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MALIGNANT GLOMANGIOPERICYTOMA : A CASE REPORT AND REVIEW OF LITERATURE

Pathology

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ABSTRACT

Glomangiopericytoma is a rare tumor of head and neck region with nasal cavity as a common location. It is a tumor of adult age group and patients present with nasal obstruction. It has characteristic histomorphologic and immunohistochemical features with indolent behavior . However rarely it may show aggressive behavior with atypical morphology including increased mitosis and necrosis. These patients have more recurrences and may metastasize.

KEYWORDS

Glomangiopericytoma, Immunohistochemistry, Nasal cavity

INTRODUCTION:

Glomangiopericytoma is a rare sinonasal tumor comprising around 0.5% of all sinonasal tumors and showing perivascular myoid differentitiation. [1] The common location is nasal cavity and is seen in both males and females with slight female predominance and common age is 5-7th decade. It has an indolent behaviour and on histopathological examination it shows oval to spindle cells with bland nuclear features without significant nuclear atypia, mitosis or necrosis. Admixed many branching blood vessels are also seen. There are case reports of glomangiopericytoma describing its classic morphology [2,3,4], but glomangiopericytoma with malignant features are very rare. Here we describe one such case.

CASE HISTORY

A 55 year old man presented to our hospital with history of recurrent nasal obstruction and epistaxis. Patient had a history of similar mass previously and was operated thrice but the mass again recurred. Previously at one time patient was operated in our hospital and the histopathological examination showed features of glomangiopericytoma. Twice he was operated outside and no records were available. This time CT scan showed a mass involving bilateral nasal cavities, left maxillary ethmoid and sphenoid sinus with destruction of sellar floor, bilateral nasal turbinates and medial wall of left maxillary sinus and radiological diagnosis was in favour of malignant lesion. The mass was removed in pieces and tissue was sent for histopathological examination. Grossly it consisted of multiple fragments ranging from grayish brown to hemorrhagic fragments.

Histopathological examination showed multiple fragments showing variable morphology with some showing necrosis, some showing a subepithelial tumor(fig.1, 2) composed of branching hyalinized blood vessels with surrounding spindle cells with elongated nuclei and fine chromatin. No nuclear atypia or mitosis was seen in these areas. Also identified many fragments showing oval to spindle cells with marked nuclear pleomorphism, brisk mitotic activity (13-33/10 hpf) including atypical mitosis and bizarre cells (fig.3). Focal areas of osteoid production and many hyaline globules were also seen in these areas. Immunohistochemistry was done which showed diffuse positivity for vimentin (fig.4) in both benign and malignant areas and focal actin positivity in the benign areas. Cytokeratin, S100, CD34 and desmin were negative. Ki67 labelling index was high (20-25% in the malignant areas). Based on these histopathological and immunohistochemical features, a diagnosis of glomangiopericytoma with malignant features was made.

DISCUSSION:

Glomangiopericytoma is a rare sinonasal tumor with characteristic histopathological and immunohistochemical features and indolent behaviour and is placed in the borderline/low malignant potential category in the WHO classification. It classically shows a submucosal tumor composed of bland oval to spindle cells with interspersed branching hyalinised blood vessels. Rarely it may show malignant features characterized by nuclear pleomorphism, necrosis and increased mitosis (>4/10 HPF). Though morphologically it resembles hemangiopericytoma of the soft tissue based on histopathological features, immunohistochemically and biologically, it is different from the latter. Compagno et al [5] in 1976 described this tumor as hemangiopericytoma like intranasal tumor which is currently called as glomangiopericytoma. In their study of 23 cases, the authors described the clinicopathological features of these tumors. According to this study, nasal cavity was the most common site and age was 6-7th decade. Nasal obstruction and epistaxis were common symptoms. The tumors showed little nuclear pleomorphism with minimal mitotic activity and no necrosis. No evidence of malignant features were seen on follow up. However according to authors, one case in a young patient, which was not included in the study, showed features of malignant hemangiopericytoma (pleomorphism, mitosis and necrosis) with metastasis.

Study by Thompson et al [6] evaluated 104 cases of sinonasal type haemangiopericytoma which included 57 females and 47 males with a mean age of 62.6 years and common symptoms of nasal obstruction and epistaxis. Nasal cavity was the commonest site. The tumors were submucosal and vascular with oval to spindle shaped nuclei and showed positivity for actin and vimentin. In this study, 18 patients developed recurrence within 1-2 years of diagnosis, 97 patients were either alive or dead but free of disease and three patients died with the disease. According to this study the majority of sinonasal type hemangiopericytoma behave in a benign fashion.

Tse et al [7] and Kuo et al [8] described six and five cases each of sinonasal hemangiopericytoma with their clinicopathological and immunohistochemical features. According to these studies, the tumor showed bland spindle cells with interspersed blood vessels and positivity for actin.

Recently study by Park et al [9] analyzed 337 cases of glomangiopericytoma and described the clinicopathological, immunohistochemical and prognostic factors of these tumors. In this study, pleomorphism and necrosis were present in 6 and 2 cases respectively. Recurrence or metastasis was present in 45 patients. Actin was positive in 86% and negative in 14% of patients whereas CD 34 positivity and negativity was seen in 24% and 76% of patients respectively . According to this study, actin immunonegativity or CD34 positivity were predictable of poor prognosis in patients with glomangiopericytoma. In the present case, patient had history of recurrences, histopathologically the tumor showed marked nuclear pleomorphism with brisk mitotic activity and atypical mitosis, high Ki67 labelling index and only weak patchy actin positivity and CD34 negativity. According to study by Kowalski et al [10] which included 2 cases of nasal cavity hemangiopericytomas, a proliferation index of 10% or greater may indicate a more aggressive behaviour of these tumors. The differential diagnosis of glomangiopericytoma includes solitary fibrous tumor, glomus tumor, synovial sarcoma, hemangioma and glomangiopericytoma can be differentitiated from other entities based on histopathological and immunohistochemical features.

In conclusion, we have described a rare tumor showing features of glomangiopericytoma with malignant features characterized by nuclear pleomorphism, brisk mitosis and high proliferation with multiple recurrences.

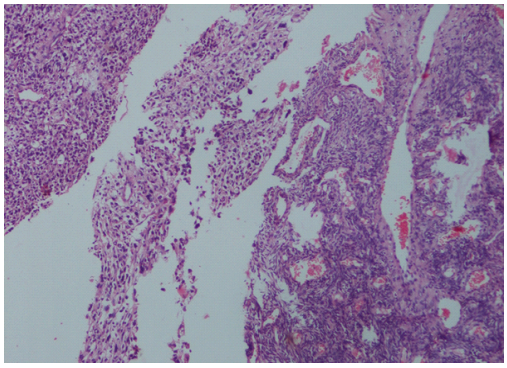

Fig.1-Low power view (4X) showing glomangiopericytoma and atypical focus

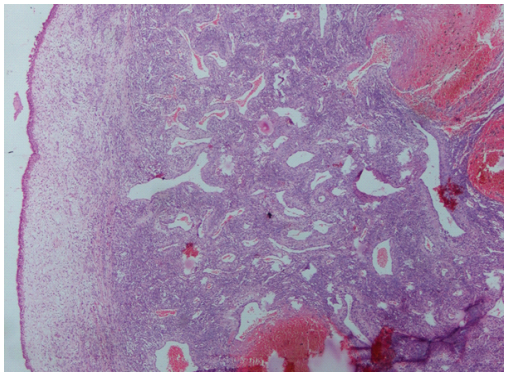

Fig 2 low power view showing subepithelial location of glomangiopericytoma

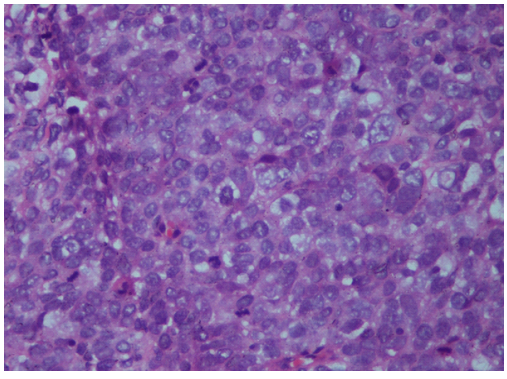

Fig .3 high power view (40X) showing nuclear atypia with brisk mitosis

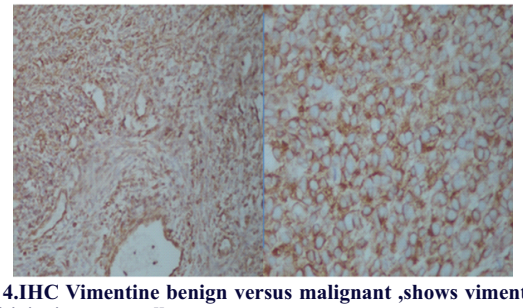

Fig 4.IHC Vimentine benign versus malignant ,shows vimentine positivity in tumor cells

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