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Commentary

The Arced Proboscis-Sinonasal Glomangiopericytoma

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Sinonasal glomangiopericytoma is an infrequently discerned, soft tissue neoplasm demonstrating differentiation akin to perivascular myoid cells. Tumefaction is pervaded with hemangiopericytoma-like vasculature. Previously designated as sinonasal type hemangiopericytoma, glomus tumour or intranasal myopericytoma, neoplasm is additionally denominated as glomangiopericytoma, sinonasal type.

Commonly, tumefaction is confined to nasal cavity and manifests with nasal obstruction, epistaxis or sinusitis. Neoplasm is accompanied by genomic mutation within CTNNB1 gene along with oncogenic activation of β catenin and immune reactivity to nuclear β catenin.

Singular surgical extermination of the neoplasm is accompanied by superior prognostic outcomes wherein proportionate tumour reoccurrence emerges at ~27%.

The exceptionally encountered sinonasal glomangiopericytoma comprises of < 0.5% of primary sinonasal neoplasms. Mean age of disease emergence is the 7th decade whereas tumefaction may implicate paediatric subjects of ~5 years to elderly individuals of ~90 years.

A mild female predilection is encountered with female to male proportion of 1.2:1. Surgical outcomes appear non concurrent with gender predisposition [1,2].

Sinonasal glomangiopericytoma preponderantly incriminates nasal cavity. Occasionally, nasal turbinate or nasal septum may singularly be implicated. Apart from nasal cavity, maxillary sinus or ethmoid sinus may be involved. Bilateral neoplasms are uncommonly discerned in ~5% instances.

Sinonasal glomangiopericytoma may be engendered from pericytes concurrent with vascular articulations confined to nasal cavity. Tumefaction exemplifies mutational activation of β catenin and manifests with overexpression of cyclin D1 [1,2].

Sinonasal glomangiopericytoma originates from and exhibits genomic mutations within CTNNB1 gene. Polymerase chain reaction (PCR) and genetic sequencing enunciates single nucleotide substitution within exon 3 codon of CTNNB1 glycogen serine kinase 3 beta phosphorylation region [1,2].

Sinonasal glomangiopericytoma exhibits cogent clinical symptoms as dyspnoea, nasal obstruction, polyps or tumefaction confined to the nasal cavity. Besides, sinusitis, nasal discharge, nasal haemorrhage, parosmia or headache may ensue. Features such as oncogenic osteomalacia are exceptionally encountered.

Cytological examination exhibits spindle shaped, epithelioid or spherical cells with indistinct cellular perimeter [1,2].

Frozen section exemplifies sub epithelial accumulation and proliferation of bland, spindle shaped and spherical cells segregated from superimposed respiratory epithelium. Foci of haemorrhage, stromal or perivascular hyalinization and staghorn vascular articulations may be encountered [2,3].

Upon gross examination, a polypoid, beefy red or greyish pink tumefaction with superficial haemorrhage is observed. Mean tumour diameter of 3 centimetres is enunciated. Cut surface appears as soft, oedematous and fleshy [2,3].

Upon microscopy, superimposed respiratory epithelium or metaplastic squamous epithelium appears intact. Sub epithelial neoplastic cellular proliferation appears demarcated from superimposed epithelium by a distinct grenz zone [2,3].

Generally, neoplasm exemplifies a diffuse, fascicular, solid or focally whorled pattern of tumour evolution besides a perivascular configuration which demonstrates miniature, prominent, thin walled, submucosal vascular articulations surrounded by aggregates of tumour cells.

Spindle shaped, spherical or elliptical tumour cells are pervaded with variable cytoplasm, spherical nuclei and an indistinct cellular perimeter. Occasionally, multinucleated tumour cells are observed.

Tumour cell aggregates are permeated with inflammatory cells as mast cells and eosinophils. Foci of minimal cellular or nuclear atypia may emerge [2,3].

Mitotic activity is absent or exceptional. Tumour necrosis is absent. Incriminated vascular articulations appear as prominent blood vessels with 'staghorn' countenance and perivascular hyalinization.

Ultrastructural examination exhibits a basal lamina which encompasses individual cells, tapered cytoplasmic extensions and orderly dissemination of filament bundles [2,3].

Sinonasal glomangiopericytoma appears immune reactive to β catenin, cyclin D1, CD99, vimentin, smooth muscle actin (SMA), muscle specific actin (MSA), factor XIIIa, laminin or D2-40.

Sinonasal glomangiopericytoma is immune non reactive to ERG, CD31, CD34, factor VIII or keratin [4,5].

Sinonasal glomangiopericytoma requires segregation from neoplasms such as glomus tumour, lobular capillary haemangioma, solitary fibrous tumour, Ewing sarcoma or Ewing-like sarcoma.

Sinonasal glomangiopericytoma can be appropriately discerned with cogent clinical symptoms and discernible polypoid tumefaction confined to the nasal cavity [4,5].

Appropriate examination of surgical tissue sampling enunciates a soft tissue neoplasm delineating aggregates of tumour cells reminiscent of perivascular myoid differentiation and appear immune reactive to nuclear β catenin, cyclin D1 or smooth muscle actin (SMA).

Tumour cells appear immune non reactive to diverse vascular markers. Majority of neoplasms are devoid of diagnostic biochemical or haematological parameters [4,5].

Tumefaction may exceptionally be associated with osteomalacia, serum hypophosphatemia and elevated levels of serum alkaline phosphatase. Computerized tomography (CT) and magnetic resonance imaging (MRI) exemplifies a lobulated, polypoid tumefaction confined to nasal cavity or paranasal sinus. Besides, bone erosion or sclerosis may accompany the neoplasm.

Sinonasal glomangiopericytoma can be appropriately managed with surgical extermination of the neoplasm [4,5].

Prognostic outcomes following surgical intervention are superior with 5 year survival at ~90%. Tumour reoccurrence ensues in ~27% lesions. Therefore, extended clinical monitoring is recommended as tumour reappearance may be delayed.

Tumour relapse is associated with extended duration of clinical symptoms, neoplastic invasion within circumscribing bone and profound nuclear pleomorphism [4,5].

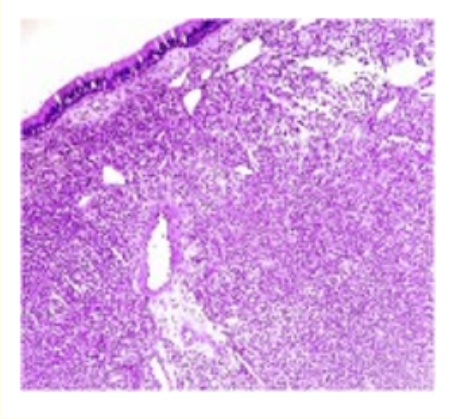

Figure 1: *Sinonasal glomangiopericytoma composed of spindle shaped and elliptical cells imbued with variable cytoplasm, spherical nuclei and indistinct cellular perimeter. Fascicles of tumour cells appear subjacent to superimposed respiratory epithelium. Tumour is invaded with inflammatory cells and prominent vascular articulations with staghorn appearance [6].*

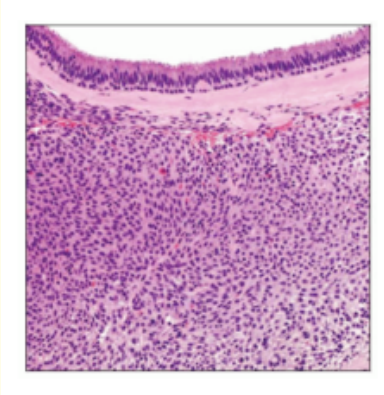

Figure 2: *Sinonasal glomangiopericytoma delineating superficial respiratory epithelium with a distinct grenz zone and subjacent aggregates of spindle shaped and elliptical tumour cells pervaded with variable cytoplasm and indistinct margin. Tumour cell aggregates are infiltrated by mast cells, eosinophils and innumerable vascular articulations with staghorn appearance and perivascular hyalinization [7].*

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6. Image 1 Courtesy: Science direct.

7. Image 2 Courtesy: Basic medical key.

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