Unilateral intra-orbital pseudo tumor in infant- a rare entity with review of literature

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Abstract

"Pseudo tumor" is an idiopathic orbital inflammatory disease characterized by non-specific inflammation involving the orbit. This condition being non-infectious, and non-neoplastic disease with unknown etiology. After thyroid orbitopathy and lymphoproliferative disorder, this is a common condition in paediatric patients. It may pose difficulties in diagnosis. We report a case of 9 months old male child presented with unilateral proptosis, swollen painful lids, with restricted movements for 3 days. In the beginning patient was misdiagnosed as orbital pre-septal cellulitis. We diagnosed this case on ultrasonography and contrast enhanced computerized tomography of brain with orbit. This condition needs to be differentiated from other orbital condition which are common in pediatric age group.

Keywords: Pseudo tumor, Orbit, Infant, Inflammation, Proptosis.

Introduction

Intra orbital pseudo tumor is a non-specific inflammation involving the orbit. It is a hypo cellular lymphoid lesion, often incompletely replacing the orbital fat, lacrimal gland and extraocular muscles in which mature lymphocytes, plasma cells, histiocytes are seen.

Rootman and Nugent classified acute orbital pseudo tumors according to their orbital location, and described five patterns: anterior, diffuse, apical, myositic, and lacrimal.(1) Orbital pseudo tumor is the third most common ophthalmologic disease of the orbit after thyroid orbitopathy and lympho-proliferative disorders and accounts for approximately 8–11% of all orbital tumors. The most frequent subtype of intra orbital pseudo tumor is a local mass within the orbit (50%), followed by dacryoadenitis (29%), myositis (8%), peri-neuritis (4%), eyelid pseudo tumor (4%), and diffuse orbital inflammation (4%).(2) Pediatric cases account for 11.5% of the total population of cases with intra orbital pseudo tumor,(3) and several conditions such as orbital cellulites, pre-septal cellulitis. Retinoblastoma, rhabdomyosarcoma, and leukemia are considered in the differential diagnosis in children.(4)

Case History

A 9 months' male infant presented with painful tender swelling of left eye involving eyelids, with proptosis, external ophthalmoplegia, with fever since 3 days (Fig. 1). Blood investigations revealed hemoglobin- 10.4 gram%, total leukocyte count- 8900 per mm^3. Rest of blood chemistry and coagulation profile was normal. At initial presentation patient was misdiagnosed as orbital pre-cellulitis and put on injectable amoxicillin and clavulanic acid 30mg/kg. Patient didn't respond to systemic antibiotics. Then patient was switched to injectable higher systemic antibiotics with combination of Vancomycin, Amikacin and Ceftriaxone. Even after five days of antibiotic treatment, patient didn't improve clinically. Then patient was further subjected to ophthalmic ultrasonography (USG) orbit (B-mode), contrast enhanced computed tomography (CECT) brain with orbit. USG orbit demonstrated thickening of superior and lateral recti suggesting unilateral pseudo tumor. CECT brain and orbit revealed peripheral enhancing thickened periorbital soft tissue on left side with maximum thickness of 8mm to 9mm. A well-defined non-enhancing iso to hypodense soft tissue density of approximate size 14x12x8 mm was noted, extending into extraconal and intraconal compartment of left eye involving lateral rectus and superior rectus, superior oblique of left eye (Fig. 2, 3). In view of possibility of pseudo tumor, the child was treated with intravenous high-dose steroids (Hydrocortisone 5mg/kg) before considering a biopsy. Patient demonstrated of clinical improvement, including reduced proptosis, ptosis and improved eye movements within 36 hours after starting high-dose steroid treatment (Fig. 4). He continued with intravenous high-dose steroids for 3 days before he was discharged on a gradually tapering dose of oral prednisone.

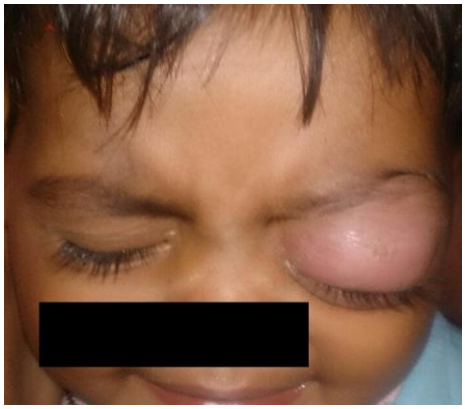

Fig. 1: Clinical image at presentation showing swelling and redness of left eye involving eyelids, with proptosis

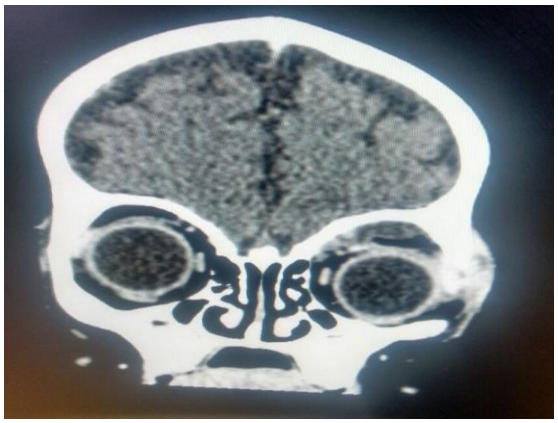

Fig. 2: Coronal section of computed tomography of brain and orbit showing a well-defined non-enhancing hypodense soft tissue density of approximate size 14x12x8 mm, extending into extraconal and intraconal compartment of left eye involving lateral rectus and superior rectus, superior oblique of left eye

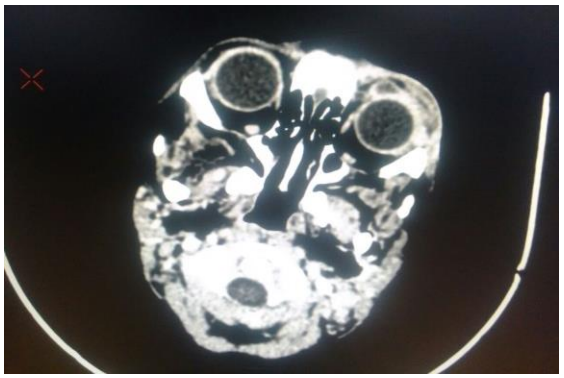

Fig. 3: Axial section of CECT brain and orbit showing a peripheral enhancing thickened periorbital soft tissue on left side with maximum thickness of 8- 9mm

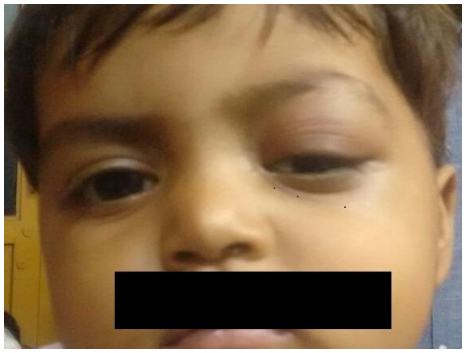

Fig. 4: Post-treatment clinical image showing improvement in swelling and redness of left eye and playful gesture of patient

Discussion

Idiopathic orbital inflammatory syndrome was first described by Birch-Hirschfield in 1905. It is a nonspecific, non-granulomatous inflammatory process of the orbit with spontaneous resolution also known as IOP.[3] Involvement of muscles is a common scenario in IOP. It is bilateral in 45% of pediatric cases, in general 90–95% of the cases are unilateral in adults.[4,5] In 50% of the child patients, systemic signs of IOP may include headache, emesis, anorexia, lethargy, and fever, although these symptoms are rarely reported in adult patients as all these symptoms were present in our patient.[4,6] Ocular motility restriction, swollen eyelid, proptosis, and high orbital pressure are the most common presenting signs in children with intra orbital pseudo tumor.[2] Although these features suggest an active inflammatory process, they are not specific for intra orbital pseudo tumor. A high degree of clinical suspicion combined with neuroimaging results is more helpful in making a correct diagnosis.

Differential diagnosis of paediatric intra orbital pseudo tumor may include orbital cellulites, rhabdomyosarcoma, leukemia, orbital trauma with retained foreign body, ruptured dermoid cyst, lymphangioma, neuroblastoma, metastatic retinoblastoma, and thyroid related orbitopathy.[4] Enlargement of only a single extraocular muscle or a combination of several muscles occur frequently in intra orbital pseudo tumor. The tendons enlarge together with the muscle bundles and lead to a tubular configuration. This muscle involvement is in contrast to thyroid ophthalmopathy, in which all muscles are effected and have a reveal a spindle-shaped configuration with normal tendons.[7] Intra orbital pseudo tumor is differentiated from cellulitis by the absence of infections of contiguous paranasal sinus.[8] Systemic corticosteroid therapy is main stay of treatment. Similar to our case, over 75% of patients show dramatic improvement within 24 to 48 hours of treatment.[9]

In conclusion, intra orbital pseudo tumor is rarely seen in infants, and involvement of extraocular muscle is rare. CECT in case of intra orbital pseudo tumor

provides a better differential diagnosis and prevents unnecessary biopsy. Follow-up after the initiation of steroid treatment, helps in showing response to treatment and confirm suggested diagnosis.

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