Post Traumatic Epidermoid Inclusion Cyst Of The Orbit

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INTRODUCTION:

Epidermoid cysts arising within the orbit are extremely rare. Approximately 30 cases of epidermoid cysts arising within orbit have been reported. Epidermoid tumors are inclusions of ectodermal elements in sites not normally containing these structures. They can be divided into primary or secondary lesions. Primary or congenital epidermoid cysts are related to implantation of ectoderm at time of closure of the neural groove, or of other epithelial lines, also called as intra-diploic epidermoid cyst. Secondary or acquired epidermoid cysts are usually caused by post traumatic inclusion of surface epithelium. We are reporting a case of post traumatic epidermoid inclusion cyst of orbit clinically presenting with proptosis, which is extremely rare.

CASE REPORT:

A 51 years old male patient presented with gradually progressive protrusion of right eye since 15 years. Patient had history of blunt injury to the right eye 22 years back. Seven years later he first noticed protrusion of right eye. At the time of presentation, patient had difficulty in near vision.

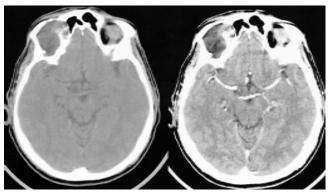


Figure 1a: Plain and contrast CT axial sections of the orbit shows well circumscribed rounded non enhancing cystic lesion in the superotemporal quadrant of right orbit a mean attenuation value of the lesion at 29 Hounsfield units (HU).



Figure 1b: Contrast CT coronal sections of the orbit shows, lesion is displacing the globe anteriorly, medially and inferiorly. Posteriorly the lesion is extending upto the orbital apex and causing destruction of orbital roof, extension of mass into the right frontal sinus is presently seen.

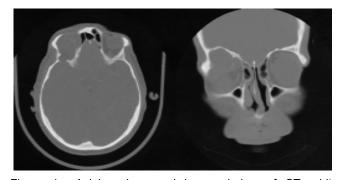


Figure 1c: Axial and coronal bone window of CT orbit shows no e/o sclerosis of adjacent bones.

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On examination, the eyeball was displaced forwards, downwards and medially. The irreducible proptosis measured 9 mm. The visual acuity was 6 / 9 in the right eye. Upward movement and abduction of right eye was limited at extremes. The visual fields and fundi were normal.

CT scan was performed with GE Prospeed Helical CT Scanner which demonstrated a well circumscribed rounded non enhancing cystic lesion in the superotemporal quadrant of right orbit (figure 1a, 1b & 1c). The lesion was displacing the globe anteriorly, medially and inferiorly. Posteriorly, the lesion was extending upto the orbital apex. The orbital roof was destroyed and extension of mass into the right frontal sinus was seen. Viewing console analysis of the scans revealed a mean attenuation value of the mass at 29 Hounsfield units (HU).

A frontal mucocele was suspected and an endoscopic resection of the tumor through the right frontal sinus approach was performed. A cystic pearly mass was seen in the right frontal sinus.

Histopathologic examination showed a ruptured squamous epithelial cyst wall and desquamated layers of keratin towards the center of cyst (figure 2), confirming the diagnosis of an epidermoid cyst.



Figure 2: Histopathological slide shows ruptured squamous epithelial cyst wall and desquamated layers of keratin towards the center of cyst.

DISCUSSION:

An epidermoid cyst was first reported by Pinson in 1807 (1). Epidermoid tumours consist of two main parts: capsule and contents. The capsule is usually opaque, white and glistening. The contents of epidermoid tumors consist of concentric laminae of keratin and epithelial debris which have a white soapy or waxy appearance(2). Epidermoid tumors are inclusions of ectodermal elements in sites not normally containing these structures. They can be

divided into primary or secondary lesions. Primary or congenital or intra - diploic epidermoid cysts are related to implantation of ectoderm at time of closure of the neural groove, or of other epithelial lines. Secondary or acquired epidermoid cysts are usually caused by post traumatic inclusion of surface epithelium(3).

In 1838, Muller first described an intradiploic epidermoid cyst (3). Intradiploic epidermoid cysts arise in the diploe of the bones of orbit and their growth expands both inner and outer tables, thus producing sharply demarcated bone defects on the radiographs (4). The typical radiologic appearance of an intradiploic epidermoid cyst is a widening of diploic space and bone defect with smooth demarcated edges associated with thickened sclerotic margins. Bone window CT scans demonstrate the sclerotic margin of the bone surrounding the epidermoid cysts (3).

Post - traumatic epidermal inclusion cysts are rare and occur mainly in the fingers, palms, and soles (5). Post - traumatic epidermal inclusion cysts are very rare in the skull region. The typical radiologic appearance of an intradiploic epidermoid cyst is a cystic lesion with bone defect without widening of diploic space.

These lesions can be easily differentiated from the frontal mucocele, mixed tumors of the lacrimal gland , sphenoid meningioma and lymphoepithelioma of the nasopharvnx. Retension cvsts or mucoceles of paranasal sinuses, particularly the frontal and ethmoid , may give rise to dehiscence or a hiatus in the orbital wall with marginal sclerosis. Inflammatory changes of the paranasal sinuses, however, are invariably demonstrated with mucocele. Mixed tumors of the lacrimal gland may cause deepening of the lacrimal fossa with or without surrounding sclerosis, but the specific location helps in differential diagnosis. Sphenoid meningioma may simulate epidermoid tumors of the orbit, although hyperostosis with actual proliferation of the sphenoid wings rather than sclerosis facilitate differential diagnosis. Lymphoepithelioma of the nasopharynx may rarely cause hyperostosis of the orbital wall (6).

Post - traumatic epidermal inclusion cysts are very rare in the skull region. One such case was seen by Acarturk and Stofman in the region of deep infratemporal fossa 12 years after the patient sustained blunt trauma to that region (5).

In our case, patient had history of trauma 22 years back & presented with proptosis of the right eye. CT scan demonstrated a well circumscribed rounded non enhancing cystic lesion in the superotemporal quadrant of right orbit causing proptosis. The orbital roof was

destroyed and extension of mass into the right frontal sinus was seen. The radiologic differential diagnosis included frontal mucocele, mixed tumors of the lacrimal gland, sphenoid meningioma and lymphoepithelioma of the nasopharynx. With the given CT findings in our case, we reached to a diagnosis of frontal mucocele. Unfortunately, subsequent histopathological examination was not in congruity with the radiological diagnosis and showed the lesion to be an epidermoid cyst.

In conclusion, the diagnosis of orbital epidermoid cyst should be considered as a differential diagnosis of frontal mucocele.

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