A mysterious upper eyelid mass in a child

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DESCRIPTION

A 7-year-old girl presented with a history of gradually progressive painless mass over the right upper lid for 2 years. Her birth and developmental history was normal. There was no history of trauma to the face. No similar swellings were noted elsewhere on the body. She had no other systemic ailments. On ophthalmological examination, her visual acuity was 20/20 both eyes. A pedunculated, globular, non-pulsatile upper eyelid mass (Figure 1) with minimal crusting on the surface was noted, measuring around 3 × 3 cm. Few dilated vessels were seen on the surface of the mass. On palpation, the mass was soft, non-fluctuant and non-tender. No bruist or thrill was observed. Surface of the mass was covered by healthy eyelid skin, and base of the mass was pedunculated. There was no change in the size of the mass on Valsalva manoeuvre. Upper lid margin and tarsal conjunctival surface appeared normal. Mechanical ptosis of 3 mm was present on the right side. The anterior segment and fundus examination of both eyes was otherwise unremarkable, with an intraocular pressure of 16 mm Hg. CT of the orbit revealed a homogenous, isodense mass, entirely located in the preseptal area of the right upper eyelid, with well-delineated margins, but no distinguishing features contributing to the probable diagnosis. No orbital or postseptal involvement was noted on CT. The mass was excised completely through a superior eye lid crease incision. Intraoperatively, severe bleeding was encountered, which was not anticipated. Histopathological examination revealed proliferation of capillaries and thick-walled congested blood vessels with extravascular haemorrhagic areas, suggestive of an arteriovenous malformation (AVM). (Figure 2).

AVMs in orbit are high flow conduits between arteries and veins, circumventing the intervening capillary beds, resulting in the characteristic presentation: proptosis, pain, hyperaemia, dilated corkscrew vessels, palpable thrill or bruist, and commonly associated with raised intraocular pressure.1 Orbital AVMs are extremely rare, seen in about 0.14%–0.5% of the population.2 AVMs are congenital hamartomas, which appear as a result of erroneous vascular maturation during 4–6 weeks of embryonic development, but may not necessarily be present at birth. The timing of their appearance depends on the type of vessel involved.3 Precipitating factors for the subsequent enlargement may be trauma, menarche or even pregnancy.4 5 They may grow proportionately with the patient but do not involute thereafter.2 Clinicians as well as pathologists may find it difficult to distinguish between AVMs and capillary hemangiomas. Differentiating AVMs from hemangiomas has diagnostic and prognostic ramifications. Classically, hemangiomas appear within a few weeks after birth. They obtain maximum growth during the first year of life, and typically involute within the first decade,6 whereas AVMs are complex communications between mature arteries and veins. Although AVMs are present at birth, their growth can manifest anytime later in life. The growth of AVMs is progressive and they do not involute unlike capillary hemangiomas. However, orbital AVMs are a rare entity, and

Figure 1 Clinical photograph of the patient showing pedunculated, globular, soft, non-tender, non-pulsatile, upper lid mass in the right eye. Skin over the mass was stretched and shiny with minimal crusting on the surface. Upper lid margin and tarsal surface appeared normal.

Figure 2 10× H&E showing keratinised stratified squamous epithelium (black arrow) and subepithelium showing proliferation of capillaries and extravascular haemorrhagic areas (the yellow arrowhead). 40× H&E (inset) describes congested capillaries (the star symbol), with intravascular haemorrhagic areas (the black arrowhead). The interrupted line shows the thickened vessel wall.
their presentation can be varied. Histopathologically, presence of thick-walled vessel wall is suggestive of AVM (figure 2).

Here, we have described a rare case of a preseptal AVM in a young child, which presented as a pedunculated, globular upper eyelid mass, with none of the diagnostic features of a typical AVM. The vascular nature of the lesion is a risk factor for uncontrolled intraoperative bleeding. It becomes crucial, therefore, to evaluate the patient thoroughly, and to adopt a multidisciplinary approach in the management of the case.

**Learning points**

- Arteriovenous malformations (AVMs) are one of the rare vascular abnormalities, which affect the eyelid.
- Clinical presentation of AVM may be varied, leading to a diagnostic dilemma.
- Severe, uncontrolled intraoperative bleeding should be anticipated during surgical excision of vascular lesions.

**Contributors**

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