Clinicopathologic Case Report

Congenital cyst of the orbit: A case report

ABSTRACT

Dermoid cyst is an example of choristoma (i.e., tumors that originate from aberrant primordial tissue which result in normal appearing tissue in an abnormal location). This particular type of cyst is formed at the site where 2 suture lines of the skull close during embryonic development, and during this time, dermal or epidermal elements are pinched off which later convert to form cysts. Approximately, 50% of these tumors that involve the head are found in or adjacent to the orbit. This article presents a similar case of the orbital dermoid cyst with its management and also a review on other varieties of dermoid cysts of the head and neck region.

Keywords: Dermoid, congenital, cyst, orbit

INTRODUCTION

Dermoid cyst is the commonly used clinical term for benign cystic teratoma, derived from sequestration of surface ectoderm into underlying mesenchyme along embryonic lines of closure. Both dermoids and epidermoids are lined by keratinized stratified squamous epithelium and have a fibrous wall. A dermoid has dermal appendages, whereas an epidermoid has none.[1]

Orbital dermoid cysts may be of three categories [Figure 1]. Orbital dermoid cysts are not attached to the skin, which helps differentiate them from sebaceous cysts. The cyst usually is tethered to the periosteum of the bone near suture lines, including the sinuses or intracranial cavity.[2]

Approximately, 10% of head and neck dermoids are orbital, but they may occur virtually anywhere within or adjacent to the orbit. Twice as many of these lesions develop in the superotemporal orbital quadrant compared with the superonasal quadrant [Figure 2]. They compose 3%–9% of all orbital masses with an average of 4.7% in pooled series.[3]

Shields and Shields had classified orbital dermoid cysts according to their relation to suture lines into juxtasutural, sutural, and soft tissue cysts. Juxtasutural cyst is not firmly attached to suture while a sutural dermoid is firmly attached to and usually associated with bone erosion.[4]

Differential diagnosis includes mucocele, encephalocele, echinococcus cyst, and sebaceous cyst.

Histologic features include a cyst-like structure with a thick fibrous wall, lined with squamous epithelium that may contain hair follicles, glands, and cellular debris. Histologic examination is necessary to differentiate the dermoid cyst with its squamous epithelium lining containing dermal appendages from the epidermoid cyst, which has no dermal appendages. Both lesions are filled with keratin.[5]

CASE REPORT

An 11-year-old boy reported to our department with a complaint of cosmetic deformity because of swelling near the eye for many years. The swelling was pea-sized at birth which slowly increased in size with a faster growth spurt in the last

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1 year [Figure 3]. The swelling has never been associated with any pain or discharge. On examination, the swelling appeared localized and dome shaped measuring approximately 2 cm × 2 cm. The overlying skin was normal in color and texture and was not attached to the underlying lump. On palpation, the swelling was nontender, nonfluctuant, and nonpulsatile. Ophthalmic examination revealed no decrease in visual acuity or strabismus. His paranasal sinus X-ray did not reveal any bony erosion near the orbit [Figure 4]. A lateral brow incision was taken [Figure 5], and the entire tumor mass was excised without rupturing its contents [Figure 6]. The incision was closed in layers that healed well without any complications [Figure 7a and b].

DISCUSSION

The major categories in the classification include cysts of surface epithelium, teratomatous cysts, neural cysts, secondary cysts, inflammatory cysts, and noncystic lesions with cystic component. Cysts of the surface epithelium

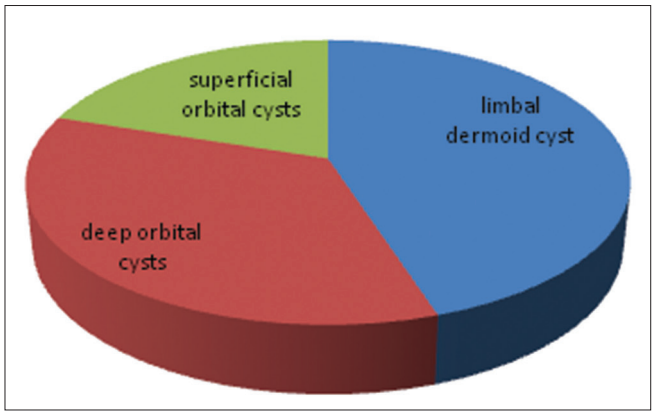

Figure 1: Types of congenital orbital cysts

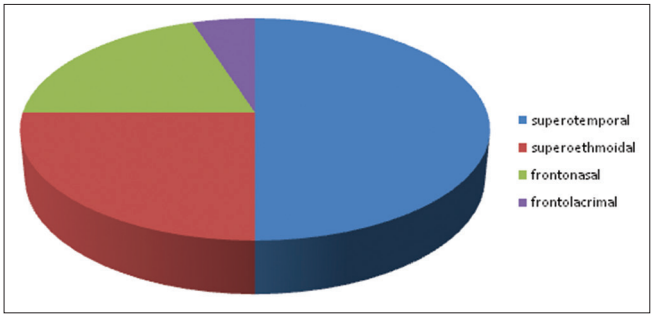

Figure 2: Frequency occurrence of various orbital dermoid cysts

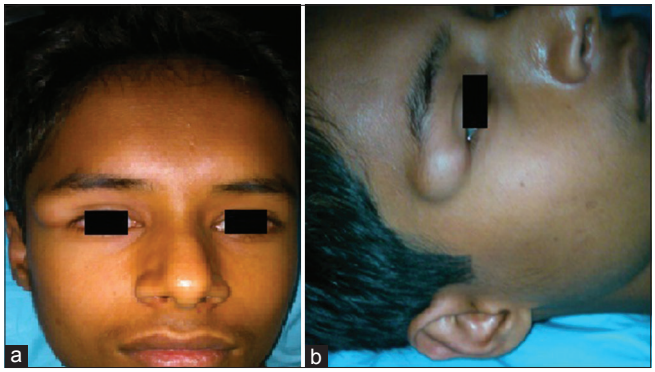

Figure 3: (a and b) Preoperative picture showing the orbital cyst

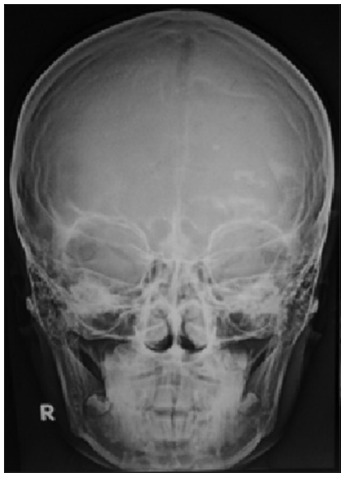

Figure 4: Preoperative paranasal sinus X-ray

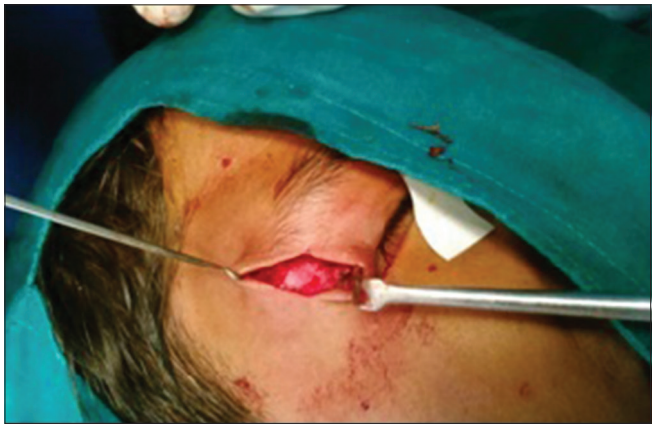

Figure 5: Lateral brow incision taken for excision of lesion

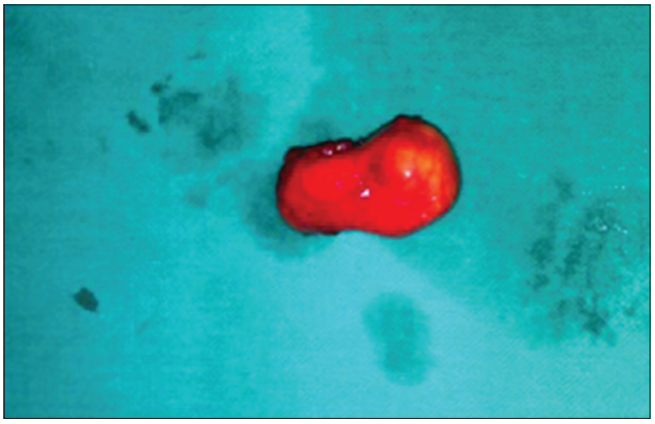

Figure 6: Completely enucleated lesion without rupture

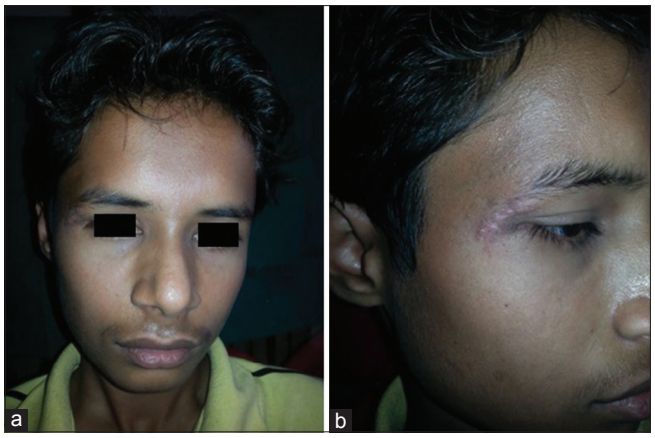

Figure 7: (a and b) Postoperative pictures showing good healing

are further divided into simple epithelial cyst (epidermal, conjunctival, respiratory, and apocrine gland) and dermoid cyst (epidermal and conjunctival). Epidermal dermoid cyst (dermoid) is by far the most common orbital cystic lesion in children, accounting for over 40% of all orbital lesions of childhood and for 89% of all orbital cystic lesions of childhood that come to biopsy or surgical removal. Neural cysts include those associated with ocular maldevelopment (congenital cystic eye and colobomatous cyst) and those associated with brain and meningeal tissue (cephalocele and optic nerve meningocele). The most important secondary cyst is mucocele that can occur in children with cystic fibrosis. Inflammatory cysts are generally due to parasitic infestations and are more common in tropical areas of the world. Noncystic lesions that can have a cystic component include adenoid cystic carcinoma, rhabdomyosarcoma, lymphangioma, and others.

Bilateral limbal dermoids are found in patients with Goldenhar's syndrome.

The various approaches mentioned in literature for managing the orbital cysts depending on their location are as follows: the superficial medial orbital dermoids were excised through medial skin incision or frontoethmoidal (Lynch) incision. The superficial lateral dermoids were removed by subbrow incisions. Deep orbital dermoids were removed with a lateral orbitotomy.[6]

Orbital dermoids or epidermoids should be excised because they enlarge, and the contents leak into adjacent tissues. The material within these cysts is highly irritant and provokes a severe inflammatory reaction, often followed by fibrosis. Every effort should be made to remove the tumor in one piece since the contents of the cystic lesion are irritating and result in lipogranulomatous inflammation of the adjacent orbital tissue.[7]

Deep orbital cysts with intracranial extension will require neurosurgical assistance for removal. In a child patient, if the cyst is very extensive, requiring bone removal for into dissection, then surgery should be delayed until bone growth has ceased.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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