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Case Report

Solitary fibrous tumor of the lacrimal sac mimicking lacrimal sac mucocele: A case report with review of literature

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ABSTRACT

Solitary fibrous tumors (SFT) are benign neoplasms, composed of spindle-shaped cells. It has been reported in numerous locations in the body, but SFT of lacrimal sac is very rare. A 67 year old man clinically diagnosed as lacrimal sac mucocoele. During DCR, there was a yellowish mass with vascularisation on the surface. The mass was removed in-toto. Histopathology and immune-histochemisty confirmed SFT of lacrimal sac. Patient was followed up for 6 months without any recurrence.

1. Introduction

Solitary Fibrous Tumor (SFT) was described as a primary spindle cell tumor of the pleura.[1] It is diagnosed in different parts of the body including viscera of the abdomen, pelvis and trunk. Six percentages of all SFTs arise in the head and neck region.[2] SFT of the lacrimal sac is rare, very few cases of SFT have been reported worldwide. Because of its varied clinical presentations and histopathologic appearances, it can be misdiagnosed. We present a rare case of SFT of the lacrimal sac which presented like lacrimal sac mucocoele with review of literature.

2. Case Presentation

A 67 year old man, presented with history of watering and discharge in the right eye for 2 years, associated with painless progressive swelling in the medial canthus. He has a history of ruptured abscess treated from elsewhere 2 months back. His best corrected visual acuity (BCVA) was 6/12 in both eyes. External examination showed a firm, well-circumscribed, non-tender mass of size 13*10 mm in right lacrimal sac region, not extending above medial canthal tendon (Figure 1). There was no regurgitation of mucopurulent discharge or pus on applying pressure over lacrimal sac area. Anterior segment examination revealed immature cataracts in both the eyes while posterior segment examination was unremarkable. Obstruction in the lacrimal passage was confirmed by probing and syringing. He was clinically diagnosed as chronic dacryocystitis with lacrimal sac mucocele and was posted for dacryocystorhinostomy (DCR). During DCR, after skin incision and blunt dissection, there was a yellowish mass with vascularisation on the surface. The mass was removed in-toto and specimen sent for histopatological examination, after explaining to the patient and the bystanders. Haemostasis was achieved and wound closed in layers.

Histopathological examination revealed structures of lacrimal sac tissue with subepithelial circumscribed tumor tissue formed by plump to spindle cells arranged in perivascular pattern with extensive stromal fibrosis. There were numerous stag horn vessels with epitheloid cell proliferation. Interspersed mitosis, pseudovascular spaces lined by epitheloid cells and giant cells seen (Figure 2A,B). Immunohistochemisty showed diffuse and strong positivity

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for CD34, Bcl2, vimentin and CD99 (Figure 3 A,B,C, and D), suggestive of solitary fibrous tumor, intermediate grade.

Patient was followed up for 6 months without recurrence. The study adhered to the ethical principles outlined in the Declaration of Helsinki. A written informed consent was taken from the patient about the use of clinical data and clinical photographs for the purpose of the study.

3. Discussion

Solitary fibrous tumors are composed of spindle-shaped cells and were originally considered a neoplasia of the mesothelium. It was first described in 1931 by Klemperer and Rabin as a distinct mesothelial tumor arising from the pleura and mediastinum.[3] Many cases have been described other sites like subcutaneous tissues, paranasal sinuses, nasal cavity, meninges, extremities, thoracic wall, and abdomen (liver, adrenal, peritoneum, urogenital system).[4] Earlier, some such tumours may probably have been described as hemangiopericytoma. SFT of the orbit is rare and typically present as benign orbital masses in middle-aged adults.

Lacrimal sac SFTs may present as nasolacrimal duct obstruction, recurrent dacryocystitis, or as a slow-growing mass in the medial canthal region. Lacrimal sac SFT is extremely rare and only around 15 cases were reported in literature in English language (Table 1). Most of these cases were presented as mass in the medial canthal area and epiphora as the presenting complaint. 4 cases had a previous history of DCR.[5–8] They may easily be misdiagnosed due to their rarity, heterogeneity of clinical presentations and histological appearances.[9] Definitive diagnosis can be made by histopathological and immunohistochemical examination only, due to the variability in radiological appearances.[10]

Microscopically, SFTs consists of ovoid to spindle cells that are haphazardly arranged with varying cellular densities in admixture with stromal collagen bundles.[11] SFTs can be a mimicker of other tumours such as mesotheliomas and sarcomas .Hence immunochemical staining has proven useful in establishing the diagnosis.[12] The tumour stains positively for CD-34, CD-99, BCL-2 antigens and vimentin and negatively for S-100 protein, desmin, cytokeratin and actin.[13] Less than 15% of solitary fibrous tumours are aggressive. Malignancy is evidenced by hypercellularity, cytologic atypia, necrosis, infiltrative margins, and high mitoses.[14] Such features were not present in this case.

Solitary fibrous tumours are chemo-resistant, total excision is the preferred treatment. In cases of partial resection, local recurrence or malignant transformation may occur.[5] As there is a high chance of recurrence, complete surgical excision and continued follow-up should be emphasized.[10]

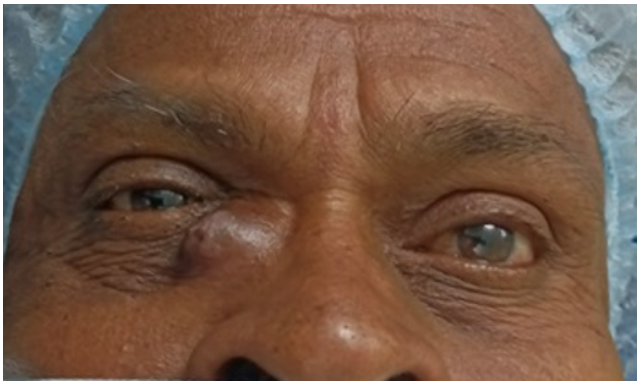

Fig. 1: Clinical picture showing mass in the medial canthal area

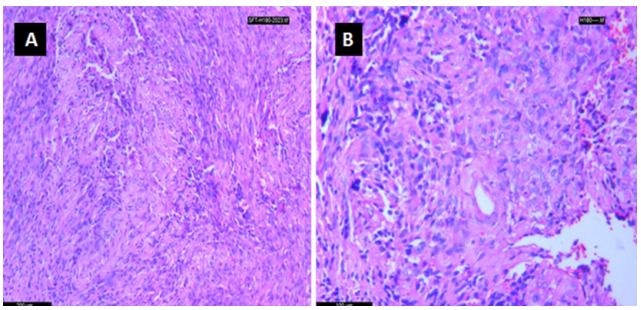

Fig. 2: Histopathological examination of lacrimal gland tumor section (H and E section,revealed structures of lacrimal sac tissue with subepithelial circumscribed tumor tissue forme by plump to spindle cells arranged in perivascular pattern (arrow marks) with extensive stromal fibrosis.

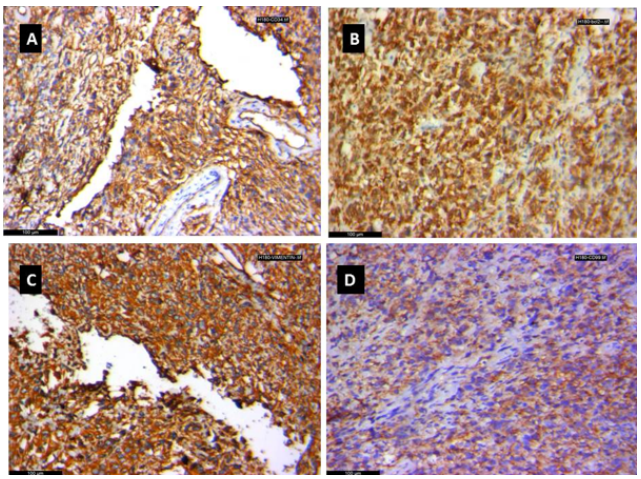

Fig. 3: Immunohistochemistry of lacrimal sac tumor showing strongly positive for **A:** CD-34 cells, **B:** Bcl2, **C:** Valentin and **D:** CD99, suggestive of solitary fibrous tumor

Table 1: Profile of previously reported cases of solitary fibrous tumor of the lacrimal sac

Case	Author	Age/sex	Clinical presentation	CT scan findings	Histopathology	Immuno histochemistry	Follow up period
1	Woo, Kyung[5] (1999)	23/M	medial canthal mass	Heterogeneously enhancing mass extending to NLD	Densely cellular areas,proliferation of spindle cells with less cellular sclerotic areas,anastomosing staghorn vessels	NA	2 years
2		34/F	medial canthal mass, h/o DCR 6 years back	Homogenously enhancing mass extending to NLD	proliferation of spindle cells,anastomosing staghorn vessels	CD34+,Vimentin+ S100 –,Desmin -	7 years
3	Rumelt, S[15] (2003)	67/F	RE-epiphora 10 years	NA	The bland spindle cells were arranged in a 'patternless' pattern without a distinctive whorl or cord pattern.These mesenchymal-like cells were uniform.no pleopmorphism	CD-34 +, CD-99+ BCL-2 +, Vimentin - S-100 –,Desmin –	1 year
4	Kim HJ[16] (2008)	26/M	Periocular mass	Well defined, heterogenous, extending to NLD	NA	NA	NA
5		51/M	Periocular mass	Well defined isodense, extending to NLD	NA	NA	NA
6	Kurdi M[17] (2014)	44/F	Watering, medial canthal area mass	Discrete lesion in the lacrimal sac fossa consistent with a mucocele of the lacrimal sac.	Moderately cellular, spindle cell neoplasm with prominent vascularity. The cells, admixed with thick collagen bands, were arranged in fascicle.	CD34+, Vimentin+ S100 –, Desmin –,SMA-	1 month
7	Moriyama[18] (2017)	71/M	Medial canthal mass	Homogenously enhancing mass in the anterior part of right orbit, extending to nasal cavity,	Spindle shaped cells, in pattern less arrangement within a collagenous matrix ,dilated vascular spaces	CD 34+, Bcl2+, CD99+,vimentin+, S100-	2 years
8	Caroline[9] (2018)	63/F	Watering, medial canthal area mass	Mass in the right lacrimal sac with extension into the proximal right nasolacrimal duct.	A cellular spindle cell lesion featuring fascicles and storiform architecture with occasional foci of staghorn vessels, perivascular hyalinization and keloidal collagen bundles.	NA	16 months

Continued on next page

Table 1 continued

9	Gudkar (2019)	65/F	Watering, medial canthal area mass	Well-circumscribed, homogenous, right medial canthal mass extending into the nasolacrimal duct	Partially capsulated tumor with hyper and hypocellular areas and scattered thin-walled blood vessels. Spindle-shaped and epithelioid cells were distributed haphazardly	CD34 +, Bcl-2+Cytokeratin-,S-100 –	6 months
10	Maria Araújo[6] (2019)	35/M	Medial canthal area mass	Round lesion with intense and homogeneous contrast enhancement, with mass effect at the inferomedial wall of the orbit	Highly vascularized lesion with cancer cells sometimes ovoid, sometimes fusiform, inside a collagenous stroma with slight pleomorphism and marked capillary vascularization	CD 34+, Bcl2+, CD117−, S100−, SMA−.	3 years
11	Morawala A[7] (2020)	35/M	Watering, medial canthal area mass	Moderately well-defined, isodense, relatively homogenous mass arising from the lacrimal fossa and extending into the bony NLD	Fascicles of spindle cells arranged in a pattern-less pattern with occasional whorls. The cells were plump with moderate eosinophilic cyto-plasm, oval, and vesicular nucleus	CD34+ , Bcl-2 +, CD99 +, Cytokeratin-.	8 months
12		66/F	Watering, medial canthal area mass	a large lesion extended into the orbit up to the junction of mid and posterior orbits with globe	Fascicles of spindle cells arranged in a pattern-less pattern with occasional whorls. The cells were plump with moderate eosinophilic cyto-plasm, oval, and vesicular nucleus	CD34+, Bcl-2 + CD99 +,Cytokeratin-.	6months
13		39/M	Medial canthal area mass	Isodense lesion extending from the bony lacrimal fossa into orbit with globe compression	Fascicles of spindle cells arranged in a pattern-less pattern with occasional whorls. The cells were plump with moderate eosinophilic cyto-plasm, oval, and vesicular nucleus	NA	7months
14	Kumar P[(10)] (2021)	34/F	Watering, medial canthal area mass	Well-circumscribed enhancing lesion seen that is centered at the lacrimal fossa	Oval to spindle cells with high cellularity, and these cells were arranged in staghorn and fascicular pattern. These cells are arranged around the compressed vessels.	CD-34 +, CD-99+ Vimentin +, S-100 –	6months
15	Agrawal S[8] (2021)	47/F	Watering, medial canthal area mass	Homogeneous well-defined lesion in the lacrimal sac area,which was extending into the nasolacrimal duct withadjacent bone remodelling	A spindle-cell tumor with variablecellularity and areas of collagenization and normal lacrimal sac lining of stratifiedcolumnar epithelium with gobletcells overlying the tumor. Prominentvascular pattern. There was nocytological atypia or mitosis seen.	CD-34+ S-100 - cytokeratin -	6months

4. Conclusion

The lacrimal sac SFT can present as nasolacrimal duct obstruction, recurrent dacryocystitis, or slow-growing mass in the sac area. Even though lacrimal sac SFT is very rare, it should be considered as a differential diagnosis of medial canthal region mass in patients with epiphora.

5. Conflict of Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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None.

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