Primary lacrimal gland teratoma in an adult: A clinicopathologic and radiological correlation

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A 20-year-old male presented with a gradually growing mass in the left lateral brow region for 2 years. On imaging, the mass appeared to arise from the lacrimal gland, was cystic, without any bony abnormality. With a diagnosis of dermoid cyst, an excision biopsy was performed. The histopathology revealed an epithelium-lined cyst, with the wall of the cyst comprising dermal adnexa and inflammatory cells. The wall also contained fragments of skeletal muscle, nerve bundles, adipose tissue, exocrine and secretory acini, few blood vessels with occasional hair shafts, with the lacrimal gland in the periphery of the tumor. There were no immature elements, atypia, and malignant cells, thus confirming the diagnosis of a mature cystic teratoma. Primary lacrimal gland teratoma is a rare diagnosis and its presentation in adults is extremely uncommon.

Key words: Lacrimal gland, orbit, teratoma

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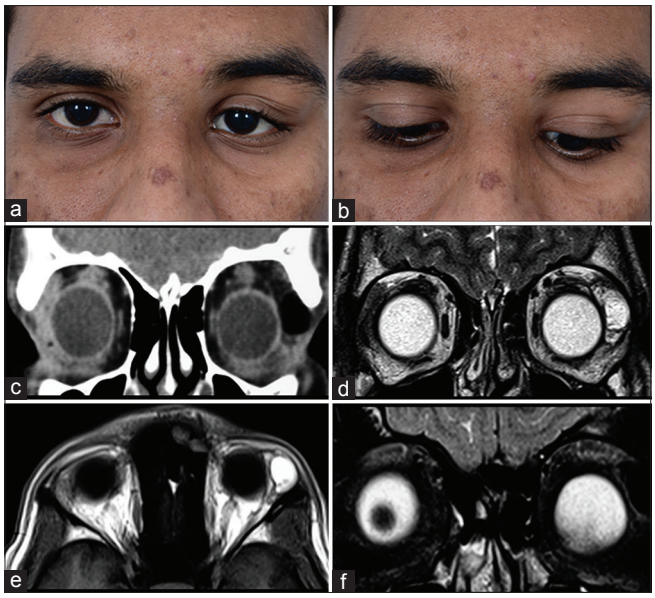

Figure 1: Primary lacrimal gland tumor in an adult, clinical and radiological imaging. (a and b) External photograph of a 20-year-old male with a mass in the left lacrimal gland. CT orbit coronal view (c) reveal a well-defined hypodense mass with no bony abnormality. MRI orbit reveals a hyperintense signal on both T1 (d) and T2 sequences (e and f)

Teratomas are embryonic tumors that contain tissue elements derived from all the three embryonic germ layers—the ectoderm, mesoderm, and endoderm.[1] The most frequent location of a teratoma is in the gonads, followed by sacrococcygeal, retroperitoneal, and mediastinal regions.[2] In contrast, orbital teratoma is rare, but a well-known entity.[1-5] Orbital teratoma almost always presents as a unilateral mass, and the stimulus for differentiation of the pleuripotential germ cells toward adult structures in the orbit is not clear.[1] The most characteristic presentation of this tumor in the orbit is extreme proptosis present at birth.[1-6] Herein, we report an unusual presentation of a primary lacrimal gland teratoma in a 20-year-old patient.

Case Report

A 20-year-old male presented with a gradually growing mass in the left lateral brow region for two years and reported rapid growth in the past one month. There was no other significant ocular, medical, or family history.

On examination, the best corrected visual acuity in both eyes was 20/20. A distinctly palpable orbital mass was present in the superotemporal part of the left orbit just below the superior orbital rim [Fig. 1a and b]. It measured approximately 20 × 15mm, and was firm, nontender, and mobile in all directions. There was no proptosis on Hertel's exophthalmometry. Ocular motility was full and normal in all directions. Palpebral part of the lacrimal gland appeared normal and Schirmer's test did not reveal any tear function abnormality. On computed tomography (CT), a well-defined oval mass in the superotemporal orbit with the lacrimal gland not seen separately was evident [Fig. 1c]. It had a density similar to that of the adjacent fat with no calcifications or surrounding bony abnormality. On magnetic resonance imaging (MRI), the lesion appeared T1 and T2 hyperintense [Fig. 1d and e] with suppression of signal on fat-saturated sequences [Fig. 1f]. A clinical and radiological diagnosis of an external angular orbital dermoid cyst was made.

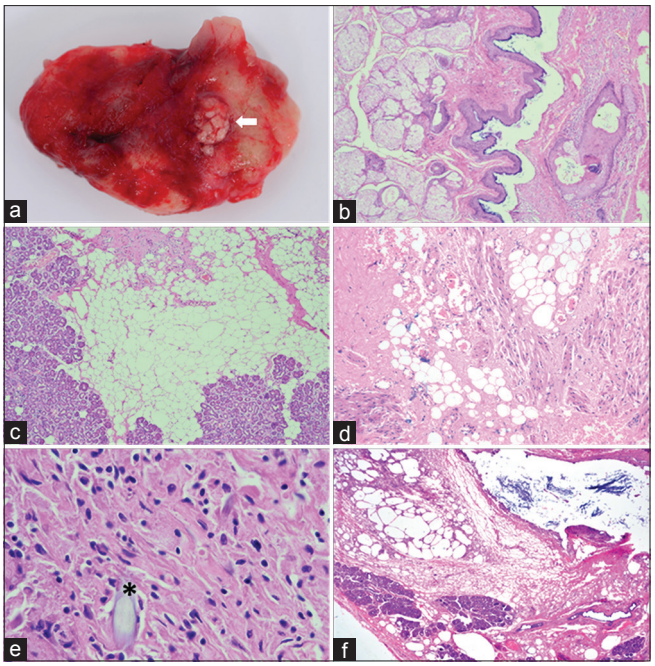

Figure 2: Primary lacrimal gland teratoma in an adult, gross and histopathological features: (a) Gross photograph of the excised tumor reveals a lobulated cystic mass with lacrimal gland in the periphery (white arrow). Histopathology of the tumor shows (b) stratified squamous keratinized lining with dermal adnexae [HE, ×20], (c) lacrimal gland acini with adipose tissue [HE, ×20], (d) tissues of mesodermal origin: skeletal muscle and adipose tissue [HE, ×20], and (e) hair shaft appearing as a refractile structure (black asterix) inciting a foreign-body reaction [HE, ×400], (f) Lacrimal gland (left bottom corner) showing tissues from all three germlines: adipose tissue (mesoderm), stratified squamous epithelium with lamellated keratin (ectodermal, top right), and ducts (endoderm, just above the lacrimal gland on the right bottom) [HE, ×20]

A lateral orbitotomy was performed to completely excise the tumor. On gross examination, the tumor was a single cystic mass with a part of the lacrimal gland in the periphery [Fig. 2a], and pultaceous material filling the cystic cavity. On histopathology, the cyst was lined by stratified squamous keratinized epithelium, filled with lamellar keratin [Fig. 2b]. The wall was infiltrated by histiocytes, lymphocytes, plasma cells, and multinucleate foreign-body giant cells. Skeletal muscle fragments, nerve bundles, adipose tissue, exocrine and secretory acini, few blood vessels, and occasional hair shafts with a surrounding foreign-body reaction were also seen [Fig. 2c-f]. There was no evidence of immature elements, atypia, or malignancy. A diagnosis of a mature cystic teratoma with a foreign-body reaction was made. At 41 months follow up, the patient is doing well with no recurrence.

Discussion

Teratomas of the head and neck represent 2% of all teratoma cases, and orbital presentation is uncommon.[6] Although orbital teratoma can be malignant, most frequently the tumor is benign in nature.[1-6] Several classification systems have been

proposed for orbital teratomas.[1] While Duke-Elder classified orbital teratomas based on the degree of differentiation of the three germ layers, Kivela *et al*. have proposed a classification depending on the location of the tumor within the orbit.[1] Histopathological classification includes mature or benign, immature but probably benign, immature and possibly malignant or cancerous, and frankly malignant orbital teratomas.[2]

Orbital teratomas usually present in a neonate as unilateral proptosis.[1-3] Primary orbital teratoma in an adult has been reported earlier, but is extremely uncommon.[3-5] Although Levin *et al*. described a primary orbital teratoma in a 15-year-old-female, the teratoma had been present for at least 13 years with gradual progressive growth.[4] In a yet another report, a 28-year-old-female with acute onset of proptosis was discovered to have a primary orbital teratoma with a tooth.[3] Whitham too described a teratoma with tooth in a 21-year-old male that arose from the lacrimal gland.[5] Our adult patient presented with an apparent lacrimal gland tumor, and on histopathology, a mature teratoma was confirmed with a part of the lacrimal gland in the periphery. On imaging and surgery, the lacrimal gland was not seen separately from the tumor; thus, the benign teratoma seemed to arise from the lacrimal gland.

Conclusion

In conclusion, primary teratomas are unusual tumors of the orbit, and lacrimal gland teratoma is extremely rare. Adult presentation of an orbital teratoma can be misdiagnosed as an orbital dermoid cyst, and is confirmed only on histopathology.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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