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Major review

Management of neonatal proptosis: A systematic review

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ABSTRACT

Gross proptosis presenting at birth is an uncommon manifestation of a variety of lesions that can compromise vision and result in disfigurement or even loss of life. Notably, many disease entities have different presentations and prognoses in neonates compared to older children. A structured mental framework is essential to an efficient and coordinated response. We present three challenging cases of neonatal proptosis and discuss the clinical presentation and biological behavior of the lesions that are most often implicated.

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1. Introduction

Gross proptosis at birth, a source of profound anxiety for both families and clinicians, is an uncommon, but well documented, presentation for a variety of lesions that can result in vision loss, disfigurement, and even death. Despite its relative rarity, all obstetricians, neonatologists, pediatricians, ophthalmologists, and orbital surgeons must be prepared to evaluate and manage neonatal proptosis expeditiously. An organized and evidence-based approach is paramount.

Although the differential for neonatal proptosis does overlap with that for proptosis in infancy and early childhood, there are also important differences in terms of relative incidence, presentation, treatment considerations, and tumor biology.

With advances in fetal ultrasound and magnetic resonance imaging (MRI), many ocular and orbital conditions can be detected reliably as early as the first trimester.[51] Ideally, this permits a proactive approach in which the obstetrician obtains prenatal consultations with a pediatric ophthalmologist, orbital surgeon, and oncologist. The goal of this multidisciplinary team should be to implement a coordinated plan of care at the time of delivery, thereby maximizing the chances of preserving life, normal anatomy, and vision. Occasionally, however, prenatal visits are missed or significant orbital lesions are not identified on fetal ultrasound, and massive proptosis at delivery comes as a surprise and creates much consternation among medical staff.

We present three challenging cases of gross neonatal proptosis, a review of the literature, and a discussion of the

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clinical presentation and biological behavior of each lesion. A hierarchical approach is suggested as a starting point for evaluation and management by obstetricians, neonatologists, pediatric ophthalmologists, and orbital surgeons. A mnemonic is offered to ensure an orderly sequence of clinical examination and intervention immediately after delivery.

2. Case presentations

2.1. Case 1

A 2-day-old white male neonate was born with massive proptosis of the right eye. An abnormality was first observed on fetal ultrasound at 20 weeks. Maternal hypertension did not require medical intervention. The neonate was delivered vaginally at 38 weeks to a 33-year-old gravida 1, para 0. APGAR scores were 8 and 9, and the birth weight was 3.13 kg. Neonatal heart rate, respiratory rate, and blood pressure were all high normal or slightly elevated.

An orbital MRI obtained on day 1 revealed a right-sided retrobulbar mass, measuring 5.6 × 4.6 × 4.4 cm and containing T2 voids consistent with vascular channels. The irregular post-contrast enhancement pattern was considered "almost pathognomonic for cavernous hemangioma" by the interpreting radiologist (Fig. 1).

On initial evaluation, the patient's right eye was grossly proptotic with injection and foci of subconjunctival hemorrhage (Fig. 2). The ipsilateral eyelids were diffusely stretched and retracted. The cornea was hazy and the anterior chamber shallow with prominent iris vessels. The orbital mass resisted retropulsion and could not be transilluminated. There was an absent direct pupillary response to light with a brisk consensual reaction. The left eye and periocular structures were within normal limits.

There were several firm, bluish subcutaneous nodules on the trunk, extremities, and tongue. B-scan ultrasound disclosed a highly reflective, irregular mass with low to medium sound attenuation, occupying nearly the entire orbit. These characteristics were considered most consistent with teratoma, but a diagnosis of neuroblastoma was also entertained, given the presence of tachycardia and skin nodules. Abdominal ultrasound revealed a 1.4 × 1.1-cm lesion of the right adrenal gland. Biopsy of a superficial lesion was consistent with metastatic neuroblastoma, and this diagnosis was subsequently confirmed with bone marrow aspiration.

The mass was excised on day 3 to minimize corneal exposure–related morbidity and to decrease the risk of necrosis and infection during chemotherapy. Access was achieved via lateral canthotomy and conjunctival peritomy (Fig. 3). The mass was dissected free of the superior, medial, and inferior rectus muscles, but the optic nerve and lateral rectus were grossly infiltrated and had to be sacrificed. The globe itself was successfully preserved.

Microscopic analysis revealed undifferentiated, round basophilic cells in a matrix of vascularized connective tissue. The partially encapsulated mass was 90% necrotic with scattered calcification and 6 mitotic figures per 10 high-powered fields. Immunostaining was positive for neuron-specific enolase (NSE) and S-100, but negative for glial fibrillary acidic protein (GFAP), neurofilaments, leukocyte common antigen, chromagranin, and desmin. Genetic analysis disclosed diploid DNA and intermediate Myc-N proto-oncogene expression.

The patient underwent 4 cycles of cyclophosphamide and VP-16 chemotherapy with apparent resolution of his adrenal mass and skin lesions. During the fifth cycle, however, repeat MRI revealed hydrocephalus secondary to multiple new brain metastases. The infant deteriorated rapidly, despite shunting and aggressive chemotherapy, and died at 8 months of age.

2.2. Case 2

Routine transabdominal ultrasound at 23 weeks disclosed a large cystic mass of the left orbit in the fetus of a 24-year-old gravida 1, para 0. Subsequent transvaginal ultrasound revealed a 3.3 × 3.9 × 3.0-cm lesion (Fig. 4). Follow-up examinations at 26, 30, 34, and 38 weeks' gestation demonstrated interval fetal development without lesion enlargement.

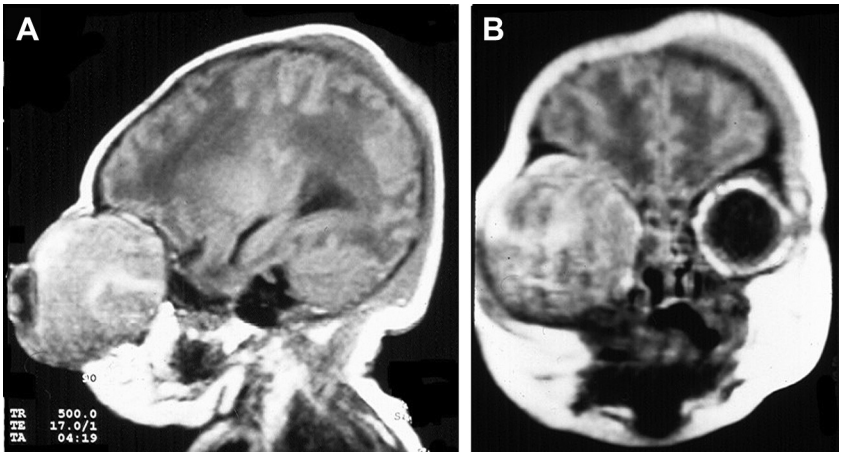

Fig. 1 – Sagittal (A) and coronal (B) T1-weighted magnetic resonance image, demonstrating a right retrobulbar mass causing profound proptosis in a 1-day-old neonate. The mass occupies nearly the entire right orbit and deforms the ipsilateral globe. Note the irregular post-contrast enhancement pattern originally thought to be consistent with cavernous hemangioma.

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