Smooth muscle hamartoma in lateral canthus simulating lipodermoid: A rare entity.

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Abstract

Smooth muscle hamartoma (SMH) is a rarely encountered neoplasm with only 2 reported cases of SMH involving conjunctival fornix. The similarity in the location, appearance and radioimaging features of the lesion with lipodermoid in our case point towards the difficulty in diagnosing SMH clinically.

A 16 year old male presented with a 12 mm × 5 mm pinkish yellow, non-tender lesion in temporal fornix with extension to superior and inferior fornices. No hair follicle was observed on its surface. There was no variation in the size of the lesion with valsalva/bending down/straining efforts. Local excision of the lesion was planned and sent for histopathological examination.

CECT brain and orbit revealed a well-defined mass in lateral part of sclera in left eye with fatty attenuation, with no post septal extension/erosions of adjacent bones/calcification/intracranial extension suggestive of lipodermoid. Histopathology report of excised lateral canthal mass revealed lobulated fat with vascularised fibrous tissue and smooth muscle suggestive of a hamartoma.

Keywords: Smooth muscle hamartoma, Lipodermoid, Cystic lesion of orbit.

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Introduction

Smooth muscle hamartoma in conjunctival fornix is a rare entity, with only 2 case reports till date in literature. In our case, temporal location with the characteristic pinkish yellow appearance of the lesion in a young male with radioimaging revealing a fat attenuated lesion point towards a differential diagnosis of lipodermoid, a choristoma usually prevalent at this age group.

Excision biopsy is gold standard for accurate diagnosis of smooth muscle hamartoma. None of the cases reported till date had post-septal extension, thus pointing towards a benign nature of the lesion. Hence, SMH should be considered as a differential for cystic conjunctival forniceal lesions.

Case Presentation

A 16 year old male noticed a swelling in the lateral canthus of left eye since one and half years, gradually increasing in size. There was no history of trauma or chronic drug intake. Patient was systemically stable.

Ocular examination revealed a visual acuity of 20/20 with projection of rays accurate in all quadrants in both eyes. There was no limitation of movement of extraocular muscles and anterior and posterior segment were within normal limits. Local examination revealed a 12 mm × 5 mm pinkish yellow, non-tender lesion in temporal fornix with extension to superior and inferior fornices (Figure 1). No hair follicle was observed on its surface. There was no variation in the size of the lesion with valsalva/bending down/straining efforts. Local excision of the lesion was planned and sent for histopathological examination.

Figure 1. *Clinical photograph of a 12 mm x 5 mm pinkish yellow, non-tender lesion in temporal fornix of left eye with extension to superior and inferior fornices.*

Investigations

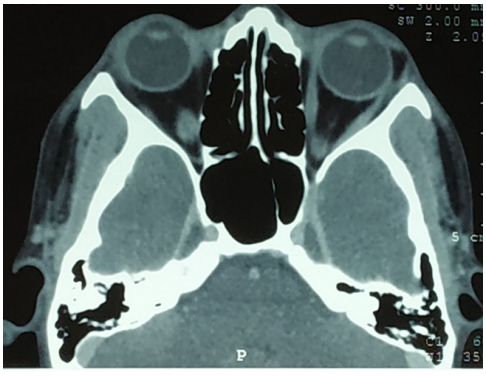

Figure 2. *CECT brain and orbit revealing a well-defined mass in lateral part of sclera in left eye with fatty attenuation, with no post septal extension/erosions of adjacent bones/calcification/ intracranial extension.*

CECT brain and orbit revealed a well-defined mass in lateral part of sclera in left eye with fatty attenuation, with no post septal extension/erosions of adjacent bones/calcification/ intracranial extension suggestive of lipodermoid (Figure 2).

Histopathology report of excised lateral canthal mass revealed lobulated fat with vascularised fibrous tissue and smooth muscle suggestive of a hamartoma (Figure 3).

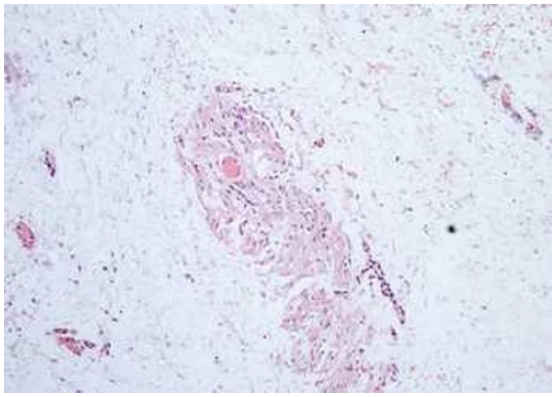

Figure 3. *Histopathology of excised lateral canthal mass revealed lobulated fat with vascularised fibrous tissue and smooth muscle suggestive of a hamartoma.*

Differential diagnosis

Our case was suggestive of a choristoma (lipodermoid) based on clinical and radiological examination.

Results and Discussion

Smooth muscle hamartoma (SMH) – first described by Sourreil et al. is a rarely encountered cutaneous neoplasm characterised by proliferation of smooth muscle bundles with the epidermis [1]. Common locations of SMH are the upper extremities, face and mammary region [2].

Other rare locations reported are scrotum, eyelid and eyebrow. Only two cases have been reported in literature with SMH involving conjunctival fornix [3-6]. Histologic sections in the studies revealed large bundles of smooth muscle with a fibrotic background and interdigitating fat, consistent with the histological appearance in our case. A male predominance has been noted [1].

The possible explanation for the origin of hamartoma in the conjunctival fornix is either from the smooth muscle of vascular endothelium or from capsulopalpebral muscle of lower lid and/or levator palpebral superioris of upper lid.

The temporal location with the characteristic pinkish yellow appearance of the lesion in a young male with radio imaging revealing a fat attenuated lesion point towards a differential diagnosis of lipodermoid, a choristoma usually prevalent at this age group. Smooth muscle hamartoma, being a rare entity with no definite characteristic features on radio imaging remains unrecognized and clinically mistaken for other cystic lesions in the orbit.

The stage III optic pathway glioma in our patient occluded the third ventricle and foramen of Monro by its compressive effect leading to hydrocephalus. Dysfunction of pituitary and hypothalamus due to direct extension of the tumor led to endocrinological abnormalities and concurrently developed diencephalic syndrome.

Conclusion

Surgical excision of the tumor was not feasible due to its intracranial extension. Radiotherapy was avoided in view of increased radiation induced sequalae at 10 months of age. Chemotherapy was initiated to halt the progression of the disease and preserve the vision in the left eye. Thus, smooth muscle hamartoma should be considered in the differential diagnosis of cystic appearing conjunctival fornix lesions.

Learning Points

- Smooth muscle hamartoma in conjunctival fornix is a rare entity, with only 2 case reports till date in literature.
- SMH can have similar clinical and radiological characteristics as lipodermoid.
- SMH should be considered as a differential for cystic conjunctival forniceal lesions.
- Excision biopsy is gold standard for accurate diagnosis of SMH.
- None of the cases reported till date had post-septal extension, thus pointing towards a benign nature of the lesion.

Patient Consent

Consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of the patient.

Conflict of Interest

The author declared that there is no conflict of interest.

Financial Interest

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