Indian Journal of Pathology and Oncology 2023;10(2):112–113

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Indian Journal of Pathology and Oncology

Journal homepage: www.ijpo.co.in

Editorial

An enigmatic rare tumor of sino-nasal cavity

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ARTICLE INFO

Article history:
Received 07-06-2023
Accepted 11-06-2023
Available online 17-06-2023

Hemangiopericytoma was first described by Stout and Murray as highly vascularized tumor arising from Zimmermann's pericytes present in pericapillary connective tissue.[1] Subsequently, many soft tissue tumors showing various types of cellular differentiation were called as hemangiopericytomas. Campagno[2] as early as in 1976 described Sino-nasal hemangiopericytoma-like tumors as distinct group from other soft tissue hemangiopericytomas. Although Mursahima[3] reported first case of nasal hemangiopericytoma.

Haemangiopericytoma is a rare entity comprising only 1% of all vascular tumors and only 5% are localized in the nasal cavity or paranasal sinuses.[4] Generally, Haemangiopericytoma behaves aggressively but literature review and our case study suggest benign nature of this lesions occurring in paranasal sinuses and skull base.[5]

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 like 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 like glomus tumors.[8] Recent WHO classification includes them together as. 'glomangiopericytomas',[9] and further reports on immunohistochemistry of glomus tumors showed positivity for caldesmon which is not seen in pericytes.[10]

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.[11] Granter 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

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https://doi.org/10.18231/j.ijpo.2023.025

and palely eosinophilic cytoplasm.[12]

Watanabe et al[10] 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[13] and solitary fibrous tumors[14] 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 from or adjacent to nasopharyngeal wall.[13] 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.[9]

Fletcher in his review considered Sino-nasal hemangiopericytomas to be of pericytic origin.[15] Despite using immunohistochemistry and electron microscopy for the differential diagnosis and to elucidate the origin of these tumors, the finality of their cell of origin remains inconclusive partly due to the rarity of these unusual lesions and hence the diagnosis of hemangiopericytoma-like tumor is still remains mainly histopathological[16] and on clinical follow-up. Wide surgical resection was done in both cases. Five years after surgery, there was no recurrence in case report 1 patient neither by endoscopy nor by MRI. Patient of case report 2 is asymptomatic on follow-up after two years of surgery. Recently, Georg et al treated Sino-nasal hemangiopericytoma by an endoscopic controlled endonasal tumor resection after embolization with Onyx and there was no recurrence after one year follow-up.[17] Duval reviewed 194 cases of Sino-nasal hemangiopericytoma till 2013 and found no significant difference between rate of recurrence for endoscopic or open resection. Incomplete excision was the most important predictor of recurrence. Radiotherapy may be advantageous in cases of incomplete surgical resection.[18,19]

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Cite this article: Bhanushree C S. An enigmatic rare tumor of sino-nasal cavity. *Indian J Pathol Oncol* 2023;10(2):112-113.