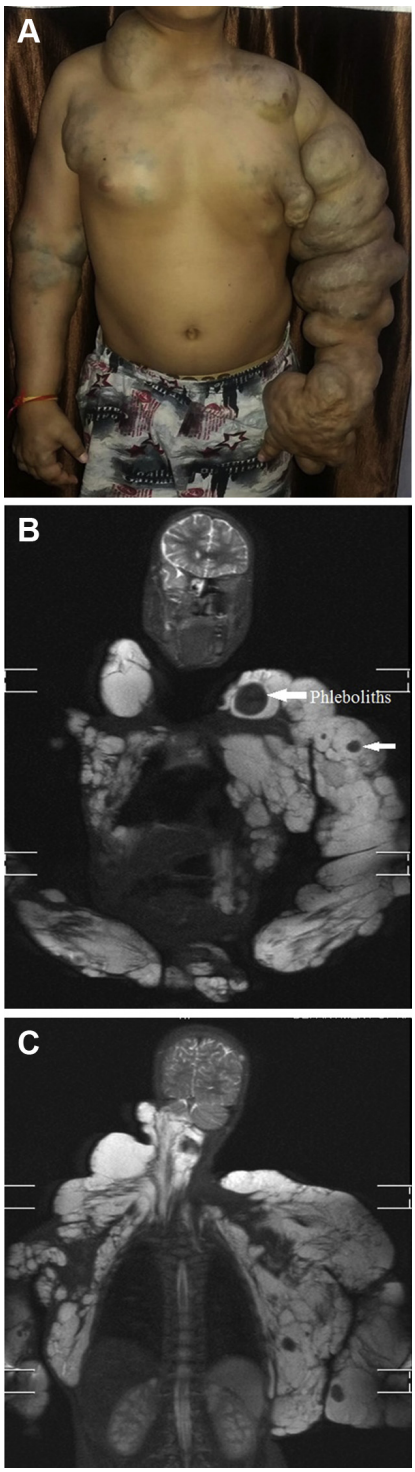
A 14-year-old boy presented with multiple large bluish swellings of both upper limbs, neck, and chest. These swellings were small at birth and became extensive with age (*A*). The lower limbs were spared. There was no family history of vascular anomalies. The lesions were soft, without any thrill or bruit, and they emptied on elevation and compression.

Magnetic resonance imaging of the whole body was performed. Coronal short-tau inversion recovery images (*B* and *C*) showed multiple large, T2-hyperintense lesions with multiseptated cystic-appearing spaces and phleboliths within (*B*, *arrows*, and *C*/Cover) involving the chest wall (anteriorly and posteriorly) and upper extremities. The lesions involved almost all the muscle compartments and neck spaces, projecting into the pharyngeal mucosal space and encasing the mediastinal structures. In the abdomen, similar lesions were seen in the retroperitoneum. The lesions showed gradually progressive filling in of the spaces on dynamic time-resolved magnetic resonance angiography. Diagnosis of multiple giant and extensive venous malformations was made.

According to the International Society for the Study of Vascular Anomalies classification,[1] these were simple venous malformations; and according to the Hamburg classification, they corresponded to extratruncular venous defects of infiltrating and diffuse subtype.[2] There was absence of bone overgrowth or undergrowth, dilated superficial veins, or any focal bone lesion (enchondroma) to suggest syndromic associations like Servelle-Martorell syndrome, Klippel-Trénaunay syndrome, or Maffucci syndrome. Proteus syndrome was also unlikely as it comprises asymmetric body growth, macrodactyly, and cerebriform connective tissue nevi in addition to venous malformations.[3] Overall prevalence of low-flow vascular malformations is approximately 1%, with extremity and head and neck involvement in 80% of cases and trunk involvement in 20%.[4] This kind of extensive involvement of blood-filled vascular lakes without syndromic association is rare. Treatment options include multiple sessions of sclerotherapy with polidocanol or sodium tetradecyl sulfate solution or oral sirolimus,[5] although these patients usually respond poorly to any kind of management.

The paper is in concordance with Institutional Review Board ethics, and required consents were obtained. The patient's parent agreed to publication of the case details and images.



REFERENCES

1. International Society for the Study of Vascular Anomalies. Classification. Available at: https://www.issva.org/classification. Accessed May 6, 2020.
2. Loose DA, Mattassi RE. Hamburg classification: vascular malformation. In: Kim YW, Lee BB, Yakes WF, Do YS, editors. Congenital vascular malformations. Berlin: Springer; 2017. p. 51-4.

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3. Nozaki T, Nosaka S, Miyazaki O, Makidono A, Yamamoto A, Niwa T, et al. Syndromes associated with vascular tumors and malformations: a pictorial review. Radiographics 2013;33:175-95.
4. Flors L, Salinas CL, Maged IM, Norton PT, Matsumoto AH, Angle JF, et al. MR imaging of soft tissue vascular malformations: diagnosis, classification, and therapy follow up. Radiographics 2011;31:1321-40.
5. Dubois J, Soulez G, Oliva VL, Berthiaume MJ, Lapierre C, Therasse E. Soft tissue venous malformations in adult patient: imaging and therapeutic issues. Radiographics 2001;21:1519-31.

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