Orbital Subperiosteal Hematoma in the Newborn Causing Unilateral Proptosis: Ultrasound as First-Line Imaging Modality

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Abstract

Proptosis in the neonatal period is relatively infrequent and has diverse underlying etiologies. One of the more common causes appears to be orbital subperiosteal hematoma. Early detection, differentiation from other causes, and regular follow-up are essential as loss of vision can occur. We describe two cases of neonatal proptosis caused by orbital subperiosteal hematoma highlighting different diagnostic and management approaches, and provide a summary of previously reported cases. Spontaneous resolution occurs in most cases; however, emergent surgical evacuation is warranted in cases of optic nerve compression. This is the first report to provide orbital ultrasound images of uncomplicated neonatal orbital subperiosteal hematoma. Orbital ultrasound followed by magnetic resonance imaging (MRI) is a valid nonradiation approach for assessing neonatal proptosis due to subperiosteal orbital hematoma.

Keywords
► newborn
► orbital
► subperiosteal
► hematoma
► proptosis
► ultrasound

Case Report

Case 1
A male infant was born at term to a 31-year-old, gravid 3 para 2 mother by precipitous spontaneous vaginal birth. Pregnancy and third trimester fetal ultrasound scan were unremarkable. Apgar scores were 7 (1 minute) and 9 (5 minutes). Birth weight was 3,815 g (75th percentile). Facial suffusion was noted immediately after birth and intramuscular vitamin K was administered shortly after birth.

At 16 hours of age, proptosis of the right eye was noted with associated subconjunctival hemorrhage, right-sided hypotropia, and impaired right lateral gaze (►Fig. 1a). The cornea appeared clear bilaterally, pupils responded to light symmetrically, both fundi and the baby’s physical examination were unremarkable. A full blood count and coagulation profile were unremarkable (results not shown).

Imaging and Clinical Course
Orbital ultrasound demonstrated a well-defined hypoechoic, nonvascular 13 × 9 mm mass in the superior aspect of right orbit, exerting mass effect and scalloping the retroorbital fat and superior rectus muscle (►Fig. 1b, calipers, contralateral eye not shown). A cranial magnetic resonance imaging (MRI) study on day 12 of life demonstrated an ellipsoid lesion measuring 19.8 × 22.8 × 11 mm with elevation of the roof of the right orbit. The lesion was hyperintense on T1 and showed mixed intensity on T2 sequences (►Fig. 1c). There
was no obvious breach involving the bony roof of the orbit to suggest the possibility of a meningocele or a meningoencephalocele. The infant was followed up closely and lubricating eye drops were applied to avoid exposure keratopathy. The proptosis resolved clinically over the course of 10 weeks. The infant had normal visual outcome at the last assessment at 12 months of age.

Case 2
A male infant was born at term by normal vaginal delivery to a 23-year-old primigravida mother. Pregnancy, antenatal course including fetal third trimester ultrasound, and family history were unremarkable. Birth weight was 3,600 g (50–75th percentile), length and head circumference were on the 50th percentile, and Apgar scores were 4 (1 minute) and 7 (5 minutes). Facial suffusion was noted at birth and prophylactic intramuscular vitamin K was administered. At about 2 hours after birth, mild swelling of the left upper eyelid was noted; this increased over the following 20 hours with worsening chemosis, proptosis, and subconjunctival hemorrhage (Fig. 2a). Empirical treatment with intravenous flucloxacillin, cefotaxime, and acyclovir was commenced. At 55 hours of life, an afferent pupillary deficit was noted. Full blood count and coagulation profile including individual factor analysis were unremarkable (results not shown).

Imaging and Clinical Course
Emergency computed tomography (CT) imaging demonstrated a nonenhancing ovoid mass measuring 2.2 × 1 × 2 cm in the superior left orbit with mass effect on the left globe and left optic nerve (Fig. 2b). No fracture was observed. On orbital MRI, a uniform mass without enhancement, calcification, or cysts was noted (Fig. 2c). The left optic nerve was displaced inferiorly (Fig. 2d).

Emergency orbital decompression at 65 hours of age via left upper lid sub-brow incision was uncomplicated. Follow-up at 3, 6, and 12 months of age demonstrated normal fundi and visual capacity. Repeat orbital MRI at 4 months showed normal anatomy and normal-caliber optic nerves (images not shown).
Discussion

Proptosis at or shortly after birth appears to be relatively uncommon but may potentially be underreported. Differential diagnoses include congenital tumors (dermoid cyst, teratoma, retinoblastoma, rhabdomyosarcoma, neuroblastoma, neurofibroma, myofibroma), vascular malformations, lymphangiomas, cysts, encephalocele/meningomyelocele, bony abnormalities, abscesses, and thyrotoxicosis. Subperiosteal orbital hemorrhage is a recognized but rarely reported entity. A systematic literature review yielded seven cases with available imaging along with relevant details, which are summarized in Table 1 (see Supplementary Material for search strategy [online only]). The majority of these cases were unilateral; bilateral involvement was described in two cases.

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The mechanism of neonatal subperiosteal orbital hemorrhage is not entirely clear. A history of traumatic delivery has often been quoted. Among reported cases, only one case was associated with forceps and one case with vacuum-assisted delivery. Two cases occurred in the context of shoulder dystocia (Table 1). Extrapolating from adult case series, the presumed pathogenesis could be a sudden rise in intra-abdominal and intrathoracic venous pressure, and transmission of this pressure into the valveless orbital veins during birth. Another contributing factor could be the vacuum created upon disimpaction of head during birth. This has been postulated in a newborn after failed rotational forceps delivery, disimpaction of the head, and subsequent delivery by cesarean section. In this case, marked proptosis was noted; noncontrast CT, however, excluded intraorbital hemorrhage. In our cases, both infants had facial suffusion, which would lend support to a rise in facial venous pressure. The loose attachment of the periosteum to the bone in the superolateral aspect of the orbit in infants may explain the characteristic location in all reported cases, including the two cases presented here. In one previous case, a subperiosteal hematoma with periosteal defect leading to leaking of blood into the orbit was found at the time of surgical exploration.

Coagulopathy does not seem to be common as evident from the normal results of coagulation screens obtained in the reported cases. Administration of vitamin K at birth needs to be ascertained in all cases. In one infant, preexisting coagulopathy and thrombocytopenia were aggravated by anticoagulation for extracorporeal membrane oxygenation (ECMO) for cardiorespiratory failure; this infant developed proptosis secondary to subperiosteal orbital hematoma, which was confirmed by contrast-enhanced MRI and which resolved spontaneously. Proptosis and hyphema were the only initial manifestations in a premature, growth-restricted infant subsequently diagnosed with neonatal alloimmune thrombocytopenia (NAIT) without any other clinical signs suggestive of thrombocytopenia such as petechiae. Details...
Table 1  Cases of neonatal orbital subperiosteal hemorrhage

<table>
<thead>
<tr>
<th>Cases (references)</th>
<th>Age of presentation, eyes involved</th>
<th>Mode of birth, associated factors, and presumed mechanism</th>
<th>Initial imaging, follow-up imaging</th>
<th>Additional investigations</th>
<th>Treatment</th>
<th>Follow-up and outcome</th>
</tr>
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<tbody>
<tr>
<td>1^2</td>
<td>10 h of life (imaging: day 9)</td>
<td>Forceps delivery, forceps mark</td>
<td>Ultrasound, (^a) contrast-enhanced CT (at 9 days of life), repeat CT at 3 weeks and 3 months</td>
<td>Coagulation profile (normal)</td>
<td>Conservative</td>
<td>Regular clinical follow-up to 3 months: normal visual development, Radiological resolution at 3 months</td>
</tr>
<tr>
<td>2^3</td>
<td>Day 4</td>
<td>Spontaneous vaginal birth, periosteal defect with leaking of blood into orbit</td>
<td>Contrast-enhanced CT, serial ultrasound (^a)</td>
<td>Coagulation profile and blood count (normal)</td>
<td>Surgical anterior infra-brow approach</td>
<td>Follow-up with serial USS; resolution at 4 months</td>
</tr>
<tr>
<td>3^4</td>
<td>Birth</td>
<td>&quot;Difficult&quot; vaginal birth, shoulder dystocia</td>
<td>CT</td>
<td>–</td>
<td>surgical transeptal approach via sub-brow incision (day 16 of life)</td>
<td>Normal fundoscopy and vision at 3 months</td>
</tr>
<tr>
<td>4^5</td>
<td>Day 1</td>
<td>Vacuum-assisted vaginal delivery, PPHN, ECMO, preexisting coagulopathy exacerbated by anticoagulation for ECMO</td>
<td>Gadolinium enhanced MRI</td>
<td>Coagulation profile: thrombocytopenia and PT prolongation; tonometry</td>
<td>Conservative</td>
<td>Normal at 3 months</td>
</tr>
<tr>
<td>5^6</td>
<td>At birth</td>
<td>Spontaneous</td>
<td>CT, MRI</td>
<td>Tonometry, CBC, testing for HbS, G6PD, coagulation profile (normal)</td>
<td>Conservative</td>
<td>Complete resolution at 2 weeks and normal visual outcome at 12 months</td>
</tr>
<tr>
<td>6^7</td>
<td>12 h post birth</td>
<td>Spontaneous</td>
<td>CT</td>
<td>CBC, coagulation profile</td>
<td>Needle aspiration</td>
<td>Phthisis, corneal clouding, severe visual impairment</td>
</tr>
<tr>
<td>7^8</td>
<td>Day 1</td>
<td>Traumatic vaginal delivery, shoulder dystocia</td>
<td>MRI</td>
<td>Slit lamp examination, tonometry</td>
<td>Conservative</td>
<td>Clinical follow-up, complete resolution at 2 weeks</td>
</tr>
</tbody>
</table>

Abbreviations: CBC, complete blood count; CT, computed tomography; ECMO, extracorporeal membrane oxygenation; G6PD, glucose-6-phosphate dehydrogenase; HbS, hemoglobin S; MRI, magnetic resonance imaging; PPHN, persistent pulmonary hypertension of the newborn; PT, prothrombin time; USS, ultrasound scan.

\(^a\) Sonographic images not provided.
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