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Case report

Clinico-pathological features of sino-nasal hemangiopericytoma: A case report and review of literature

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ABSTRACT

Hemangiopericytoma-like tumors are considered to originate from modified perivascular glomus-like myoid cell. Though it may arise from any part of the body, it is an uncommon tumor in the sino-nasal region. Sinonasal hemangiopericytoma-like tumor is important as it is recognized as a special variant of hemangiopericytoma because of its considerably more favorable prognosis. We encountered this rare case of sino-nasal hemangiopericytoma-like tumor in a 46-year-old- female with typical histological findings which helped in predicting the behavior of tumor and establishing proper line of management.

1. Introduction

Hemangiopericytoma-like tumors of nasal cavity are a peculiar form of vascular neoplasms probably related to traditional hemangiopericytoma but with somewhat different morphologic features, location, clinical setting and biological behavior [1]. Hemangiopericytoma-like intranasal tumors are thought to originate in a paranasal sinus and extend into the nasal cavity secondarily. They have been reported in elderly in their sixth and seventh decades of life and clinically mimic allergic polyps. These patients most commonly present with symptoms of nasal obstruction and epistaxis [2]. Imaging studies show nasal cavity or paranasal sinus opacification by a polypoid mass lesion. Sinonasal hemangiopericytoma-like tumor is an indolent lesion with an overall excellent survival (> 90 % 5-year survival) achieved with complete excision [3].

2.Case report

Forty-six year-old female presented with nasal obstruction for the past six months and few episodes of minimal epistaxis. Clinical examination revealed polypoid nasal mucosal thickening and showed congestive surface. Computed Tomography (CT) of paranasal sinuses was done to look for the extent and nature of the lesion and showed subtle hyperdense polypoid opacification of right maxillary sinus extending into right nasal cavity through mildly widened infundibulum. CT findings were consistent with a diagnosis of antro-choanal polyp. The patient underwent complete excision of lesion. Gross examination revealed fragmented pieces of gray white soft tissue of about 2 cc volume with fleshy consistency.

3.Microscopy

Sections showed subepithelial well delineated hypercellular tumor composed of tightly packed cells surrounding thin walled endothelial-lined spaces forming storiform pattern and short fascicles [Fig 1]. Large vascular spaces showed staghorn configuration [Fig 2]. The individual tumor cells were uniform, elongated with spindle-shaped nuclei and indistinct cytoplasmic borders. Occasional mast cells were noted. No cellular atypia, mitotic activity or necrosis noted in the sections studied. Histopathological diagnosis was that of 'hemangiopericytoma-like tumor'.

4.Discussion

Stout and Murray first described a highly vascularized tumor arising from Zimmermann's pericytes present in pericapillary connective tissue and named them as hemangiopericytomas [4]. Many soft tissue tumors showing various types of cellular differentiation were subsequently called as hemangiopericytomas. Although Mursahima [5] reported first case of nasal hemangiopericytoma, Campagno [2] as early as in 1976 described sino-nasal hemangiopericytoma-like tumors as distinct group from other soft tissue hemangiopericytomas.

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Figure 1. Photomicrograph (x10) of H & E stain shows hypercellular tumour composed of tightly packed cells surrounding the thin walled endothelial lined spaces forming storiform pattern and short fascicles. No cellular atypia or mitotic activity or necrosis noted.

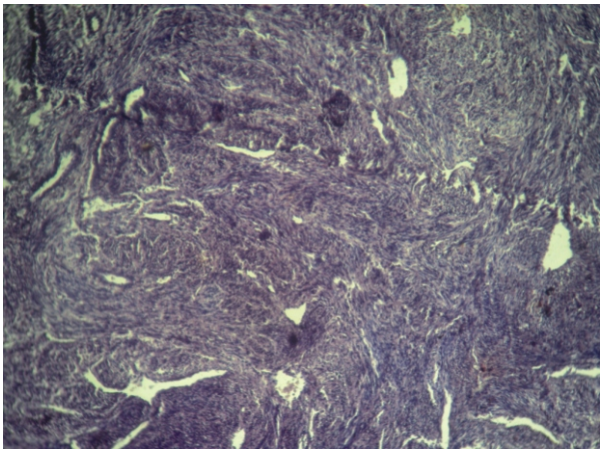
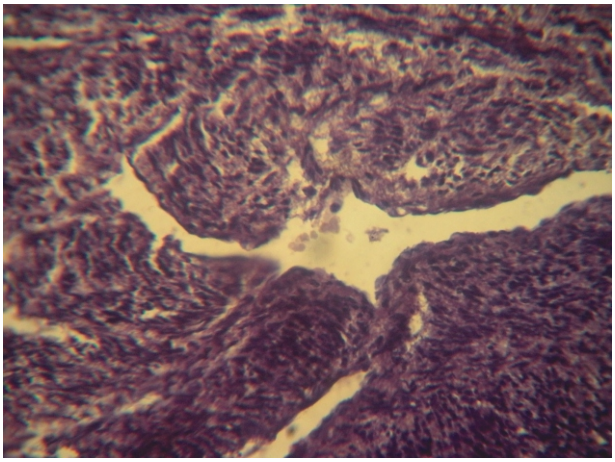


Figure 2. Photomicrography(x40) of H&E stained section showing large endothelial lined vascular space forming "stagorn configuration" with occasional mast cells.



Compagno et al described clinical, microscopic and gross features of 23 cases of intranasal hemangiopericytoma-like tumors and considered these lesions as a peculiar form of vascular neoplasm within the histological spectrum of traditional hemangiopericytoma. The microscopic criteria for these tumors were absence of mitotic activity, clear distinction of normal vessels from tumor cells, uniform spindle cells with little or no overlapping of cell borders, absence of necrosis and presence of scattered mast cells. Follow-up of these patients did not reveal any evidence of malignant or biologically unpredictable behavior [2].

But Eichhorn et al [6] considered them as similar to other soft tissue hemangiopericytoma with local recurrences in as much as 4 out of 9 cases with follow-up (44%) but low metastatic rates and thought to reflect on the factors like early presentation and small tumor bulk apart from difficulty of complete local resection.

Eichhorn et al found more cases of nasal origin compared to Compagno who reported sinus origin of these lesions with secondary nasal involvement. Eneroth et al [7] reported cases with recurrence and late metastasis and found poor correlation between histological findings and grade of malignancy.

Immunohistochemical studies have shown sino-nasal hemangiopericytomas are positive for vimentin and focally for actin, but negative for desmin and S-100 and these features were similar to glomus tumors [8]. Recent WHO classification includes them together as 'glomangiopericytomas' [3], and further reports on immunohistochemistry of glomus tumors showed positivity for caldesmon which is not seen in pericytes [9].

Initially described as arising from pericytes, hemangiopericytomas both sino-nasal and soft tissue types are now believed to arise from modified perivascular (actin-positive) glomus-like myoid cell [10]. Granter [10] described perivascular spindle and round cell tumors of myoid origin and divided them as myofibromatosis, hemangiopericytomas and myopericytomas. Hemangiopericytomas showed short, spindled or ovoid cells, with uniform nuclear morphology and palely eosinophilic cytoplasm [11].

Watanabe et al [9] in their comprehensive review subdivided the reported cases of sino-nasal hemangiopericytoma into three groups – soft tissue hemangiopericytomas with plump spindle cells showing nuclear atypia, true hemangiopericytomas (previously called as hemangiopericytoma-like tumors) showing myoid differentiation with good clinical outcome, and those reported as nasal glomus tumors.

Hemangiopericytoma-like tumor may be confused with angiofibromas [1] and solitary fibrous tumors [12] in the sino-nasal location clinically and on imaging. Imaging cannot differentiate these rare lesions from the more common antro-choanal polyps and inverted papillomas. Angiofibromas are locally destructive lesions composed of fibrovascular tissue of varying maturity arising form or adjacent to nasopharyngeal wall [1]. The tumor occurs exclusively in young men and is much less cellular than the hemangiopericytoma-like tumor and contains mainly fibrous component. Solitary fibrous tumors are rare lesions showing spindle cells with 'ropy' keloidal collagen bundles and thin-walled vascular spaces apart from CD34 and bcl positivity in immunohistochemistry [3].

Fletcher in his review considered sino-nasal hemangiopericytomas to be of pericytic origin [13]. In spite of using immunohistochemistry and electron microscopy for the differential diagnosis and to elucidate the origin of these tumors, the finality of their cell of origin still remains inconclusive partly due to the rarity of these unusual lesions and hence the diagnosis of hemangiopericytoma-like tumor is still remains mainly histopathological [14] and on clinical follow-up.

5.References

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