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CASE REPORT: HAEMANGIOPERICYTOMA – A RARE SINONASAL TUMOUR

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ABSTRACT Hemangiopericytoma (HPC) is a soft tissue tumor arising from Zimmermann's pericytes, which are modified smooth-muscle cells in the periphery of blood vessels. These pericytes are located outside the reticulin sheath of the endothelium [1,2]. HPC is found mostly wherever there is increased vascularity seen. A high incidence of local recurrence and metastasis is reported in HPCs. [3,5]

KEYWORDS : Haemangiopericytoma, Zimmermann's pericytes, laser, Vascular tumor

INTRODUCTION

HPC is an uncommon mesenchymal tumor, accounting for 1% of all blood vessel-related neoplasms and approximately 3% of all soft tissue sarcomas. HPC has a predilection for the long bones, pelvis and scapula; but, 15% occur in head and neck region, incidence of sinonasal HPC is less than 1%. Origin in oral cavity is less common.

HPC is a rare tumor of adult life mostly occurring in 5th decade and is very uncommon in children and accounts for only 10% of cases.[8] The architectural pattern of HPC can be seen in other mesenchymal neoplasms. The diagnosis of HPC is one of the exclusions and relies on the presence of characteristic histological features. As sarcomas, HPCs are graded based on histologic and biologic parameters. However, the pericytes, in which they originate, possess characteristics of both smooth muscle and endothelial cells. Differentiating these from other cell types is often challenging. Accordingly, the diagnosis of HPC is made on the basis of distinct architectural patterns exhibited histologically.[9,10]

Prior to the routine use of immunohistochemistry (IHC) in the diagnosis of HPC, misinterpretation for synovial sarcoma was common. However, unlike HPC, synovial sarcomas are often immunoreactive for both keratins and epithelial membrane antigen (EMA). Electron microscopy is also helpful in separating the two tumors, as epithelial differentiation is identified in some synovial sarcomas studied ultrastructurally.[12]

Increased mitotic activity, a higher cell density, an appearance of undifferentiated cells; and presence of necrotic and hemorrhagic areas in the tumor tissue are the major features seen during malignant transformation. HPC shows cytogenic abnormalities.[13]

Depending upon the size and location of the tumor, the HPC generally present as painless mass, but may have symptoms.[14]

Case study

A 32-year-old male patient presented to the ENT OPD with chief complaint of recurrent epistaxis from the right nasal cavity for the past 10-12 days. There was no history of trauma, no history of any bleeding disorders, and no significant past surgical or medical history. On Examination,

Ear: Bilateral Tympanic membrane intact

Nose: A small, bright red , 0.5cm X 0.5cm, polypoidal mass seen below the Inferior Turbinate on the lateral wall of Right Nasal cavity on Anterior Rhinoscopy, about 1.5 cm from the nostril. Right sided mild DNS present

Oral Cavity & Oropharynx: No abnormality clinically

Throat: No abnormality clinically

Nasopharynx: No abnormality clinically

Neck: No abnormality clinically

On INVESTIGATION,

XRAY NOSE & PNS showed Inferior turbinate shadows hypertrophied and PNS hazy.

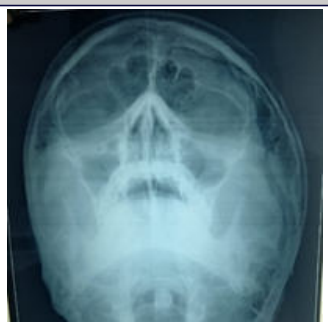

Fig. 1. X-Ray Nose & PNS

Routine blood investigations were within normal limits. Nasal Endoscopy showed a small, bright red ,0.5cm X 0.5cm , polypoidal mass seen below the Inferior Turbinate on the lateral wall of Right Nasal cavity . No other masses seen in the nasal cavity or nasopharynx. Histopathology showed sections of many small capillary spaces lined by flattened layer of endothelial cells with sheets of pericytes and collagenous tissue surrounding the vascular spaces. There is an area of ulceration with infiltration of moderate no. of polymorphs. A flattened layer of squamous epithelium is also seen wrapping the tissue at one area.

Impression: Haemangiopericytoma (Benign)

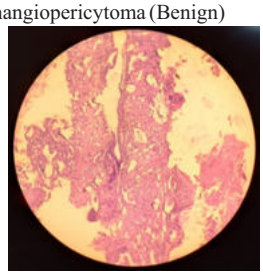

Fig.2. Histopathology slide on Low power magnification (10x)

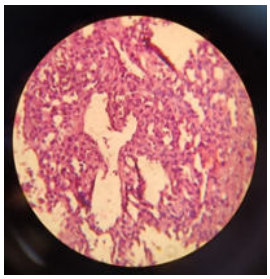

Fig.3. Histopathology slide on High power magnification (100x)

Treatment

Excision of Right lateral wall nasal mass under Local Anaesthesia by Diode Laser was done. No active bleed noted. Specimen was sent for Histopathological examination. Topical antibiotic and topical haemostatic agent was applied at the site of excision.

Per Op Findings :
0.8cm X 0.8cm haemorrhagic polypoidal mass seen below the inferior turbinate in the right nasal cavity. Nasal mass excised completely using diode laser

Fig. 4. Post-operative specimen

Patient was advised for a follow-up visit every 6 months.

DISCUSSION

In 1942, the tumor was first described by Stout and Murray.[4] Hemangiopericytoma is a soft tissue tumor derived from mesenchymal cells with Zimmermann's pericytic differentiation. It can be both benign and malignant in nature. Hemangiopericytoma constitutes only 3%–5% of all soft tissue sarcomas and about 1% of all vascular tumors. The head and neck incidence is about 15%.[4]

There has been dispute regarding the use of term for the tumor, some authors prefer “intranasal hemangiopericytoma—like tumors” and others prefer “sinonasal hemangiopericytoma” due to strong bound with soft tissue hemangiopericytoma.[25] However, in 2005, the World Health Organization (WHO) classification of head and neck tumors proposed the term glomangiopericytoma due to their similarity with glomus tumors. Nowadays, in clinical practice, the term “hemangiopericytoma” for all tumors with hemangiopericytoma-like histology has become accepted.[15]

Hemangiopericytoma can occur in any age groups, mostly in sixth to seventh decade of life. There is no gender predilection.[7]. Based on the patient's medical history and clinical findings, factors such as trauma, long-term steroid use, arterial hypertension and hormonal or metabolic imbalance could be the predisposing risk factors.[16]

Usually, tumor is located in the nasal cavity and presents with epistaxis and nasal obstruction.[17] Pain is usually absent and if present should be regarded as a sign of local infiltration.[18] Other infrequent symptoms are vision impairment, headache and local swelling. Majority of the tumors grow rapidly so the history is of short duration.[19]

On examination the mass is pinkish to whitish in color and firm in nature. On CECT scan of paranasal sinuses, well circumscribed soft tissue mass can be seen without any bone erosion giving the differential diagnosis of nasal polyps, inverted papilloma, solitary fibrous tumor and lobular capillary hemangioma. However, in large lesions, bone erosion of nasal cavity, paranasal sinuses and adjacent orbital walls and skull base have been reported.[20]

Based on cellular pleomorphism, mitosis and cellularity, these tumors are classified as low intermediate or high grade. Characteristics “staghorn” pattern is seen in sinonasal hemangiopericytoma but is not specific. Tumors consists of tightly packed cellular areas surrounding thin-walled branching blood vessels. Cells are ovoid to spindle shaped showing ill-defined cell boundaries. Immunohistochemical studies play an important role in strengthening the diagnosis. Vimentin and CD34 are considered the most reliable markers.[4]

The treatment of choice is wide-field excision with negative margins.[25] Other treatment modalities such as chemotherapy, radiotherapy and immunotherapy are reserved for metastatic disease, unresectable and recurrent tumors. However, the efficacy of radiation therapy is doubtful as these tumors are considered to be radioresistant.[21,22]

The prognosis of these tumors is not definitively predictable. According to the literature, the tumor has been reported to have recurrence rate from 9.5% to 50%, with an average time of recurrence rate of 6–7 years.[23] However, recurrence has been even reported 26 years after tumor resection, so it is mandatory to have regular, lifelong surveillance of such patients. The primary factor in recurrent disease is an incomplete primary excision.[24]

CONCLUSION

HPC is an uncommon vascular tumor and rarely occurs in the nose and paranasal sinuses. It's biologic behavior is difficult to predict when based solely on conventional histological parameters. It can be benign or malignant. Long-term follow-up is mostly necessary in patients even after radical resection because recurrence or metastases may be delayed by many years. [4]

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Conflicts Of Interest : None

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