Acta Scientific Neurology (ISSN: 2582-1121)

Volume 6 Issue 8 August 2023

Clinical Case Report

An Interesting Case of Sinonasal Tumor

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Received: May 28, 2023

Published: July 06, 2023

Abstract

Aim: Sinonasal Hemangiopericytomas are rare Uncommon Soft Tissue Tumor of perivascular myoid differentiation with hemangiopericytoma-like vasculature. The clinical behaviour of the tumor is difficult to predict and surgical excision remains the treatment of choice. We present one such case of Sinonasal Hemangiopericytoma operated at our centre recently.

Materials and Method: 28 year old female presented with a painless swelling over the forehead since 6 months with history of gradual increase in size of the swelling and complaints of double vision in left eye and anosmia. Radiological evaluation showed a T1 isointense T2 hyperintense lesion arising in the left nasal cavity with an intracranial extension. Patient was operated upon for the same and Histopathological evaluation suggested the lesion to be a sinonasal hemangiopericytoma. Post operative period was uneventful.

Conclusion: Sinonasal Hemangiopericytoma are rare vascular tumors with a variable clinical discourse and a difficult diagnosis. Surgical excision is the treatment of choice and patients require long term follow ups.

Keywords: Sinonasal Hemangiopericytomas; Anosmia

Introduction

- 28-year-old female with painless swelling over the forehead since 6 months
- Gradual increase in size of the swelling
- % Pain over left eye with diplopia since 1 month
- % Loss of smell (Lt > Rt) since 15 days
- No other significant associated history

Clinical examination

- 4X3 cm ovoid shaped swelling over the glabella
- Firm in consistency, non-fluctuant, non transilluminant, non pulsatile
- Anosmia (LT > RT)

Histopathology report

Section studied showed fragments lined by respiratory type ciliated pseudo stratified squamous epithelium, with underlying submucosa and bone. There is spindle shaped tumour beneath the bony layer. The tumour is highly cellular and shows spindle cells arranged in fascicles and storiform pattern. Tumour also shows stag horn vasculature with a few mitotic figures and the tumour is seen ulcerating through the sinus mucosa at a few places.

Impression: Spindle Cell Neoplasm suggestive of Sinonasal Hemangiopericytoma

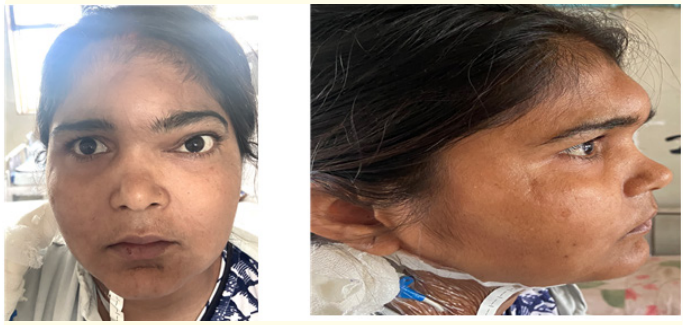

Figure 1

Discussion

Epidemiology

- Uncommon Soft Tissue Tumor of perivascular myoid differentiation with hemangiopericytoma-like vasculature
- 3~5% of all soft tissue sarcomas. 1% of all vascular tumors.
- Also called Glomangiopericytoma, sinonasal type
- CTNNB1 mutation with beta catenin oncogenic activation (beta catenin nuclear staining on IHC)
- Past history of trauma, Prolonged steroid use, Hypertension
- Recurrence rate - 27%
- Excellent long term outcome with surgery alone
- Broad Age Range (5 to 90 years). Mean in 7th Decade.
- F > M (1.2: 1). No difference in outcome based on gender after surgery. Bilateral Tumor uncommon
- Sites: Orbit, Nasal Cavity, Turbinate and Septum (occasionally in isolation), Maxillary and Ethmoid sinuses (in conjunction with nasal cavity), oral cavity, jaw, parotid gland, parapharyngeal space, masticator space, jugular foramen

Pathophysiology

- May arise from pericytes associated with vessels of the nasal cavity.
- Mutational activation of Beta Catenin
- Associated with cyclin D1 overexpression.

Clinical features

- Slow growing painless mass
- Nasal obstruction, mass, polyps, difficulty breathing.
- Sinusitis, Discharge, change in smell.
- Nasal Bleeding, Headache
- Rare association with oncogenic osteomalacia. Pain is characteristic

Radiology

- Plain X-rays of Sinonasal area are of limited value. Show the presence of a space occupying lesion with a mass effect distorting the adjacent bony structures.
- CT(P/C) shows the involvement of the soft tissues of the nasal cavity and paranasal sinuses and also any underlying bony destruction of the adjacent structures.
- MRI shows the tumor as a solid mass, isointense on T1 with diffuse enhancement. May appear isointense on T2 imaging. It helps in differentiating tumor tissue from inflammatory fluid caused by sinus obstruction. Contrast imaging helps in showing extension into the skull base. In highly vascular tumors, flow void may be seen.
- DSA useful in determining the vascular supply of the tumor.

Histopathology and immunohistochemistry

- Sinonasal Hemangiopericytomas may differ histologically from hemangiopericytomas arising from other tissues.
- Contain uniform oval or spindle shaped cells that form tight aggregates with small amount of intervening collagenous stroma. Mitosis is rare and necrosis is absent with stag horn like vascular spaces
- The Neoplastic cells do not stain for epithelial, melanocytic or neural markers.
- A finding of positive staining for cytokeratin excludes a diagnosis of Sinonasal Hemangiopericytoma
- The tumor cells are positive for vimentin (98%) and smooth muscle actin (92%)
- Special Stains for Reticulin show positivity surrounding individual tumor cells throughout the specimen.
- Electron Microscopy may show features such as thin cytoplasmic filaments and pinocytic vesicles which suggest pericytic differentiation.

Prognostic factors

- 5 year survival: 90%
- Recurrence rate: 27%. Long term clinical follow up recommended.

- Recurrences associated with long duration of symptoms, bone invasion and profound nuclear pleomorphism on histopathology.
- Factors to differentiate benign from malignant.

HPE - > 4 Mitosis per field, high cellularity, pleomorphic tumor cells
Gross - Size more than 6.5 cm, foci of haemorrhage and necrosis

Tumor behaviour

- Metastatic disease - more than 50% through the hematogenous spread
- Mostly metastasizes to lungs, liver, bone. Lymph node involvement is rare.
- In Head and Neck - 40% recur locally and 10% have distant metastasis
- Peculiar Biological behaviour of the tumor. Benign non mitotic Hemangiopericytomas have been reported to metastasize.

Management

- Surgery- Mainstay. Wide local excision
- Angiography and pre operative embolisation may be considered in large tumors
- Hemangiopericytomas are radioresistant. Hence Radiotherapy not considered as primary treatment and reserved for inoperable metastasis or treatment of post operative surgical fields in case of positive margins
- Adjuvant Chemotherapy has limited success. More useful in Infantile type [1-6].

Conclusion

- Relatively uncommon vascular tumors more so in head and neck
- HPC usually "defined" by reactivity for vimentin, with or without CD34 and CD57 and lacks other immunodeterminants of epithelial, neural and myogenous differentiation.
- Controversial Diagnosis - Determination between True Sinonasal Hemangiopericytoma vs soft tissue like Hemangiopericytoma
- Surgical Excision is the treatment of choice.
- Clinical Behaviour is difficult to predict.
- Close follow up for an extended period of time.

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