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Bane of consanguinity - Vision-threatening orbital sub-periosteal haematoma in hypofibrinogenaemia



Figure. (A) The patient on presentation after trivial trauma to the forehead, with swelling seen on the right eye. (B) Increased intraorbital swelling causing severe proptosis with exposure keratopathy causing decrease in vision. (C) Computed tomography orbit showing hyperdense biconvex lesion in the superior orbit, suggestive of subperiosteal haematoma with the orbital roof intact. (D) After wide-bore needle aspiration, drain was put as there was continuous bleed. (E) Postoperative one-week proptosis completely reduced with resolving corneal infiltrates. (F) At one-month follow up, the patient had an inferior corneal opacity with vision of 6/12.

A nine yr old male child[†], born of second-degree consanguineous marriage, was diagnosed with congenital hypofibrinogenaemia seven years back. He presented with progressive right eye proptosis with exposure keratopathy following trivial trauma to the forehead in September 2018, at the department of Ophthalmology, Ramaiah Medical College & Hospital, Bengaluru, India. Computed tomography of the brain showed hyperacute subgaleal and right orbital subperiosteal haematoma of about 35 × 29 × 17 mm in size. Blood parameters revealed fibrinogen levels <10 mg/dl, prothrombin time >120 sec, activated

partial thromboplastin time >120 sec and thrombin time >120 sec. Cryoprecipitate infusion was given to stabilize blood parameters. When fibrinogen levels were >200 mg/dl, wide bore needle aspiration of subperiosteal haematoma (Video) and lateral tarsorraphy was done to reduce proptosis and protect cornea. The patient improved with vision of 6/12 after one month.

Orbital subperiosteal haematoma is a rare presentation following trivial trauma and is usually self-limiting and rarely requires intervention. In

[†]The child's assent and parents' consent obtained to publish clinical information and images.

bleeding diathesis, orbital subperiosteal haematoma if not aggressively managed may lead to visual loss.

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