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HEMANGIOPERICYTOMA OF NASOPHAYRNX A RARE CASE REPORT

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ABSTRACT

Hemangiopericytoma (HPC) is a rare tumour of uncertain malignant potential arising from mesenchymal cells with pericytic differentiation. It accounts for 3-5% of soft tissue sarcomas and 1% of vascular tumors. It usually presents in 5th to 6th decade of life. Most common sites are limbs, pelvis and head and neck. About 20% of all hemangiopericytomas are seen in head and neck, mostly in adults. Usually it presents in orbit, nasal cavity, oral cavity, jaw, parotid gland, parapharyngeal space, masticator space and jugular foramen. Long term follow up is important because of imprecise nature of the histological criteria for prediction of biologic behaviour.

We report a rare case of nasopharygeal hemangiopericytoma in a 19 year old male patient presenting with recurrent epistaxis and nasal blockage since 3 years.

KEYWORDS :

BACK GROUND

Hemangiopericytoma is a rare tumour type. It originates in a specific cell type called pericytes, identified by Rouget in 1873 and subsequently described by Zimmermann in 1923.

These cells are arranged alongside capillary vessels and have smooth muscle characteristics. They are responsible for vessel caliber regulation owing to their contractile capability, modulating both flux and permeability.

Regarding etiology, a past history of trauma, prolonged steroid use and hypertension are

Diagnosis is made histologically, but it is difficult to predict the behaviour of the tumour in an individual patient . It has low potential for local recurrence or metastasis, The treatment of choice is total local excision. Adjuvant radiotherapy and chemotherapy may be employed.

Patients with hemangiopericytoma should be regularly monitored for local recurrence and systemic tumour spread. .

CASE DISCUSSION

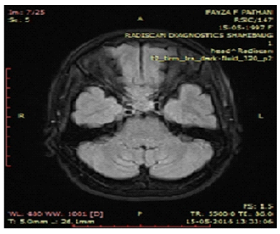
A 19-year old female presented to the ENT OPD ,Civil Hospital on with history of nasal obstruction, epistaxis, headache and difficulty in swallowing past 3 years.

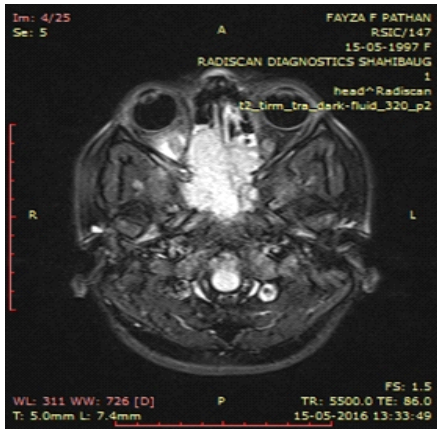
The patient has complaints of progressive painless loss of vision in right eye for past 2 months.

On examination, there was a fleshy polypoidal mass seen filling the bilateral nasal cavities(more on left side), with mild displacement of the septum to the right side. There was a minimal bleeding on touch.

Postnasal examination revealed the mass extending into the nasopharynx through the bilateral choanae.

IMAGING FEATURES





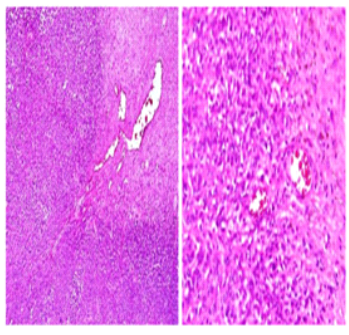
MRI reveals heterogenously enhancing mass lesion in midline in nasopharynx which is heterogenously hypointense on T1WI and hyperintense on T2/FLAIR images with intense enhancement on T1+Contrast scans.

The lesion extends into bilateral nasal cavities, with transphenoidal extension into sella and abutting optic chiasma, internal carotid arteries and anterior cerberal arteries.

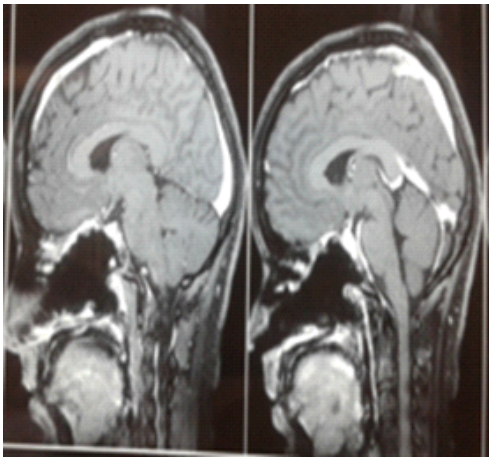
An endoscopic excision of the lesion was done.
Intraop findings suggest highly vascular tumour which bleeds on touch in bilateral nasal cavity.

BIOPSY

Showed vascular neoplasm with sheets of pericytes in between thin wall blood vessels consistent with hemangiopericytoma.



POST OP MRI



FOLLOW UP

The patient was subsequently planned for adjuvant radiotherapy to the tumor bed. He received a total dose of 45 Gy in 20 fractions at 2.25 Gy per fraction, over four weeks, by six Mega-voltage photons. The patient is on regular follow-up

DISCUSSION

HPC is a neoplasm of uncertain cell type origin. It is an uncommon spindle cell tumour, constituting 2.5% of soft tissue neoplasms and occurs primarily in adult life (median age 45 yrs, with peak prevalence in the sixth to seventh decade of life), appearing with an equal sex distribution . It is frequently aggressive and has a tendency to recur and metastasize. Approximately one-third of all HPCs occur in the head and neck. Primary parapharyngeal space HPCs are very rare neoplasms and, according to the international literature, only a few cases have been reported, with some of them referring to tumours that invade the parapharyngeal space from other sites of the head and the neck.

A painless mass with epistaxis and nasal discharge is the most common presentation of head and neck HPCs. The clinical behaviour varies depending on the different grading of each case. The diagnosis cannot be made on the basis of clinical and gross morphologic characteristics. Definitive diagnosis of HPC is provided by the accurate histopathologic assessment, which determines the accurate management and prognosis . The prediction of the clinical behaviour of HPC is not always clear and does not always correlate with the histolopathologic features of the tumour. Strict universal histopathologic criteria, for malignancy, have not been identified and vary between different studies. Generally, large size (> 5 cm), increased mitotic rate (> 4 mitosis/10 H PF), with the presence of atypical mitosis, high cellularity, pleomorphic tumour cells and foci of haemorrhage and necrosis predict a highly malignant course

MRI is the "gold standard" for accurate surgical planning. Imaging characteristics of the HPCs may be non-specific, demanding a differential diagnosis from other types of vascular or non-vascular tumours, especially those with prominent vascularization, such as juvenile haemangioma, glomus tumour, angiosarcoma, leiomyoma, leiomyosarcoma, schwannoma, mesothelioma, liposarcoma, benign and malignant histiocytoma, synovial sarcoma, chondrosarcoma, neuroblastoma, adenoid cystic carcinoma and mixed cell tumour .

The treatment of choice is radical excision of the HPC with a sufficient cuff of healthy tissue. Unfortunately, this is not always possible for para-pharyneal space tumours, making the need of adequate exposure mandatory, which is accomplished by stylo-mandibular tenotomy.

Adjuvant radiotherapy and chemotherapy may be employed, although the literature is not quite clear about their results. HPCs are considered to be relatively resistant to radiotherapy. Radiotherapy is reserved only as adjuvant therapy in cases of incompletely excised lesions, recurrent tumours, and tumours with high-grade histopathologic features. Although chemotherapy may have a role in the treatment of distant metastatic disease, its role in the primary treatment remains to be clearly defined

Vincristine, Adriamycin, and Cyclophosphamide have all been used with variable success in cases of aggressive HPCs . Some additional reports show that the use of interferon may be of benefit in patients with pulmonary HPC metastases .

HPC appears to present a survival rate that may vary from an overall 10-year rate of 70% to a considerably reduced 10-year survival rate in cases of > 4 mitosis/10 H PF (9%) or necrosis (29%) or tumour size > 6.5 cm (63%) . Some studies report local recurrence rates as high as 40%, with metastatic disease in 15% of the cases reported to have a latency period which varies from 63 to 107 months

Pathological appearance of resected HP is predictive of later metastatic potential. Long-term follow-up is necessary in patients even after radical resection because recurrence or metastasis may be delayed by many years.

The treatment of choice is radical excision, and the follow- up we propose includes clinical evaluation every 6 months and an annual MRI for at least 3 years

CONCLUSION

Nasopharyngeal space hemangiopericytoma is a rare soft tissue tumour of uncertain cell type origin, with high histological variability and unpredictable clinical and biological behaviour. One third of HPCs occur in the head and the neck and only a few cases have been reported regarding localization in the parapharyngeal space.

In nasopharyngeal space High index of suspicion is required to diagnose these cases. Extensive radiological as well as histological investigations need to be carried out. An attempt should be made for complete surgical resection. Postoperative radiation is indicated in cases of incomplete resection and recurrence.

The treatment of choice is radical excision, and the follow- up we propose includes clinical evaluation every 6 months and an annual MRI for at least 3 years.

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