Chronically discharging lacrimal fistula—A forerunner of Malignancy?

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Abstract

Lacrimal sac malignancies are rare cause of epiphora. Squamous cell carcinoma (SCC) is the most common type of lacrimal sac malignancy. Both sexes are equally affected[1]. The peak incidence is at the fifth decade[1]. About 300 cases were reported in literature[2]. Since it is a fatal condition early diagnosis and treatment are important. We present a case of fifty nine years old chronic smoker with history of lacrimal trauma presented as lacrimal sac SCC.

Key word: Discharging fistula, Lacrimal sac malignancy, Squamous cell carcinoma

Case Report

A fifty nine year old diabetic, hyperlipidemic, chronic smoker with history of coronary artery disease presented with swelling near left medial canthus for last six years. The swelling gradually enlarged for last six months and became painful for last three months. He gave history of trauma to the left medial canthus at the age of ten with a pencil, following that he had persistent watery discharge from that area. Examination showed facial asymmetry due to periorbital oedema on left side and minimal proptosis. There was resistance to retropulsion, and positive Nafziger's sign. He had dystopia on left side, all extraocular movements were absent except for minimal abduction. FDT and FGT were positive. There was lid oedema, mechanical ptosis, conjunctival congestion on left side (Fig. 1). Bilateral cornea were clear, pupils reacting to light briskly, and BCVA of 6/9, N_6 both eyes.

Fundus was within normal limits. Supra clavicular lymphnodes were enlarged on left side. CT scan orbit showed left lacrimal sac malignancy with infiltration and extension to surrounding structures (Fig. 2). FNAC of lymphnodes showed metastasis from a poorly differentiated carcinoma possibly squamous cell carcinoma and biopsy of the lesion showed poorly differentiated squamous cell carcinoma (Fig. 3). Staging was $T_4N_1M_0$. Patient improved with six cycles of chemotherapy with Paclitaxel and Carboplatin. Then he lost follow up. After few months he reported with lung metastastasis. Even though two more cycles of palliative chemotherapy were given he expired with systemic metastasis.

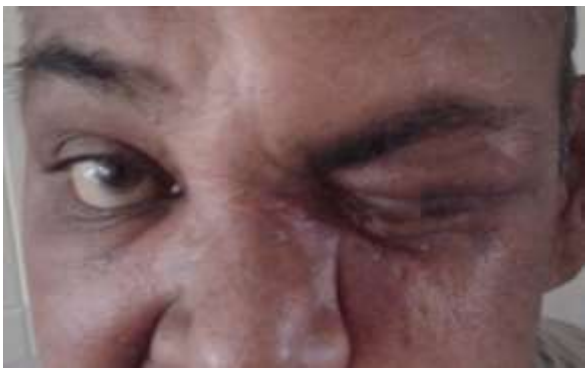

Fig. 1: Gross appearance

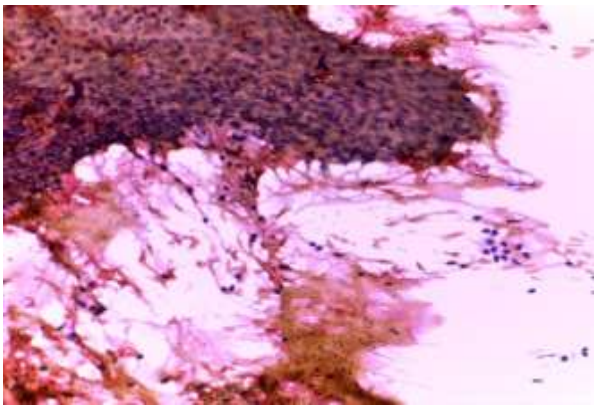

Fig. 2: Histopathology of lacrimal sac malignancy

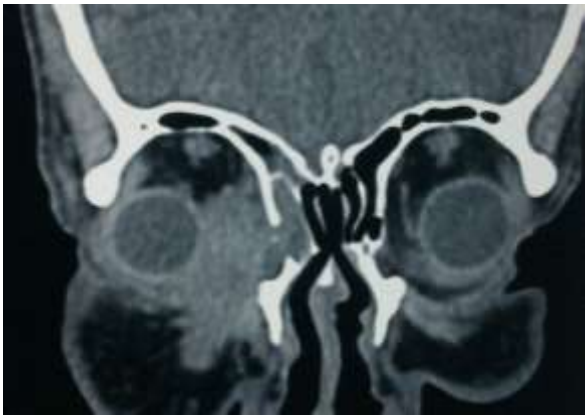

Fig. 3: Axial and coronal CT

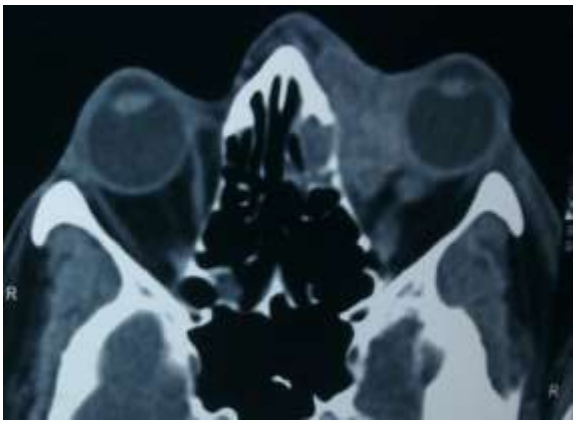

Fig. 4

Discussion

Primary lacrimal sac tumours are rare but most of them are malignant epithelial tumours[3,4,5]. Lacrimal sac malignancies can develop in post trauma dacryocystitis[6]. According to Stefanyszyn MA, etal the most common clinical presentations of lacrimal drainage malignancies are epiphora, recurrent dacryocystitis, lacrimal sac mass, mass emanating through the lacrimal puncta[2,3,4,7,8,9]. Malignant melanoma of lacrimal sac presented with bloody nasal discharge and bleeding from the punctum[3,5,10]. The lacrimal sac malignancies were classified into epithelial and nonepithelial. Squamous and transitional cell papillomas, oncocytomas, and benign mixed tumours are benign epithelial tumours. Squamous cell carcinoma, transitional cell carcinoma, adenocarcinoma, mucoepidermoid, adenoid cystic, and poorly differentiated carcinoma are malignant epithelial tumours. Fibrous histiocytoma, lymphoid lesions, malignant melanoma, hemangiopericytoma, lipoma, granulocytic sarcoma, neurofibroma, and very rare types like small cell carcinoma, solitary fibrous tumours, multiple myeloma,etc. are included under nonepithelial tumours[7,11,12]. Melanoma of conjunctiva can involve lacrimal drainage apparatus[13]. Stefanyszyn MA, etal reported that the lacrimal sac tumours had 55% malignancy rate[7]. Squamous cell carcinoma is the most common type of epithelial malignancy[2,4].

Dacryocystography help to diagnose the lacrimal sac tumours[1,14]. Plain X ray may be normal or shows bone marrow erosion of lacrimal fossa and calcification in lacrimal sac malignancy[1]. In lacrimal sac malignancies CT scan showed soft tissue mass with widening and erosion of nasolacrimal canal and inferior turbinate[1,2,9,10,14]. MRI helps to diagnose subtle bone marrow invasion, perineural and intracranial extension[1,2,10]. Diagnosis of lacrimal sac malignancies are by histopathological examination[2]. Even though lacrimal drainage system malignancies are uncommon cause for lacrimal duct obstruction all DCT specimen should be sent for histopathological examination since these neoplasms are potentially life-threatening and are often difficult to diagnose[4,10,15,16]. Malignant epithelial tumours usually recur locally and can metastasize and be fatal[3,7,10]. The differential diagnosis of the conditions of medial canthal swellings are dacryocystitis, pseudotumour, anterior ethmoid mucocele, sarcoidosis of the sac or extensions of sinonasal malignancies [1]. Necrotizing sialometaplasia is an inflammatory reaction against an ischemic insult or local trauma within a glandular tissue, most commonly seen in the minor salivary glands of the oral mucosa[17]. It can rarely affect lacrimal sac mucosa (necrotizing dacryocystometaplasia)[17]. It is considered clinically and histopathologically as a differential diagnosis of well-differentiated squamous cell carcinoma[17].

Lacrimal sac malignancies can have lymphnode and systemic metastasis[10]. Lacrimal sac SCC spread by direct invasion to orbit, paranasal sinuses, and skull[1]. Lymphnode metastasis is late, involve preauricular, submandibular, jugulodigastric and cervical nodes[1]. Effective method of managing lacrimal sac malignancy is a combined sinus orbit approach with simultaneous reconstruction of the bony defect with titanium mesh[18]. Epithelial malignancies of the lacrimal drainage system grow along the epithelium, therefore should be treated by wide surgical excision of the tumor and the entire lacrimal drainage system combined with a lateral rhinostomy and radiation therapy[7]. Since some of non-epithelial lacrimal sac malignancies are life threatening, early diagnosis and treatment are important[3]. If tumor is confined to orbit at the time of diagnosis complete excision with pre and postoperative radiotherapy improves the survival[1,10]. For tumors with orbital extension exenteration can be tried[19]. Lacrimal sac malignancy has a recurrence rate of 50%[2,14].

Bilateral simultaneous lacrimal sac malignancies are associated with HIV, HSV infection in young individuals[2]. Bilateral SCC of lacrimal sac can develop denovo or seeding of tumour from other side, or as metastasis or as local recurrence, due to loss of antiangiogenic factors following excision of primary

tumour[2]. Lacrimal sac fistula may be either congenital or acquired. Pseudo and true lacrimal fistula can develop following trauma to lacrimal region or following dacryocystitis or rupture of lacrimal abscess and can be treated by conventional DCR or endoscopic DCR[20,21,22,23]. Literature search showed increased risk of squamous cell carcinoma in recurrent, chronic anal fistula[24].

Conclusion

Even though benign lesions are common causes for epiphora, malignancies should be suspected especially in blood stained epiphora. Since lacrimal sac malignancies are life threatening ophthalmologist should be aware of this rare condition since early diagnosis and treatment are lifesaving. So all DCT specimen should be sent for histopathology examination. Since it has high recurrence rate follow up is mandatory.

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