Shining Pearls Sign: A New Identity for Venous Malformations on Computed Tomographic Imaging

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Abstract

Vascular malformations, in particular venous malformations (VM), are common lesions involving the pediatric and adolescent population. VM occur at approximately 1:5,000 to 10,000; approximately 40% of them occur in the head and neck regions. Classical appearance of VM on imaging is a demonstration of near-normal-sized or mildly dilated feeding arteries which subsequently lead to dilated venous structures of varying caliber, thus constituting the malformation. Phleboliths are the hallmark of VM. Plain radiography has been an established modality for demonstrating phleboliths. The emergence of computed tomographic (CT) as a preferred imaging modality for the evaluation of a complex vascular malformation necessitates familiarity with the spectrum of CT appearances of this lesion. This presentation illustrates an additional CT sign, “shining pearls sign,” highlighting the striking display of phleboliths in the vascular malformations, prompting correct diagnosis.

Keywords

- venous malformation
- shining pearls sign
- phlebolith
- CT

Sluggish blood flow in the venous malformation (VM) leads to stagnation, thrombosis, and subsequent formation of phleboliths, which represent calcified thrombi. Phleboliths are ubiquitously seen in many regions of body where chronic venous thrombosis occurs. Typical locations where phleboliths are observed are in the female pelvis as a sequel of postpartum thrombosis of pelvic veins or in large varicosities in lower limbs. Large mixed vascular malformations also associated with phleboliths in soft tissues. Soft tissue VM are identified and diagnosed by the presence of phleboliths. Plain radiography is losing its primary role in this imaging context to magnetic resonance imaging (MRI) and multi-detector computed tomography (MDCT), which have become the mainstay for diagnosing many soft tissue masses including VM.

MRI also has a greater role in the differentiation and characterization of many soft tissue masses. The purpose of this illustration is to highlight the role of volume rendered

reconstructions of CT data for the demonstration of phleboliths. A new sign has been described to aid in the diagnosis of VM by identifying the presence of phleboliths within the lesion. Three clinical examples of VM with phleboliths are included to illustrate the point.

Case Series

Case 1

An 11-year-old female patient presented with a gradually increasing soft tissue swelling on the right side of the face. She had difficulty in eating and had limited mouth opening. On examination, dilated vessels were visible over the right cheek. CT evaluation was recommended on suspicion of a complex vascular mass with bony involvement. MDCT demonstrated a large cluster of vascular channels supplied by the branches of the external carotid arteries from both sides. Dilated veins were also visualized in the early and the late arterial phase,

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Fig. 1 Late venous phase of coronal computed tomographic reconstruction demonstrating a large venous malformation involving the masticator space, infra-temporal fossa with extension into oral cavity. Note extensive expansile lytic lesion involving the mandible. (a) A few phleboliths are also demonstrated within the soft tissues. (b) Three-dimensional rendered images illustrate the classical appearance of multiple pearls due to phleboliths in the soft tissues.

thus confirming a VM. Mandible was deformed by extensive involvement by the malformation. The constellations of phleboliths seen on the three-dimensional (3D) image resemble a cluster of pearls of varying size, thus constituting the “shining pearls sign” (→Fig. 1 a,b).

Case 2

A 30-year-old male patient presented with a difficulty in swallowing and a diffuse swelling of the neck for the past few months. On clinical examination, a diffuse nontender soft tissue swelling was noted on left side of the neck. Transoral examination revealed a soft tissue mass bulging from the left

lateral pharyngeal wall and adjacent tonsil. CT evaluation was recommended after a clinical suspicion of a mass lesion from the oropharynx. MDCT images revealed multiple phleboliths in the soft tissues, which are typically illustrated in the 3D rendered images as multiple shining pearls (→Fig. 2a–c). Subsequent assessment of the total extent of the lesion and secondary changes in the surrounding area was completed with evaluation of the multiphasic dynamic CT data.

Case 3

A 29-year-old male patient presented with right-sided chest pain that radiated to the right upper limb. Chest X-ray was

Fig. 2 Coronal and sagittal images of the neck demonstrating extensive venous malformation involving the oro and nasopharynx. (a–c) There are foci of calcification within the soft tissues, mostly lying behind the sternomastoid muscle. Three-dimensional rendered image demonstrating a string of multiple pearls representing the phleboliths, illustrating the shining pearls sign.

Fig. 3 Contrast-enhanced axial computed tomographic image demonstrating the subtle soft tissue lesion in relation to the first rib. A small phlebolith is noted lying medial to first rib. (a, b) Three-dimensional rendered volumetric image illustrates the shining pearls sign by revealing three small phleboliths within the soft tissues.

unremarkable. CT evaluation demonstrated an irregular osteolytic lesion involving the right first rib. There were a few rounded calcifications in relation to the first rib. In addition, a vague enhancing soft tissue mass was noted in relation to the first rib. 3D rendering of the region of interest demonstrated the aforementioned shining pearls (►Fig. 3a, b). Subsequent MR evaluation confirmed a T2-hyperintense lesion, consistent with a hemangioma. The patient underwent surgical removal of the lesion with the confirmed diagnosis of a cavernous hemangioma.

All the CT images were acquired using 5-mm spiral acquisition data with subsequent reconstruction using 0.0625 mm thickness. Philips IntelliSpace portal (Koninklijke Philips Electronics NV, Amsterdam, The Netherlands) was used for 3D rendering. Presets for head and neck using carotid and cervical spine protocols were used. Images were carefully adjusted to the optimal setting for the demonstration of the phleboliths.

Discussion

Significance of phleboliths in the diagnosis of a vascular malformation, particularly of the venous type, is well established.¹⁻³ Exact incidence of phleboliths in VM is not reported, despite of numerous case reports demonstrating multiple phleboliths in VM.^{4,5} Approximately 15 to 25% incidence of phleboliths are reported in intramuscular hemangioma.^{6,7} We have encountered 4 patients with phleboliths of 11 in the age groups between 11 and 30 years. Plain radiography played a main role in demonstration of soft tissue calcification. However, cross-sectional imaging techniques such as CT have replaced plain radiography in many situations. CT technology is well known for its better sensitivity in the detection of subtle soft tissue calcifica-

tion. Sectional CT images demonstrate areas of calcification and phleboliths as rounded areas of high Hounsfield units. Also, CT is more sensitive than MRI in detecting phleboliths.⁸ A familiarity with CT equivalents (display of sectional as well as volumetric data) of previously well-established plain radiological signs is needed for accurate radiological interpretation. Interestingly, unlike plain films, on which phleboliths demonstrate a central radiolucency, CT studies show no such corresponding hypodensity consistently.⁹ Phleboliths are composed of calcium phosphate and calcium carbonate at the center of the thrombus, with an extension of mineralization to the periphery with characteristic concentric lamination.⁴ Phleboliths in abdomen are visible on thin-slice CT in 97% of phleboliths with 93% specificity and show a well-defined rounded configuration.¹⁰ Structure of calcification in a phlebolith makes it a perfect sphere on 3D rendered image leading to realistic semblance of shining pearl. A new sign is therefore proposed; using 3D-rendered CT data to describe the typical appearance of the phleboliths. Demonstration of a single or multiple shining pearls is highly suggestive of the possibility of VM or occasional hemangioma, thus rendering this sign highly effective and sensitive in the diagnosis of this lesion. Familiarity with this sign augments the diagnosis of small subtle lesion such as the one documented in Case 3 of our series.

Patients with uncomplicated VM are often managed conservatively. Indications that prompt treatment include disfigurement, hemorrhage, pain, and venous thrombosis.¹¹ Patient with small focal lesions undergoes complete surgical resection or laser photocoagulation. In addition, sclerotherapy using ethanol or ethanolamine oleate is used as first-line treatment of many low-flow anomalies including VMs.¹²

Conclusion

The presence of phleboliths enables a diagnosis of soft tissue venous vascular malformation and hemangioma. Classical description of phleboliths is well documented on plain radiography and conventional sectional CT imaging. Present illustration highlights the role of volume-rendered images in the display the soft tissue phleboliths. A new radiological sign “shining pearls sign” is described with three clinical examples. Awareness of this sign will complement detection process of vascular malformation.

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