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Case Report

Pathology Section

Primary Orbital Liposarcoma: A Case Report

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ABSTRACT

Liposarcoma is the most common malignant softtissue sarcoma in adults, commonly involving retro peritoneum and extremities. Liposarcoma of orbit is extremely rare. We report a case of primary orbital liposarcoma who underwent, exenteration in our centre. We have presented the clinical, radiological findings and histopathological features of the case along with brief review of literature.

Keywords: Diplopia, Malignant tumour, Soft tissue sarcoma

CASE REPORT

A 34 years old male reported to Ophthalmology OPD, Government Rajaji Hospital, Madurai, with the complaint of diplopia of six months duration. On examination the right eye was oedematous with soft swelling over upper eyelid. Extra ocular movements were restricted. Vision was 6/9. Cranial nerves and fundus were normal. Imaging studies [CT] revealed ill defined multi septate lesion containing fat and soft tissue arising from right eye suggestive of lipomatous tumor [Table/Fig-1]. After obtaining informed consent from the patient, biopsy was done and a diagnosis of liposarcoma was made. Later right orbital exenteration was done.

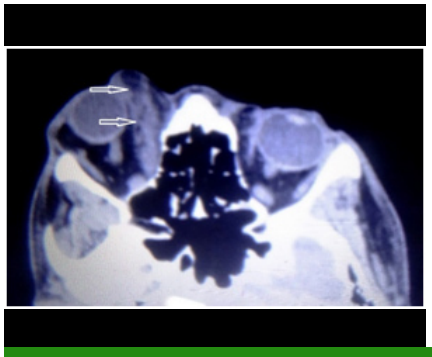
Orbital exenteration specimen was received measuring 6x5x3cm with attached skin and eyelid measuring 5x4cm. On cut surface, globe measured 2.5x2.5x2cm, Optic nerve 2cm with surrounding yellowish white soft tissue mass measuring 5x3x3cm. Skin margins were free [Table/Fig- 2].

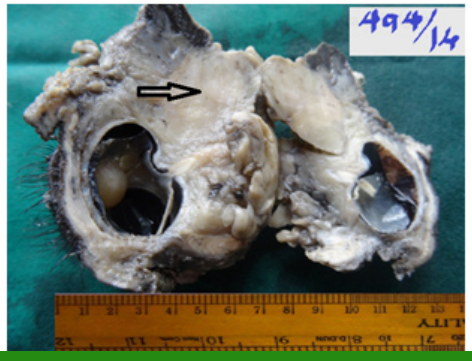
On microscopic examination the tumour was composed of predominantly mature fat cells intersected by fibrous bands and spindle cells with mild to moderate pleomorphism [Table/Fig-3]. Lipoblasts were seen as shown in [Table/Fig-4]. This was consistent with the diagnosis of well differentiated liposarcoma.

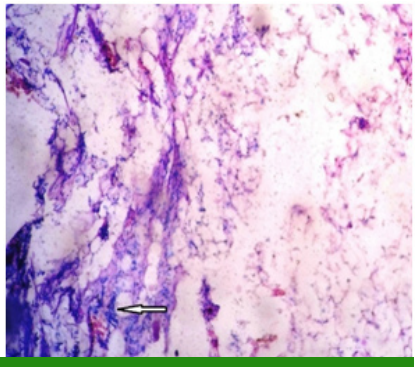
Unfortunately patient did not come for follow up after the surgery and hence further details are not available.

DISCUSSION

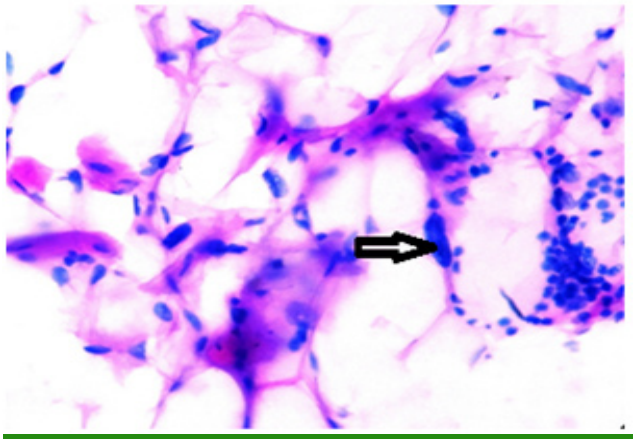
Liposarcoma is the most common soft tissue sarcoma in adults mostly involving the thigh and retroperitoneum [1]. Though large amount of fat is present in the orbit , it is a rare site for primary liposarcoma with only around 40 cases reported in the literature till date [2,3]. Metastatic liposarcoma of the orbit is even rarer [4]. Both genders are equally affected and the median age of occurrence is 31.5 years (range 17-71







[Table/Fig-1]: CT scan image showing ill defined multiseptate lesion arising from right eye **[Table/Fig-2]:** Orbital exenteration specimen showing yellowish white soft tissue mass (indicated by arrow) **[Table/Fig-3]:** sheets of adipocytes and stromal cells exhibiting nuclear atypia. (indicated by arrow)(Haematoxylin and eosin stain- H & E, 10x)



[Table/Fig-4]: shows lipoblasts (indicated by Arrow) (Haematoxylin and eosin stain- H & E, 40x)

years) [5]. There are no characteristic clinical features specific for liposarcoma. Patient may present with progressive mass, diplopia, proptosis, optic nerve compression with pain and visual impairment [6]. In primary liposarcoma no similar tumor will be evident in other parts of the body [7] as in our case. CT findings can vary from a homogenous lesion of soft tissue density to a heterogeneous mass containing fat [3].

Histologically, liposarcomas are characterized by the presence of lipoblasts. Lipoblasts have sharply outlined cytoplasmic vacuoles that indent or scallop atypical hyperchromatic nuclei. Liposarcomas are sub typed into well differentiated, myxoid, dedifferentiated, and pleomorphic variants. Among these subtypes, myxoid variant is the most common (56.8%) followed by well-differentiated (29.7%) [5].

This case was a well differentiated liposarcoma and had predominantly mature lipocytes with a variable number of pleomorphic spindle cells and multi vacuolated lipoblasts.

Main line of treatment is wide excision, which may necessitate exenteration in some cases. The role of radiotherapy and chemotherapy in the management of primary orbital liposarcoma is yet to be proved. The overall survival rate of orbital liposarcoma also is not well known because of its rarity [7].

CONCLUSION

Orbital Liposarcoma is a very rare malignant tumor .Treatment is surgical excision. Long term prognostic data of this tumor is not available due to its rarity. Ophthalmologists and Pathologists should consider orbital liposarcoma in the differential diagnosis when dealing with orbital masses.

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