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Case Report

Eyelid metastasis as presenting feature of disseminated leiomyosarcoma – A rare case report

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Abstract

Soft tissue sarcomas with smooth muscle differentiation are termed leiomyosarcoma. Leiomyosarcoma is an aggressive tumor commonly originating from smooth muscle cells of uterus or retro peritoneal areas. Distant metastasis occurs by hematogenous route to liver and lung. Eyelid metastasis without involvement of other ocular structures is extremely rare. A case of eyelid metastasis which on extensive investigations was proved to be from upper gastrointestinal tract leiomyosarcoma is reported here.

Keywords: Leiomyosarcoma, Eyelid metastasis

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Introduction

Sarcomas are rare malignant tumors arising from mesenchymal cell lines accounting for just 0.7% of all malignancies.[1] Soft tissue sarcomas with smooth muscle differentiation are termed leiomyosarcoma. In a review of 26,758 cases of soft-tissue sarcoma by Toro et al., leiomyosarcoma was most common type of soft tissue sarcoma (24%) followed by malignant fibrous histiocytoma (17%), liposarcoma (11.5%) and dermatofibrosarcoma (10.5%).[1] Common sites of origin include smooth muscle cells of soft tissue, uterus or retro peritoneal areas.[1]

Leiomyosarcoma is an aggressive tumor which can have distant metastasis which occurs generally by hematogenous route to liver and lung.[2] Cutaneous leiomyosarcoma rarely metastasizes, whereas retroperitoneal and large blood vessel leiomyosarcoma display high metastatic rate.[2] Metastasis from leiomyosarcoma to orbit has been reported. However, eyelid metastasis without involvement of other ocular structures is extremely rare.

We report a case of eyelid tumor which on extensive investigation was proved to be a metastasis from disseminated esophageal leiomyosarcoma. At the time of presentation with lid tumor, patient was not a known case of systemic leiomyosarcoma. Eyelid tumor as presenting feature of disseminated leiomyosarcoma has not been reported previously.

Case report

A 60-year-old male patient presented at our tertiary care hospital with complains of raised nodular swellings in right lower lid since last four months [Fig. 1]. The patient had noticed two small nodular growths in right lower lid which had increased gradually. It was not associated with any history of pain, ocular discharge or bleeding.

Ocular examination showed best corrected visual acuity (BCVA) for distance 6/6 in both eyes. BCVA for near N6 in both eyes.

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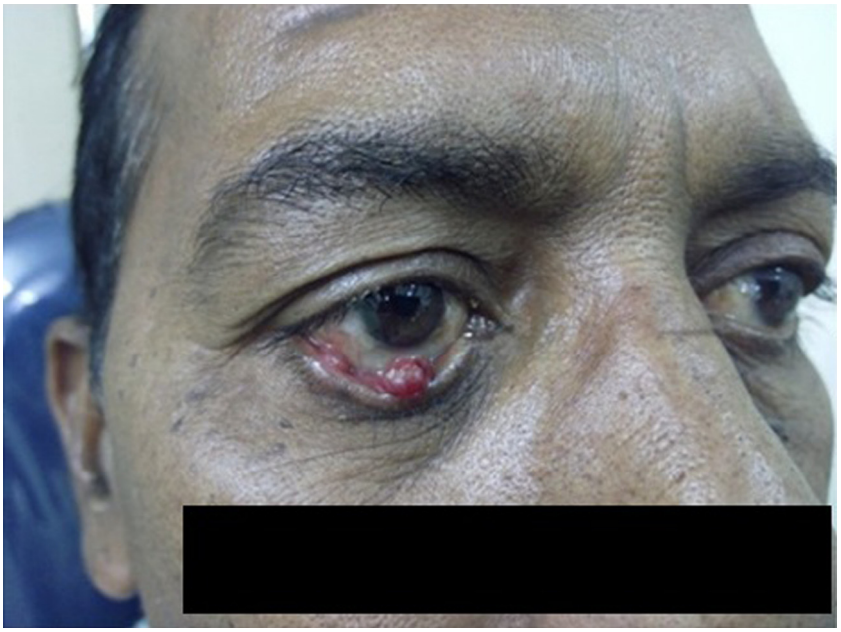

Figure 1. Raised nodular swellings in right lower lid.

Right eye showed two nodular reddish colored swellings in lower lid extending from conjunctival side toward lid margin. Larger swelling was round 12 mm diameter, raised, with keratinized surface. Smaller swelling was oval shape, raised and 8 mm in diameter. Larger swelling located in central part of the lid while smaller swelling was located near outer canthus. Growths were fixed and not moveable over underlying tarsal plate. There was no loss of eye lashes or destruction of lid margin. It was not associated with bleeding, discharge or local lymphadenopathy. Both swellings caused mild mechanical ectropion of lower lid. Rest of anterior segment and fundus was normal. Left eye showed unremarkable anterior and posterior segments.

A differential diagnosis of sebaceous gland carcinoma was considered at this stage. Patient started on topical antibiotics and was taken up for incision biopsy of mass under local anesthesia after routine blood and urine investigations which were normal.

Histopathology of biopsy specimen showed tissue bit lined by stratified squamous epithelium with focal ulceration. Sub epithelial tissue comprised of numerous dilated capillaries along with sheets of atypical cells which were round to oval in shape with moderate amount of eosinophilic to clear cytoplasm. Some hyperchromatic spindle cells were also seen. Brisk mitotic activity noted. Immunohistochemistry (IHC) showed specimen positive for Vimentin and Smooth Muscle Actin (SMA) but negative for cytokeratin (CK), CD 34, and HMB 45. This showed mass had smooth muscle component while ruling out epithelial tumors and melanomas [Fig. 2].

In view of atypical histopathological and IHC features, smooth muscle tumor was suspected. Also possibility of metastasis from distant primary was suggested by pathologist due to atypical histopathological features.

The patient was reassessed and thorough systemic examination revealed hepatomegaly. Whole body computed tomography (CT) scan showed disseminated metastatic deposits in liver, lung and retroperitonium [Fig. 3]. Magnetic resonance imaging (MRI) brain showed features suggestive of metastasis to right occipital lobe, subarachnoid and subdural spaces.

Patient was evaluated further by oncologist. Positron emission tomography (PET) CT scan showed hypermetabolic subcutaneous lesion in right lower lid, scalp, multiple lung parenchymal nodule, large necrotic liver mass, and multiple skeletal metastasis [Fig. 4]. Bone marrow biopsy showed no metastatic deposits or abnormal cell cluster. Colonoscopy showed normal study. Endoscopy of upper gastro intestinal tract revealed multiple polypoidal lesions in esophagus, stomach and duodenum. Biopsy was taken from esophagus which on histopathology and IHC was consistent with high grade leiomyosarcoma.

While under evaluation to determine primary site of malignancy, patient developed sudden onset left sided hemiplegia with subarachnoid hemorrhage due to bleed in intracranial metastasis. Patient was managed conservatively and underwent palliative brain radiotherapy (20 Gy over 5 days). Underwent shave excision of lid tumors as they increased in size. Patient was placed under the care of oncologist and started on palliative systemic chemotherapy.

Discussion

Leiomyosarcoma is a notably rare and highly aggressive tumor. Overall reported survival for patients diagnosed with soft tissue leiomyosarcoma range from 50% 3-year survival to 64% 5-year survival, making this tumor one of the more aggressive soft tissue sarcomas.[3] Occurrence of leiomyosarcoma in the ophthalmic region is extremely rare. Primary orbital leiomyosarcoma has been reported in the literature as sporadic cases.[4,5] Primary orbital leiomyosarcoma usually affect young individuals and clinically they present as painless proptosis progressing slowly over several months.[4] Vascular or sympathetic smooth muscle cells is believed to be precursor of primary ophthalmic leiomyosarcoma.

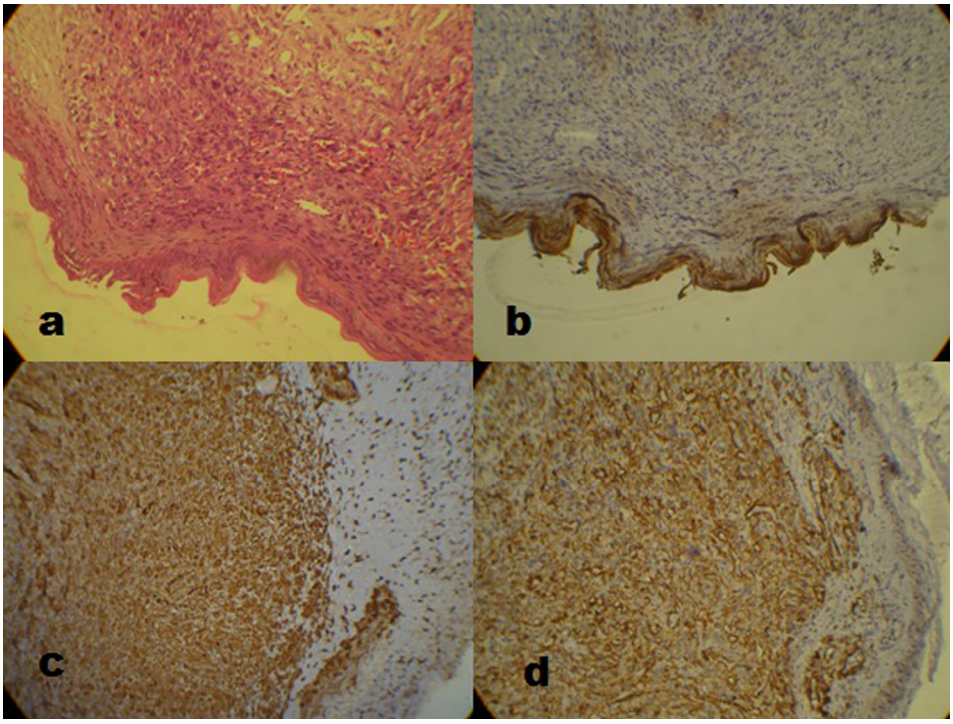

Figure 2. H &E stained section magnification 100× showing (a) presence of sheets of malignant cells in the sub epithelial region. The tumor cells are CK negative (b), vimentin (c) positive and SMA (d) positive.

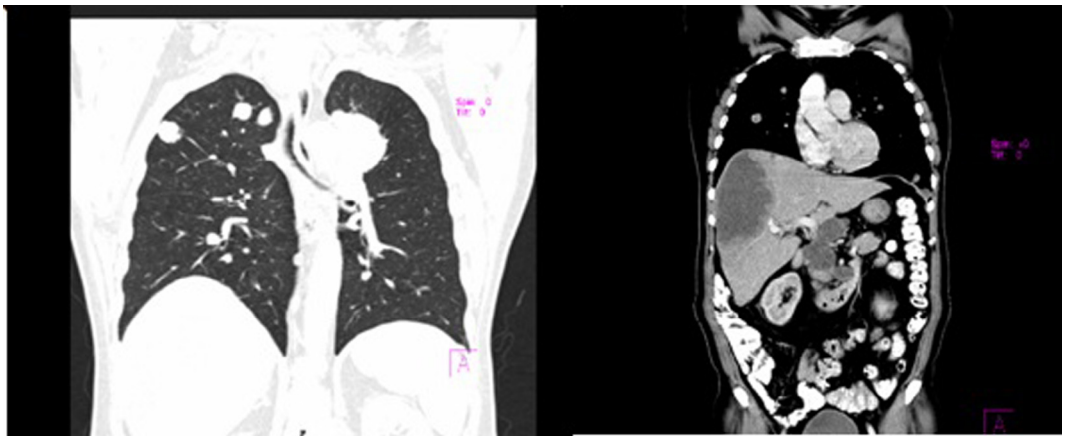

Figure 3. Whole body CT scan showed disseminated metastatic deposits in liver, lung and retroperitonium.

More common occurrence is orbital metastasis in a known case of leiomyosarcoma.[6,7] Due to its aggressive nature and tendency to have distant metastasis by haematogenous route, orbital metastasis can occur in a known case leiomyosarcoma. Such isolated cases of orbital metastasis from retroperitoneal, pulmonary or uterine leiomyosarcoma have been reported.[6,7] Such metastatic deposits can affect intraocular structures involving choroids.[6]

Metastatic lesions to eyelid are rare and occur less frequently than orbital or choroidal metastasis.[6–8] Most metastatic lesions to the eyelid are carcinomas. Metastatic deposits from disseminated leiomyosarcoma involving lids are extremely rare and extensive medline search by authors could revel only two such cases ever reported.[9,10] In both cases, patients were known cases of primary leiomyosarcoma and under treatment. In one case patient had esophageal leiomyosarcoma and esophageal tumor was reclassified after histopathological evaluation of the eyelid specimen. In other case patient had a subcutaneous leiomyosarcoma of the neck which had been excised before with subsequent radiotherapy.

In our case, patient was not a known case of systemic malignancy and had sought consultation primarily for the eyelid growth. Patient was diagnosed as a case of leiomyosarcoma with primary tumor involving upper gastro intestinal tract after atypical histopathological features of eyelid growth which prompted extensive systemic evaluation. Such presentation of eyelid metastasis as initial manifestation of disseminated leiomyosarcoma has not been reported previously.

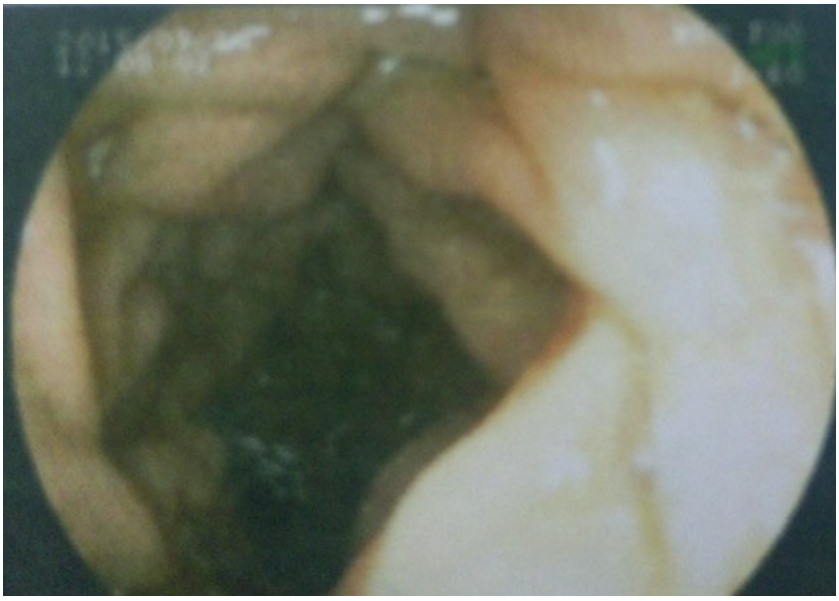

Figure 4. Endoscopic view of upper gastrointestinal tract showing multiple polypoidal lesions.

Conflict of interest

The authors declared that there is no conflict of interest.

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