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A RARE CASE REPORT OF PRIMARY ORBITAL FIBROSARCOMA

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Abstract : Fibrosarcoma is a rare slow growing but locally aggressive malignant neoplasm of mesenchymal origin. It represents less than 1 percent of adult soft tissue tumour usually present in the deep soft tissues of the extremities and bone.It is generally a diagnosis of exclusion especially as a primary tumour in the orbit. Management of this tumour requires wide local excision and even exenteration. We here report a rare case of 62 year old male who came with the complaints of protruding mass in the left eye for 6 months. This patient was diagnosed as a case of (query) Squamous cell carcinoma spindle cell neoplasm following incisional biopsy. Patient was then managed by total exenteration .Exenterated mass was later found to be primary orbital fibrosarcoma of orbit confirmed by histopathological examination and immunohistochemistry .Post operatively the wound was healthy and margins were found to be free of tumour on HPE and there is no recurrence
of the tumour on further follow up.

Keyword : Fibrosarcoma , Immunohistochemistry, Vimentin and Exenteration.

CASE REPORT :

62 years old male came to our hospital with chief complaints of mass protruding from the left eye for the past 6 months. Patient gave history of sudden increase in the size of the mass for the past 3 months associated with mucopurulent discharge and bleeding. Patient also gave history of loss of vision in the left eye for the past 4 months. There was a history of loss of weight and appetite since 3 months. There was no history of trauma, fever ,ENT symptoms or any other swelling in the body. There was no history of bone pain, cough with haemoptysis ,abdomen pain and seizures. Patient is a known hypertensive for the past 5 years under treatment. There was no history of any other systemic illness. Patient is a known smoker and alcoholic for 25years. On general examination, patient was emaciated with pallor. There was no icterus/cyanosis/clubbing/palpable lymph nodes. Vitals of the patient was within normal limits. Systemic examination of cardiovascular, respiratory, abdomen and central nervous system was within normal limits. On ocular examination, visual acuity of the right eye was 6/60 with -2.00 DSph 6/12 .Anterior segment examination was normal, pupil was reacting to light, PCIOL was present. Fundus examination was normal. On examination of the left eye, visual acuity was found to be no perception of light, Marked proptosis of the left eye was seen. Examination of the mass shows a large fungating mass measuring about 7x5 cm extending from eyebrow occupying whole of the upper eyelid till the inferior orbital rim. On palpation ,inspection findings were confirmed. Swelling was warm and tender, hard in consistency with indurated , irregular margins. Surface of the swelling shows areas of necrosis and ulceration . Globe architecture was totally distorted and details could not be made out(figure.1).

Figure 1 showing Large Left eye mass with necrosis and ulcerated areas extending hole of the upper eye lid till inferior orbital rim. Routine blood investigation like total count, differential count, haemoglobin and blood sugar was within normal limits. ESR was raised about 31mm at 30 min and 64mm at one hour. Imaging studies done with CT orbit and CT orbit contrast revealed a well defined non enhancing iso dense lesion encasing the entire left eye including the optic nerve and adjacent muscles with no significant bony destruction or intracranial extension (figure.2) . X ray chest and ultrasound abdomen was found to be normal.

figure 2 showing CT Orbit Contrast showing nonenhancing iso dense lesion encasing the entire left eye including the optic nerve and adjacent muscles with no significant bony destruction or intracranial extension Patient underwent incisional biopsy of the left eye mass. Histopathological examination of the mass was suggestive of squamous cell carcinoma or spindle cell carcinoma. Patient was planned for left eye exenteration under general anaesthesia after obtaining medical oncologist opinion who also suggested postoperative radiotherapy. The patient was then taken for exenteration and the excised specimen was sent for biopsy (figure.4).Post operative period was uneventful. Wound site was healthy(figure 6).Systemic antibiotics, analgesics was given and local antibiotic dressing was done.

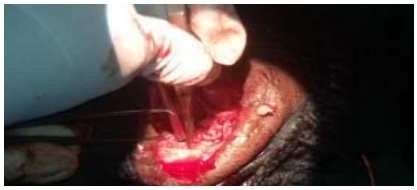

figure 3 showing intraoperative period during exenteration

Figure 4 showing exenterated specimen

Histopathological specimen of the exenterated mass showed a fascicles of spindle shaped cells with indistinct cytoplasmic borders. Nuclei elongated and pleomorphic with increased nuclear cytoplasmic ratio and prominent nucleoli with increased mitotic figures. And all margins of the exenterated mass was free of tumour cells (figure 5).Pathologist offered a differential diagnosis of Fibrosarcoma, Myoepithelial sarcoma, Malignant peripheral nerve sheath tumor and Poorly differentiated spindle cell variant of squamous cell carcinoma .

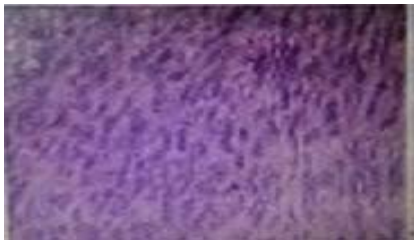

Figure 5 HPE image showing fascicles of spindle shaped cells with indistinct cytoplasmic borders. Nuclei elongated and pleomorphic with increased nuclear cytoplasmic ratio and prominent nucleoli with increased mitotic figures

Figure 6 showing exenterated socket with healthy wound The exenterated specimen was sent for immunohistochemistry. Immunohistochemistry studies revealed the mass to be positive for vimentin marker(figure 7) and negative for other markers like s100,desmin,smooth muscle antigen and epithelial membrane antigen thus confirming the diagnosis of mesenchymal tumour – fibrosarcoma .The tumour stage according to American joint committee on cancer was found to be Stage 3B(G3,T2,No,Mo) as it was a high grade tumour more than 5cm without nodal involvement or metastasis. The patient was advised to come for regular follow up. On monthly follow up for the past 6 months there is no evidence of recurrence. Patient was also advised for regular review at medical oncology department.

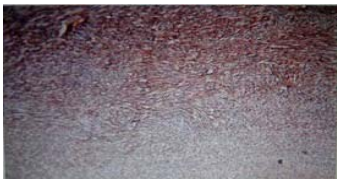

Figure 7 showing Immunohistochemistry staining showing strong positivity for **vimentin**

DISCUSSION:

Primary orbital fibrosarcoma is a rare locally aggressive malignant tumour of the orbit. It is a malignant spindle cell tumour of mesenchymal origin that occurs due to the hypercellular proliferation of immature fibroblasts. Fibrosarcoma infrequently affects the orbit as the most common occurence in the head and neck is distinctly rare other than in the nasal cavity. The orbit may be involved either primarily or by contiguity.

CLINICAL FEATURES:

Primary Orbital Fibrosarcomas commonly affects the elderly with mean age of 65 years. In children it can occur secondary to orbital irradiation for retinoblastoma. Primary orbital fibrosarcoma usually have a relative short history measured in months. They present as a enlarging orbital mass which may or may not be painful. Though they are slow growing they are locally aggressive due to infiltrative nature. Symptoms generally occurs due to mass effect of the tumour.

HISTOPATHOLOGICAL FEATURE:

Grossly the tumour is circumscribed but not encapsulated. They are firm, white and tan. Histopathologically the tumour is more cellular and composed of spindle shaped cells arranged in a collagen matrix to form fascicles giving rise to chevron like pattern. Ultrastructurally there is scanty cytoplasm, considerable rough endoplastic reticulum, no basement membrane, nuclei of the tumour cells are tapered which distinguish them from rounded nuclear ends of smooth muscle proliferations. There is little nuclear pleomorphism with 2 – 10 mitotic figures per 10 high power fields. Secondary histopathologic feature such as osseous and cartilaginous metaplasia as well as mucoid deposition can occur.

IMMUNOHISTOCHEMISTRY STUDIES:

Immunohistochemistry or IHC refers to the process of detecting antigens in cells of a tissue section by use of monoclonal/polyclonal antibodies.Fibrosarcomas are positive for vimentin and focally for smooth muscle actin demonstrating its myofibroblastic differentiation. Vimentin is a type 3 Intermediate filament protein that is a major cytoskeletal component expressed in mesenchymal cells. It is a cytoskeletal component responsible for cell integrity. Fibrosarcoma can be differentiated from amelanotic primary orbital spindle cell melanoma by their negativity for S1oo protein.

MANAGEMENT:

Management of this tumour is essentially surgical. Wide local excision and even exenteration is the treatment of choice. The tumour needs to be completely resected as the recurrent rates are higher with incomplete resection. Post operative adjunctive radiotherapy in doses of 5000-6000 cGY with shielding of the globe can be given. Chemotherapy is not effective. All patients should be followed up regularly to look for recurrences.

CONCLUSION:

This case is reported for its rarity as very few cases were reported in the literature and also to emphasise the importance of histopathological examination and imaging in diagnosing and managing the rare primary orbital tumours like fibrosarcoma and the role of immunohistochemistry in confirming the diagnosis.

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