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Case Report

Sinonasal haemangiopericytoma- An unusual swelling in the nasal cavity

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ABSTRACT

Sinonasal hemangiopericytoma, also known as glomangiopericytoma (GPC) is a benign perivascular tumor of low malignant potential, with an incidence rate of 0.5-1% of all sinonasal tumors. It often shows local recurrences with invasion to the surrounding bony tissues. It usually arises from the perivascular cells surrounding the capillaries. Etiology of the tumour is uncertain, with few postulated hypothesis like pregnancy, trauma, hypertension and corticosteroid drugs. They have an indolent course and tend to occur most commonly in the adults of seventh decade of life. We present a case report of 55 years old male presenting to the ENT OPD, with complaints of right sided nasal obstruction and repeated episodes of epistaxis since the last 9 months. History and general examination was unremarkable except for anti-hypertensive intake since the last few years. Laboratory investigations showed microcytic hypochromic anaemia, raised absolute eosinophil count of 1500 cells per microliter and positive Hepatitis B surface antigen (HBsAg) in the blood. Rhinoscopic examination showed a reddish brown nodular swelling in the right nasal cavity beneath the middle turbinate and hypertrophy of left nasal mucosa. CT imaging suggested an 18x15 mms, well defined soft tissue mass in the right nasal cavity. Complete resection of the mass with endoscopic surgery was performed. Histopathological examination coupled with immunohistochemistry confirmed the diagnosis of Sinonasal Hemangiopericytoma. This case report stresses upon various differential diagnosis of sinonasal swellings and the importance of considering long term follow up of Sinonasal Hemagiopericytoma.

1. Introduction

Sinonasal hemangiopericytoma also known as glomangiopericytoma (GPC) was first described by Stout and Murray in 1942.[1] This tumour is hypothesised to originate from the pericytes surrounding the capillaries.[2] These are uncommon perivascular tumours of low malignant potential with good prognosis after surgical resection.[3]

Hemangiopericytoma represents 3-5% of all soft tissue sarcomas and 1.0% of all vascular tumours.[4,5] Location of hemangiopericytoma is variable: 15-30% cases are found in the head and neck region; of which sinonasal hemangiopericytoma account for 5.0% of the cases.[6–9] Females are predominantly affected with peak incidence in the seventh decade of life.[2] In 2005 WHO classification of soft tissue tumours, hemagiopericytoma was identified as a distinct entity and reclassified as borderline and low malignant potential soft tissue tumour of the nose and paranasal sinus.[10]

They differ from conventional hemangiopericytoma in anatomical location, pathogenesis and microscopic features. Conventional hemangiopericytoma are more common in soft tissues of the trunk and extremities with variable and uncertain etiologies and occur as a result of diffuse proliferation of plump spindle tumour cells with characteristic staghorn shaped vascular channels. They are usually less aggressive than sinonasal

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hemagiopericytomas.[2]

2. Case Summary

A 55 year old male presented to the ENT out-patient department, with history complaint of night nasal obstruction since the last 9 months and dull intermittent pain in the nasal and malar area for 15 days. Nasal obstruction was episodic and associated with sneezing. He also complained of repeated episodes of epistaxis for the last 1-2 months; one episode every 15-20 days apart, which was relieved by local cold compression and/or application of local vasoconstrictors. Past history revealed regular antihypertensive intake since the last 5 years.

On examination, he was afebrile with mild pallor. Systemic examination was unremarkable. Rhinoscopic examination (including diagnostic nasal endoscopy) showed a reddish brown nodular swelling in the right nasal cavity beneath the middle turbinate with hypertrophy of the left nasal mucosa. Routine investigations like haemogram, liver function test, renal function test, lipid profile and electrocardiogram were normal except for the presence of microcytic hypochromic anaemia and raised absolute eosinophil count of 1500 cells per microliter. Triple test showed positive HbsAg and negative anti-HBc, IgM anti-HBc and anti-HBs in the blood.X-ray imaging of the chest and abdomen showed normal studies. CT scan of head showed an 18x15 mms, well defined heterogenous soft tissue mass in the right nasal cavity with no erosion of the bony turbinates. On the basis of clinical workup, a provisional diagnosis of benign soft tissue tumour of nasal cavity was made. Our patient underwent endoscopic sinus surgery for complete removal of the mass with 5mm clear margins.

Grossly the excised mass was single, firm brownish, 2x1.5x0.9 cms in size. Cut section showed homogenous white nodular areas with multiple foci of haemorrhage. Microscopic examination showed mucosal lining of stratified squamous cell with diffuse proliferation of plump to spindle shape tumour cells beneath the mucosa, with hyperchromatic nucleus and indistinct cytoplasm. Mild anisonucleosis with minimal mitotic activity (1/10 HPFs) was appreciated. Dense aggregates of tumour cells around staghorn shaped vascular channels lined by endothelial cells with no interconnection between vessels, with dense bands of perivascular hyalinisation was also noted (Figures 1 and 2). No foci of necrosis was seen and resection margins were free of tumour cells.

Immunohistochemical study showed strong cytoplasmic positivity of smooth muscle actin (SMA) in tumor cells (Figure 3) and diffuse strong membranous positivity of CD34 in cells lining the vascular channels (Figure 3). Immunomarkers like caldesmon, androgen receptor, cytokeratin, desmin and S100 showed negative expression. Based on histopathological and immunohistochemical findings, a diagnosis of Sinonasal Hemangiopericytoma) was given. Our patient is doing well after 2 months of follow up period.

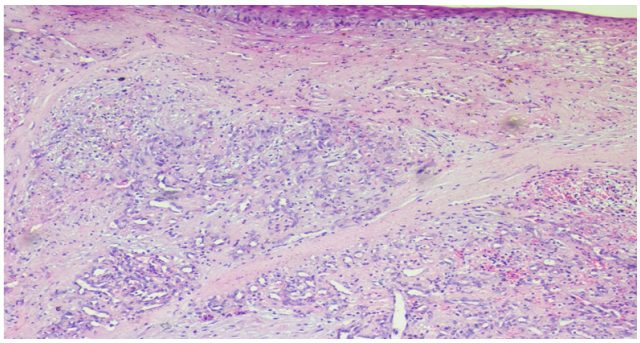

Fig. 1: Microscopic examination showed diffuse proliferation of plump to spindle shape tumour cells beneath the mucosa, with hyperchromatic nucleus with mild anisonucleosis and indistinct cytoplasm. Dense aggregates of tumour cells around staghorn shaped vascular channels lined by endothelial cells with no interconnection between vessels, with focal perivascular hyalinisation was also noted. Hematoxylin and Eosin x 10X.

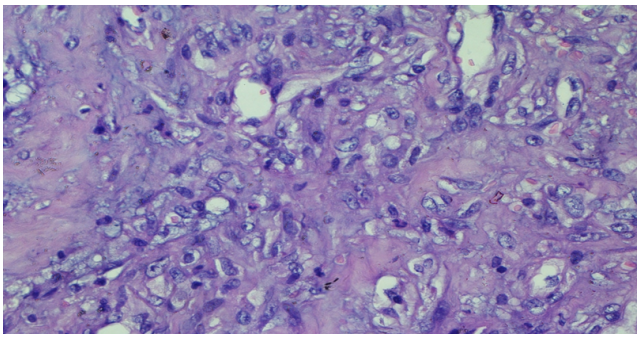

Fig. 2: High power of Figure 1. Hematoxylin and Eosin x 40X.

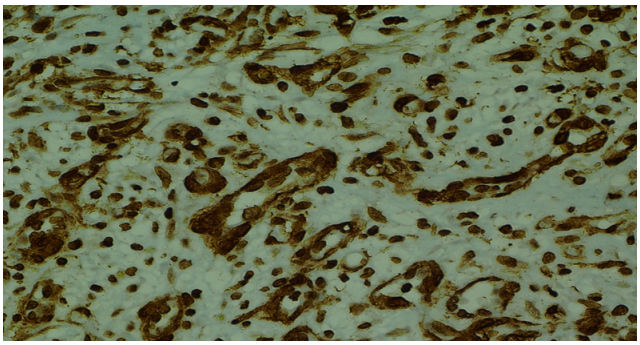

Fig. 3: Immunohistochemical study showed strong cytoplasmic positivity of smooth muscle actin (SMA) in tumor cells. IHC SMA x40X

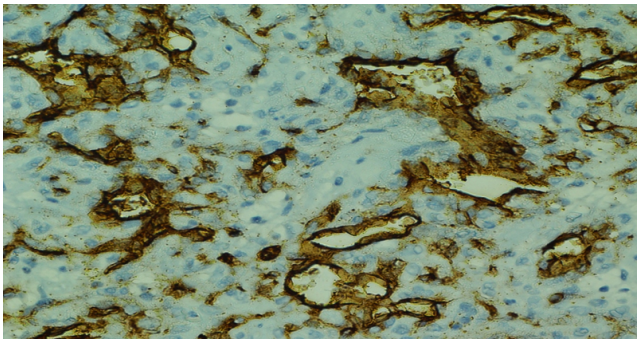

Fig. 4: Immunohistochemistry showed diffuse strong membranous positivity of CD34 in cells lining the vascular channels. IHC CD34 x40X.

3. Discussion

Hemagiopericytoma is a unique type of glomus tumour with typical vascular channel proliferaton, ranging from staghorn shaped vessels to branching of vessel with focal hyalinisation. Hemangiopericytoma arising in the sinonasal cavity tend to have an indolent course and show a characteristic morphology with pericytic proliferation similar to glomus tumor.[7,8] Therefore hemangiopericytoma arising in sinonasal cavity are now termed as sinonasal hemangiopericytoma or glomangiopericytoma and represent a separate entity from soft tissue hemangiopericytoma.[11]

Hemangiopericytoma predominantly affects females with peak incidence in the seventh decade of life.It occurs mostly in nasal the cavity as compared to the paranasal sinus.[11]The common symptoms are nasal obstruction followed by epistaxis and impaired nasal breathing. Symptoms such as proptosis, squint and visual loss secondary to nerve compression may also be observed due to invasion/compression of surrounding orbit.[12,13]

Plain X-ray is of limited value in the diagnosis, except for the presence of space occupying lesion with compression of surrounding bony structures. CT and MRI are better in assessing the extent of tumour prior to surgery. CT helps in demonstrating bony destruction in the nasal cavity, paranasal sinus and the orbits while contrast enhanced MRI helps in differentiating tumour tissue from inflammatory fluid caused by sinus obstruction secondary to other causes. Digital angiography is often useful to assess the vascularity of tumour, prior to preoperative tumour embolization.[11,12]

Grossly the tumour size ranges from 1 to 8 cm with a mean size of 3.1 cm. It issoft, solid homogenous, friable lesion with haemorrhagic and gelatinous cut sirface.[2]Microscopy shows spindle cell neoplasm of round to oval to spindle cells cell hyperchromatic nuclei, inconspicuous nucleoli and scant eosinophilic cytoplasm in syncytial pattern. Mild nuclear pleomorphism and occasional mitotic activity is seen, but necrosis is absent. Characteristic dilated staghorn like vascular configuration and frequent perivascular hyalinisation are also seen. Other growth patterns are diffuse, fascicular, storiform and whorled. Squamous metaplasia, tumour giant cells and myxoid degenerations are also seen.[13,14]Immunohistochemistry shows strong diffuse positive staining for smooth muscle actin (SMA), vimentin and β catenin in 90.0% cases. CD34 is negative in tumour cells but focally positive in vascular lining cells. These tumours are typically negative for Desmin, AE1/AE3, S100, XIIIa, CD99, CD117, STAT6, ERG, caldesmon, cytokeratin and SOX10.[15]

Major differential diagnosis of Hemangiopericytoma are lobular capillary hemangiomas, solitary fibrous tumours, leiomyomas, nasopharyngeal angiofibromas and glomus tumour.[16–18]Lobular capillary hemangioma (LCH) also called as pyogenic granuloma is a benign soft tissue tumour. Nasal septum is the common location of LCH involving the aerodigestive tract in 1/3rdof cases. Clinical profile, sign and symptoms are similar to Hemangiopericytoma with polypoidal masses with intact surface mucosa in both the tumours. LCH differs from hemangiopericytoma as it occurs in male aged less than 18 years with male to female ratio of 4:1, as against a female preponderance in hemangiopericytoma. LCH is smaller in size than hemagiopericytoma with mean size of 1.7 cm. On microscopy LCH shows a lobular configuration with small irregular vascular spaces and a large central feeding ectatic vessel with small branches with more disorganised arrangement of cells lining the vascular spaces. Mitotic figures are frequent in LCH as compared to hemangiopericytoma. On IHC LCH shows diffuse positivity of CD34 and CD31 and Smooth Muscle Actin show sparse positivity of perivascular cells. LCH are easily treatable by local excision and rarely recur.[19]

Solitary fibrous tumours (SFT) are benign mesenchymal soft tissue lesions can involve virtually any organ. They are similar to pleural solitary fibrous tumour. 1/3rd of extrapleural SFT's occur in head and neck region and 15% of these cases involves the sinonasal region. Sinonasal SFT comprise only 1% of all sinonasal neoplasms. Both sexes are equally involved with mean age of presentation of 52 years with a clinical profile similar to hemangiopericytoma. Nasal obstruction and epistaxis are the most common presenting complaints. Microscopically they are similar to hemangiopericytoma with diffuse spindle cell proliferation with area of hypo- and hypercellularity with intact surface epithelium and overlapping "patternless" growth, with cells showing irregular nuclear shape and contour with prominent coarse collagen bundles intermingled with neoplastic spindle cells. Prominent variable size "staghorn vasculature" are also noted but SFT's differs from hemangiopericytoma as they are larger in size with a mean of 5.25 cm. Perivascular hyalinisation is absent in

SFT. Immunohistochemical expression of SFT shows strong positivity of CD34 in neoplastic spindle cells and focal scant positivity of smooth muscle actin. Strong immunostaining with STAT6 is characteristic of SFT owing to NAB2-STAT6 gene fusion. Sinonasal SFT are less aggressive with low recurrence rate.[20–22]

Sinonasal Leiomyoma is very rare with incidence rate of less than 1%. Mean age of presentation is 40 years with slight female predilection with male to female ratio of 1:1.5. They have a favourable prognosis and rarely recur. They show dense spindle cell proliferation with occasional staghorn vasculature as in Hemangiopericytoma. Leiomyoma differs from hemangiopericytoma as it is more eosinophilic owing to abundant eosinophilic cytoplasm and more elongated spindled cells with coarse chromatin and occasional perinuclear vacuoles and in a prominent fascicular pattern. Immunohistochemistry of leiomyoma show strong desmin positivity.[23,24]

Nasopharyngeal angiofibroma is a soft tissue tumour of adolescent males. Most common site is posterolateral nasal cavity near sphenopalatine foramen. It is a locally aggressive mesenchymal tumour with local bony erosion and intracranial extension with a recurrence rate of 5-10%. Angiofibroma differs from hemangiopericytoma with low cellularity diffuse proliferation of evenly spaced bland spindle cells with vesicular chromatin and eosinophilic collagen fibrils in the intervening stroma. Immunohistochemical expression of angiofibroma show positivity for androgen receptor and β catenin.[25,26]

Poor prognostic factors of hemangiopericytoma are large size (>5cm), bone invasion, profound nuclear pleomorphism, increased mitotic activity (>4/10 HPFs), necrosis and high proliferation index (>10%).[17] Metastasis to distant site occur in 5-10% cases. Recurrance rate of hemangiopericytoma range from 8-53%, owing to inadequate surgical excision. In view of variable recurrence rate, lifelong follow up is advisable.[27] Our case did not show any bony invasion with no active mitosis and necrosis.

4. Conclusions

Sinonasal hemangiopericytoma is a rare condition but it should always be kept in the differential diagnosis of soft tissue swellings with nasal obstruction and epistaxis. Owing to its variable recurrence rate, lifelong follow up should be observed after surgical removal and on recurrence, prompt surgery followed by radiotherapy and chemotherapy should be administered.

5. Source of Funding

None.

6. Conflict of Interest

None.

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