Subcutaneous Forehead and Incidental Orbital Varices Diagnosed with MRI Under Sedation

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ABSTRACT

A 5-year-old boy presented for evaluation of an intermittent lesion in the left forehead that, per history from his mother, enlarged when the child slept on his right side. On clinical exam, although there were no notable findings with the child in the upright position, fullness and an enlarging mass when the child was placed on his right side could be reproduced. A diagnosis of a venous varix was suspected. Under sedation, a routine Magnetic Resonance Imaging of the orbits without and with intravenous contrast in the supine position and an axial T1 post-contrast in the right lateral decubitus position was obtained. The forehead lesion demonstrated a positional increase in size and had the Magnetic Resonance Imaging appearance of a varix. An incidental extraconal mass was also noted on Magnetic Resonance Imaging, which had the appearance of a varix and also increased in size in the right lateral decubitus position, confirming the diagnosis of an orbital varix. This case reviews the clinical presentation and imaging findings of two head and neck varices that were diagnosed with Magnetic Resonance Imaging under sedation using positional changes, thus, negating the need for radiation exposure to a child if Computed Tomography was utilized for the radiologic diagnosis.

CASE REPORT

CASE DESCRIPTION

A 5-year-old boy presented with a history of a prominent left forehead soft tissue lesion which expanded when he was placed in the right lateral decubitus position. The patient's parents noticed the lesion at birth and were told it was a mass that was a part of a birthmark. At age 5 years old, he was brought in for an evaluation as his mother was concerned that the lesion enlarged in size when the child slept on his right side (Figure 1).

On physical exam, the child was found to have a normal visual acuity and ophthalmologic examination except for a refractive error. The child wore glasses for correction of myopia and astigmatism. The protruding lesion was noted over the left forehead, especially when the child was placed on his right side. Specifically, the child was placed in the right lateral decubitus position, and within a few minutes, a venous structure became engorged and was not tender to palpation. The mother and the child denied pain or discomfort associated with this lesion. Given the positional nature of the lesion, a diagnosis of probable forehead varix was suspected.

Magnetic resonance imaging (MRI) of orbits without and with contrast was performed under sedation in the supine position. With assistance from the mother, the patient was placed in the right lateral decubitus position to reproduce the observation she had made at home when the child slept. An axial T1 post-contrast sequence was then acquired (Figures 2a, 2b), which showed an enlarging lesion in the left frontal forehead. Additionally, an incidental extraconal orbital mass was also noted on the MRI that also increased in size in between the supine and right lateral decubitus positions (Figures 2c-2f). The T2 signal hyperintensity, homogeneous enhancement, and positional increase in size for both lesions were consistent with varices.

DISCUSSION

We report a case of a young boy with a suspected left frontal soft tissue varix and an incidental orbital varix that were both diagnosed using MRI under sedation. MRI was chosen to evaluate the suspected soft tissue varix as it visibly enlarged on clinical examination between the supine to the right lateral decubitus positions. The imaging appearance and positional increase in size of the lesion were diagnostic of a varix. An incidental, extraconal orbital lesion was diagnosed as varix due to the imaging appearance and enlargement with a change in position. To the best of our knowledge, this is the first report of varices being diagnosed using MRI under sedation with positional changes. The report adhered to the tenets of the Declaration of Helsinki and was HIPAA compliant.

Etiology

Orbital varices account for approximately 1.3% of orbital mass lesions and are characterized by distensible venous walls and low blood flow [1-3]. Primary orbital varices occur from venous wall weakness resulting in dilation and proliferation of the orbital veins [4]. Acquired secondary orbital varices result from an associated dural arteriovenous fistula, a carotid-cavernous fistula, or an intracranial arteriovenous malformation [1].

Patients with primary orbital varices typically present after the onset of symptoms, which usually occur in early adulthood [4]. Symptoms range from benign and reversible to more permanent and emergent presentations. Diplopia, retro-orbital pain, and decreased visual acuity are common findings [1,4]. Positional proptosis and proptosis worsened with increased venous pressure (Valsalva maneuver, coughing, straining, etc.) are also common findings [1,5]. Orbital varices are also known to cause erosion of the orbit leading to osseous defects [6]. Acute presentations of orbital varices include acute thrombosis and spontaneous intraorbital hemorrhage, for which they are the most common cause [1,5-7]. Patients with acute variceal thrombosis may present with decreased visual acuity, proptosis, and retro-orbital pain [3,5]. Larger varices can present as an orbital mass [3].

The imaging modalities used to diagnose varices include computed tomography (CT), MRI, and magnetic resonance venography (MRV) [1]. When using CT, patients are imaged in the supine position, without and with contrast, and without and with the Valsalva maneuver [8]. Due to rapid image acquisition, the Valsalva maneuver can be performed with relative ease and shows enlargement of the varix when it is performed (Fig. 3). On CT, a varix may have a smooth contour with a linear, triangular, or clublike appearance [5]. Other signs of varices on CT include calcified phleboliths related to thrombus formation and erosion of the orbital walls that can cause orbital expansion leading to 'paradoxical enophthalmos' [6]. A disadvantage of CT is that the Valsalva maneuver requires patient compliance and may be difficult for uncooperative or young patients to perform. In addition, CT is associated with radiation exposure, where even low levels of ionizing radiation to the lens can increase the risk of cataract formation, especially in children, and when repeat imaging is required [9].

MRI can also be used to diagnose a varix and does not utilize ionizing radiation. On MRI, varices have been reported to be T1 hypo- to hyperintense, T2 hyperintense related to stagnant blood flow, and usually enhance intensely with contrast administration [5,10]. The varices in our patient had a similar MRI appearance on routine orbital MR imaging. However, the appearance of a varix on MRI may be variable depending on the acuteness or chronicity of associated blood products and the presence of phleboliths [4,5]. Thus, the Valsalva maneuver can aid in the differentiation of varices from other orbital lesions.

Due to the longer acquisition time, a Valsalva maneuver cannot easily be performed with MRI. For our patient, following routine orbital imaging in the supine position, he was placed in the right lateral decubitus position such that the potential forehead varix would enlarge and then a thin-section three-dimensional (3-D) T1 post-contrast sequence was acquired. This allowed us to diagnose both the forehead and incidental orbital lesions as varices, as both lesions enhanced with contrast and increased in size upon imaging in the right lateral decubitus position. The 3-D T1 post-contrast series can then be reconstructed and used to compare with the T1 post-contrast images that were acquired in the supine position. We elected not to use MRV as we didn't feel that it would be helpful in diagnosing the forehead varix. Potentially, an MRV may have provided additional information about the orbital varix, including the native circulation of the orbital venous structures, the varix's extent, and the inflow and outflow of the varix, and may be utilized on follow-up imaging [5,11].

Differential Diagnosis

The differential diagnosis of orbital varices includes other orbital vascular lesions such as venous and endolymphatic malformations, arteriovenous fistulas, arteriovenous malformations, carotid-cavernous fistulas, and aneurysms [1,5,12].

Treatment

The treatment of an orbital varixis primarily conservative. Intervention, including surgery, laser ablation, alcohol sclerotherapy, or embolization, may be required for symptoms such as increased orbital pressure, intractable pain, orbital hemorrhage causing visual impairment, and cosmetic disfigurement [7]. Apart from the visible forehead varix, our patient was asymptomatic, and he will be monitored and imaged periodically.

TEACHING POINT

Intraorbital varices can be diagnosed using CT scanning without and with the Valsalva maneuver; however, CT scanning requires radiation exposure, and the Valsalva maneuver may be difficult for young children to perform. The purpose of this report is to describe the MR imaging appearance and successful diagnosis of forehead and incidental intraorbital varices in a sedated child, who was imaged in the supine and lateral decubitus positions. This report may serve as the basis for further study of MRI in the supine and lateral decubitus positions in children with potential head and neck varices.

AUTHOR'S CONTRIBUTIONS

LSR. BE, and JMD contributed equally to this work. LSB, BE, and JMD were responsible for the study concept and design. Drafting of the manuscript was performed by all authors. All authors contributed to the article and approved the final manuscript.

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QUESTIONS

1. Which of the following is false about orbital varices?

a. Is a high-flow vascular lesion.

b. Is the dilation of one or more veins associated with low blood flow and distensible venous walls.

c. Are rare.

d. Result from a weakness of the ~~post-capillary~~ venous wall.

e. Acquired secondary orbital varices may result from an associated dural arteriovenous fistula.

Answer: a

Explanation: Orbital varices are low-flow vascular lesions.

2. Orbital varices can cause multiple symptoms, except which one of the following?

a. Diplopia

b. Retro-orbital pain

c. Decreased visual acuity

d. Proptosis

e. Dysphagia

Answer: e

Explanation: Orbital varices do not cause dysphagia

3. Multiple imaging modalities may be beneficial in diagnosing an orbital varix except which of the following

a. Computed tomography (CT)

b. CT with the Valsalva maneuver

c. Magnetic Resonance Imaging (MRI)

d. Plain radiographs

e. Magnetic Resonance Venography (MRV)

Answer: c.

Explanation: Plain radiographs are not useful in the diagnosis of orbital varices.

4. Radiologically, orbital varices may have the following appearances, except which of the following?

a. Enhance with contrast administration

b. T1 hypo- to hyperintense and T2 hyperintense

c. Decrease in size with the Valsalva maneuver

d. May have erosion of the orbital walls

e. May show the presence of phleboliths

Answer: c.

Explanation: The Valsalva maneuver will cause an increase in the size of an orbital varix.

Applies to article: Islam N, Mireskandari K, Rose GE. Orbital varices and orbital wall defects. Br J Ophthalmol. 2004;88(8):1092-3. PMID: 15258032.

5. Which of the following are considered to be differential diagnoses of orbital varices?

a. venous malformation

b. venolymphatic malformation

c. aneurysm

d. carotid-cavernous fistula

e. all of the above

Answer: e.

Explanation: All of the above are in the differential diagnosis of an orbital varix.

Applies to article: 1. Pappas A, Araque JM, Sarup V. Orbital Venous Varices: A Rare Bilateral Asymptomatic Presentation. Cureus. 2018;10(9):e3302. PMID: 30705795.

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FIGURES

Figure 1: Photographs of the, a 5-year-old male, in the upright and right lateral decubitus positions show an enlarging left forehead mass when placed on his right side (arrow).

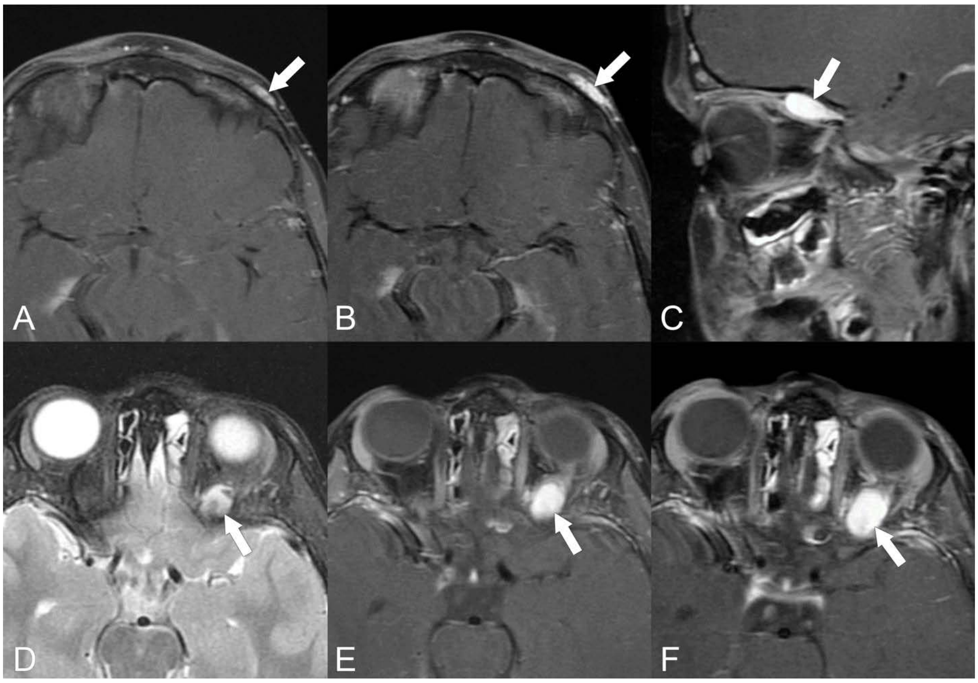

Figure 2: MRI of the 5-year-old male shows dilation of the left forehead lesion and an incidental left extraconal mass between the supine (A, C-E) and right lateral decubitus positions (B and F). (A), Axial T1 post-contrast MRI, supine position shows the enhancing left forehead subcutaneous soft tissue mass (arrow). (B**),** T1 post-contrast MRI, right lateral decubitus position shows homogeneous enhancement and enlargement of the left brow mass consistent with a varix (arrow). (C), Sagittal T1 post-contrast MRI, supine position shows an incidental homogeneously enhancing extraconal mass (arrow). (D), Axial T2 MRI, supine position, shows a hyperintense appearance of the incidental extraconal mass. (E), Axial T1 post-contrast MRI, supine position shows homogeneous enhancement of the mass (arrow). (F), T1 post-contrast MRI, right lateral decubitus position shows enlargement of the homogeneously enhancing extraconal mass consistent with a varix (arrow).

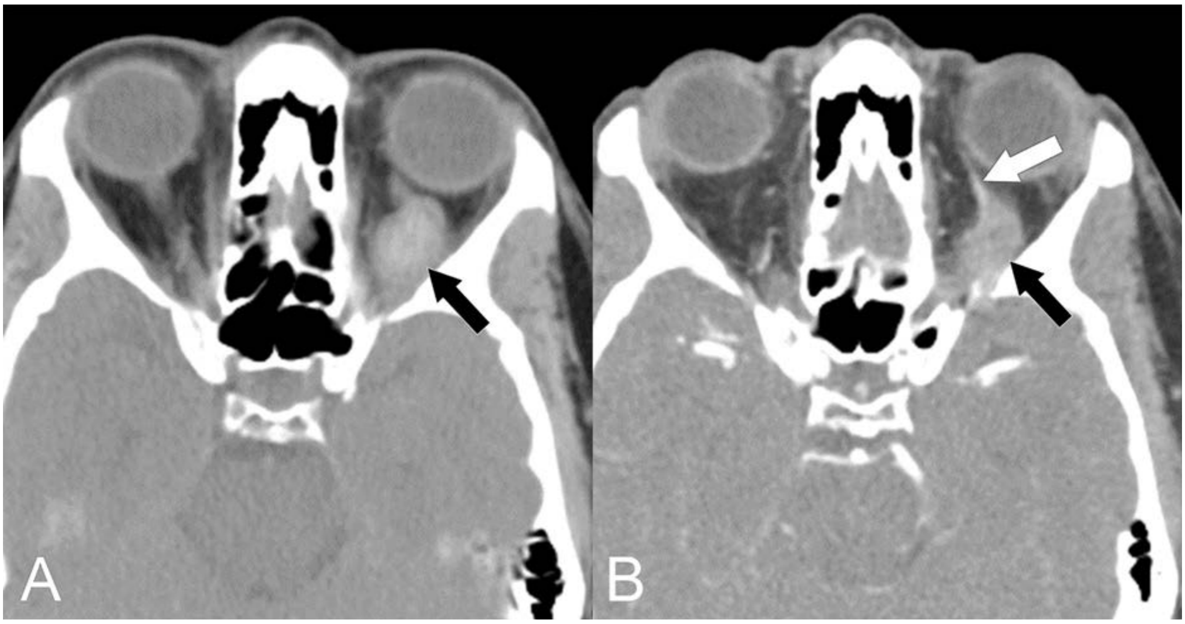

Figure 3: Selected CT images of the orbits demonstrating dilation of a left orbital varix during the Valsalva maneuver in a 37-year-old female patient. **(**A**)**, Axial CT without contrast while performing the Valsalva maneuver shows dilation of the varix (black arrow). **(**B**)**, Axial CT with contrast without the Valsalva maneuver shows a smaller size of the enhancing varix (black arrow) and an associated non-dilated vein (white arrow).

KEYWORDS

Orbit; Varix; Valsalva Maneuver; Computed Tomography; Magnetic Resonance Imaging; Lateral Decubitus Position

ABBREVIATIONS

MRI = Magnetic Resonance Imaging
CT = Computerized Tomography (CT)
MRV= Magnetic Resonance Venography
3-D = Three-Dimensional

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