ORBITAL HEMANGIOPERICYTOMA

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KEY WORDS : Hemangiopericytoma, orbit, proptosis

INTRODUCTION:

Hemangiopericytomas are uncommon tumors, but orbital hemangiopericytoma is a rare presentation.

CASE REPORT:

A 60 year old male, presented with a two years history of gradually increasing, painless proptosis of right eye. Examination revealed restriction of abduction movement of right eye with a matured cataract, CT scan showed a soft tissue shadow without bony involvement. While exploring, a single well circumscribed oblong mass of 4.5 x 3.0 x 2.0 cm was easily removed (Fig. 1). Removal of the tumor brought the abduction movement of right eye back the normal. No adjuvant therapy (Radiotherapy or chemotherapy) was given.

PATHOLOGICAL FINDINGS :

Gross : Single well circumscribed oblong mass of 4.5 x 3.0 x 2.0 cm; cut surface was solid yellowish

white with hemorrhagic and necrotic foci (Fig. 1).

Fig. 1

Gross photograph (cut surface) of orbital Hemangiopericytoma showing hemorrhagic and necrotic foci.

Microscopy : Revealed compressed vascular spaces surrounded by ovoid to spindle shaped uniform cells with scanty cytoplasm (Fig. 2). The number of mitotic figures were <5/10 high power field. At the periphery of the lesion, sinuous staghorn type vascular channels surrounded by tumor cells were seen. Reticulin stain confirmed the diagnosis of hemangiopericytoma (Fig. 3).

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Fig. 2

Photomicrograph of orbital Hemangiopericytoma showing compressed vascular spaces surrounded by ovoid to spindle shaped cells HE x 150.

Fig. 3

Photomicrograph showing tumor cells outside the darkly stained reticulin. Reticulin stain x 500.

DISCUSSION:

Hemangiopericytoma, usually an uncommon tumor, is found in deep soft tissues of lower extremities and retroperitoneum¹. An orbital hemangiopericytoma often presents with proptosis. It is circumscribed slow growing tumor of mean size upto 30 mm, rarely diagnosed preoperatively². In histologically diagnosed case it is very difficult to predict the behaviour of the tumor, whether malignant or benign. Tumor diameter more than 6.5 cm, mitotic figures >4/10 hpf, hemorrhage, necrosis, cellular anaplasia and increased cellularity associated with thrombosis are the criteria of malignancy, proposed by Enzinger and Smith³. MacMaster, et al stated that even mild degree of anaplasia with one mitotic figure /10 hpf

or moderate degree of cellular anaplasia with one mitotic figure /10 hpf are sufficient evidence to predict malignant potential of tumor⁴. In the present case although cellular anaplasia was minimal, rich cellularity, hemorrhage and necrosis were the features suggesting malignant potential of the growth. But even after 3 years of treatment patient did not show any recurrence or metastasis.

In well differentiated cases complete local excision is the primary treatment of choice but radical surgery with or without adjuvant radiotherapy is required for less well differentiated hemangiopericytoma⁵. It is found that chemotherapy gives partial or complete remission in metastatic hemangiopericytoma⁶. Local recurrence occurs in 22 to 55% of cases and some may metastasize.⁷

The prognosis of orbital hemangiopericytoma is considered better than those at other locations.

REFERENCES:

1. Searl SS, Ni C Hemangiopericytoma. Int. Ophthalmol. Clin. 1982; 22 : 141-162.
2. Roth SI, August CZ, Lissner GS, O' Grady RB Hemangiopericytoma of Lacrimal Sac. Ophthalmology 1991; 98 : 925-927.
3. Enzinger FM, Smith BH. Hemangiopericytoma, an analysis of 106 cases, Hum. Pathology, 1976; 7 : 61-82.
4. MacMaster MJ, Soule EH, Ivans JC, Hemangiopericytoma a clinicopathological study and long term follow-up of 60 patients, Cancer. 1975; 36; 2232-2244.
5. Staples JJ, Robinson Ra, Wen BC, et al, Hemangiopericytoma the role of radiotherapy, Int. J. Radiat Oncol Biol Phys 1990; 19 : 445.
6. Beadle GF, Hillcoat BL, Treatment of Advanced malignant hemangiopericytoma with combination adriamycin and DTIC, A report of 4 cases, J. Surg. Oncol. 1983; 22 : 167.
7. Jakobiec FA, Bilyk JR, Font RL, Orbit In : Spencer WH, editor, Ophthalmic Pathology : an atlas and text book, 4th ed. Philadelphia : WB Saunders, 1996 : 2545-2549.