Hemangioendothelioma of Orbit

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Abstract :

Hemangioendothelioma of the orbit is a rare condition which usually occurs in young adult with mean age group of 24 years. Very few cases had been reported in the literature. We describe here a case of hemangioendothelioma in a female of 30 years who came with history of penetrating foreign body entering into right eye and fluctuating painful swelling in the lower lid with decrease in the vision. The patient's presentation simulated malignancy of maxillary antrum, however, the diagnosis was confirmed by histopathology.

Key Words : Hemangioendothelioma, Orbit, Angiosarcoma, Maxillary antrum.

CASE REPORT

MEN, a 30 years old female presented to OPD of Dept. of Ophthalmology of our institute with history of penetrating foreign body in right eye 21 days back, which was removed and treated. Five days later she developed progressive, fluctuating, painful, swelling of the right eye. Swelling first appeared in the lower lid and progressed to involve upper lid with decrease in the size of eye. There was no other significant history of past illness, hypertension, diabetes, diplopia, polyopia, watering from eye and ha-loes. She was not on any other medication. Her sleep, bowels and appetite were normal.

On systemic examination, there was no abnormal finding and no sign of any systemic illness. Local examination revealed no abnormality in the left eye. Right eye was minimally proptosed and displaced supero-nasally, palpal fissure was markedly reduced with only 2 mm of cornea was visible. Lid was oedematous and conjunctiva revealed mild congestion and chemosis. Lower conjunctival

fornix was obliterated. Cornea was clear and anterior chamber was normal. Pupil was 3 mm in size and showed normal reaction to light. Intraocular pressure was 17.3 mm of Hg and

Fig 1 : Photograph Showing reduced palpal aperture and deviation of eye along with swelling.

Fig II : CT Scan Section showing mass in right maxillary antrum, with involvement of infratemporal fossa, pterygoid fossa and erosion of anterolateral and medial wall of maxillary antrum.

vision was 6/60, with pin hole 6/6. Inferolateral movement of right eye was restricted. Locally a nontender, horizontally mobile, ill-defined cystic swelling with poorly defined margins was present. Temperature of swelling was normal with absence of any from of pulsations. Skin over swelling was normal and mobile. Orbital margins were intact, fundus examination revealed generalised venous engorgement along with transverse choroidal fold in macular region indication pressure on posterior pole.

Hemogram revealed normal picture with normocytic hypochromic RBCs in peripheral smear. X-ray PNS (Water's view) and orbit showed no abnormality, USG demonstrated a 2.8x 2.3x 3.2 CMS hyperechoic soft tissue mass inferior to right eye ball, destruction on orbital

Fig III : Coronal CT scan showing mass in the orbit with extension into infratemporal fossa, right maxillary antrum, right nasal fossa and ethmoids.

floor and extension of mass into Rt. maxillary antrum. Patients refused CT scan.

Provisional diagnosis of carcinoma maxillary antrum with erosion of orbital floor and extension to orbit was made and patient was planned for incisional biopsy. On opening through infra-orbital incision, brownish black mass was seen between periosteum and bony floor of orbit. On probing, surprisingly there was no erosion of floor and bony wall. Mass was excised. On 4th post-operative day, ocular movement improved and fundus showed decreased in choroidal folds. However, foveal reflex remained dull and vision reached to 6/36 with pinhole. On the day of discharge vision was 6/18 with pinhole and minimal choroidal folds. Surgical wound healed without any complications.

Histopathological section showed tumour tissue composed of vascular channels of regular size and shape, endothelium lining showed pleomorphism, multilayering and formation of papillary fronds. The cells are of moderate size with large hyperchromatic nuclei and scanty cytoplasm. Intervening connective tissue showed hyalinisation and haemorrhages.

After 15 days patient reported again to Dept. of ENT for follow up with massive swelling around orbit and maxilla, vision decreased to finger counting at 1 meter with restriction of the eye movements. There was form mass around orbit and temporal region. Diplopia was absent. CT Scan done at that time showed a moderately enhancing mass of 5.1x 4.6x 3.7 CMS causing saddling and destruction of orbital bony floor. Eye ball was pushed anterosuperiorly with invasion of inferior rectus muscle. Mass was extending to maxillary antrum with erosion of lateral, medial, anterior wall and floor. Right ethmoid was eroded and mass extended to Rt. nasal cavity. Laterally it was extending to right temporal fossa and posteriorly to right pterygoid fossa without erosion of the base of skull. Since patient was not willing for surgery, she was sent to higher centre for radiotherapy, following which she did not turn up for follow up.

Discussion

Hemangioendothelioma is a malignant tumour of which cells manifest many of the functional and morphological properties. It is infiltrative, locally invasive and have high metastatic potential with metastatic rate of 60 percent (Shield J (1984), Jacobiec FA, Jones IS (1986). It is a rare monomorphic vascular tumour and constitute 3% of all orbital tumours (Enzinger & Weiss 1983) and is 1% of all sarcomas (Jacobiec & Jones 1986) Because of the origin of tumour from endothelial cells of blood vessels it's also called as Malignant Hemangioendothelioma. It develops by proliferation of endothelial cells of blood vessels (Sheild J 1984). Tumour can also develop post radio-

therapy, or can progress from paranasal sinuses (Jacobiec and Jones 1986, Sheild 1984). A case has been reported where it metastasised from carcinoma breast (Jacobiec and Jones 1986). In our case possible hypothesis could be that there was proliferation of endothelial cells following foreign body reaction and local treatment with steroids. Endothelial cell origin of hemangioendothelioma can be identified using immunohistochemical stain for cytoplasmic factor VIII and Ulex europeus agglutinin (UEA I) or by electron microscopy (Hufnagle et al 1983). Messmer et al (1983) due to inapparent endothelial cell origin in few cases preferred to use term Angiosarcoma rather than hemangioendothelioma and also reported an epitheloid variant of the angiosarcoma. They also reviewed previously reported 7 cases. Histopathologically it showed striated mass composed of cords of oval and round pleomorphic cells separated by dense fibrous tissue. With reticulin stain, endothelial cells are seen as proliferating in lumen forming fashion, with reticulin outlining the lumen (Jacobiec and Jones 1979). Clinically there is no sex predilection and is tumour of young adults with mean age of 24 years (Shield J 1984). Jacobelic & Jones (1987) reviewed 6 reported cases of hemangioendothelioma where all were infiltrative. had recurrence and metastasis; on CT all showed circumscribed and infiltrative orbital mass with enhancement with contrast. Commonest presentation being the rapidly progressive proptosis, displacement of eyeball and painful ophthalmoplegia. It can also present along with Tolosa Hunt's Syndrome (Messmer et al 1983). In our case also presentation was same except ophthalmoplegia which occurred on relapse of disease with involvement of inferior rectus muscle. In our case absence of any precipitating factor to cause sarcoma except the history of penetrating foreign body in eye makes an unique case. Initially the growth was slow and became rapid only after excision of primary growth.

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