Choroidal metastases as a presenting manifestation of neuroblastoma

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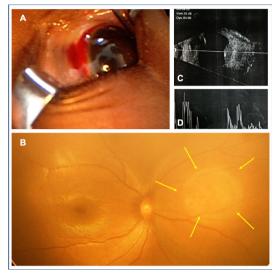


Figure 1 (A) Anterior segment image of right eye of child showing subconjunctival haemorrhage. (B) Subretinal yellowish mass in nasal retina. (C,D). Ultrasound (B) and (A) scan showing subretinal mass choroidal in origin.

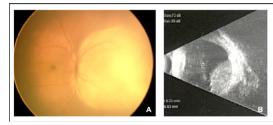


Figure 3 Enlarged choroidal mass at 2 weeks (A) with corresponding ultrasound image (B).

DESCRIPTION

Neuroblastoma is a rare childhood malignancy sometimes presenting with orbital metastasis and proptosis. Here, we present a very rare case of intraocular metastasis of neuroblastoma, with an ophthalmologist being the first physician to diagnose the underlying serious systemic ailment.

A 3-month-old healthy child, born of a term pregnancy, presented to our centre with mother reporting right eye redness for 2 days. Both eyes were apparently normal in size and there were no obvious facial dysmorphisms or pigmentation.

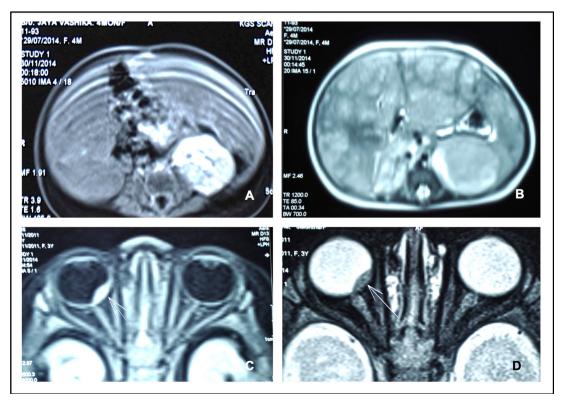


Figure 2 MRI scans of adrenal gland (A), liver (B) and right eye (C, D) showing T1 hyperintense mass originating from left adrenal gland and metastasising to liver and right eye posterior segment.



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Images in...

Anterior segment examination of the right eye revealed subconjunctival haemorrhage (figure 1) located on the nasal side, while the left eye was within normal limits. The child was fixing with both eyes. There was no proptosis or visible posterior extension of the subconjunctival haemorrhage. Both eyes showed orange glow. Dilated fundus evaluation revealed an elevated choroidal mass lesion adjacent to the optic disc in the right eye (figure 1), while the left eye was normal. Ultrasonography of right eye showed a choroidal mass lesion with no calcification (figure 1). Whole-body MRI and MRI orbits revealed neuroblastoma of left adrenal gland with extensive metastasis to the liver and right eye choroid (figure 2). When the child returned with MRI reports, the eye mass had significantly enlarged rapidly (figure 3). The child was diagnosed with Stage 4 high-risk neuroblastoma and was referred for further management to a higher centre.

Neuroblastoma, an undifferentiated malignant tumour of the primitive neuroblasts, is the most common cancer during infancy. The origin of the tumour is primarily from the sympathetic nervous system in the abdomen in 60% cases, and most cases are reported before 4 years of age. At first diagnosis, more than 50% of patients present with distant organ metastases, including lymph node, bones, liver, intracranial and lungs.

Learning points

- Choroidal lesions indicate metastatic disease even in the paediatric population.
- Metastatic neuroblastoma may rarely present as choroidal lesions, and, as such, merit thorough consideration and evaluation.
- Choroidal metastases may progress extremely rapidly and choroidal metastases in a case of neuroblastoma may indicate end stage of disease.

Metastases are mostly believed to occur via haematogenous route. Eye manifestations in neuroblastoma can be in the form of exophthalmos, periorbital ecchymoses, lid swelling, chemosis, disc oedema, strabismus, with the most common location for eye metastases being the choroid.⁴ However, choroidal metastases as a presenting manifestation of neuroblastoma are quite rare. This kind of presentation needs urgent imaging to look for the primary origin and treatment has to be titrated according to the staging of the disease. Treatment of advanced stage high-risk neuroblastoma includes chemotherapy, radiation therapy, bone marrow/haematopoietic stem cell transplantation, radioactive iodine therapy, biological-based therapy and immunotherapy with granulocyte-macrophage colony-stimulating factor and interleukin 2.⁵

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