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A Classical Case Of Histiocytic Sarcoma: A Rare Haematopoietic Neoplasm

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Abstract:
Histiocytic sarcoma is a rare haematopoietic tumor of histiocytic differentiation. Generally they are concurrent to other neoplasms or morphologically similar to them. This is the reason why they are oftenly missed on diagnosis. Here in our case report we have discussed a case of a 61 year old man who presented with a nasal mass. A long list of differentials like lymphomas, sarcoma, melanoma, metastasis etc were kept in count. However after applying large panel of various immunohistochemical markers it was finally ruled out to be a case of histicoytic sarcoma which is an aggressive haematopoietic neoplasm.

Keywords: *Histiocytic sarcoma, Immunohistochemistry, CD 68, CD 163*

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I. Introduction:

Histiocytic sarcoma is a rare but aggressive haematopoietic tumor of histiocytic differentiation. They are derived from mature monocytes/ macrophages. In many cases they get misdiagnosed as Diffuse Large B cell Lymphoma or anaplastic large cell lymphoma. Histiocytic sarcoma is an extremely rare malignancy accounting for nearly 1% of all the haematopoietic neoplasm. It is more common in males than females with an incidence ratio of approximately 3:2. The age distribution shows a bimodal peak at 0-30 years and 50-70 years. Some of the cases occur concurrent to Non Hodgkins lymphomas like ; hence they oftenly get missed on diagnosis. Histiocytic sarcoma commonly presents as a painless solitary mass at extranodal sites like gastrointestinal tract, soft tissue, skin, spleen, liver and nasal cavity. Generally the clinical features are non specific like night sweats, fever and weight loss. In this case report we have discussed a case of a 61 year old male who presented in OPD with a mass in nasal cavity with a complain of epistaxis; And after a long work up it was concluded to be a case of histiocytic sarcoma.

II. Case History:

A 61-year-old man presented with a mass in nasal cavity which was rapidly progressive in size. The mass occupied entire right nasal cavity and maxillary antrum. The patient had no co morbid conditions. There was also no significant family history. There was no association with fever, cough or voice change. However patient complained of mild dyspnoea and several episodes of bleeding from nose. The patient also had a history of a mass on level 2 right side neck which was surgically excised outside 6 months back and was revealed to be Cutaneous Lymphoid Hyperplasia on histopathology. In view of current nasal mass, surgical excision was performed from various areas of the nasal cavity and the specimens were sent in multiple containers depending on the area of excision for histopathological examination. The largest soft tissue piece measured 6x5x4 cm in size. Random sections were taken, processed and stained with haematoxylin and eosin. On microscopy, ciliated columnar epithelial lining was seen. The underlying stroma had mucosal glands and was infiltrated by atypical cels of medium to large size, round to irregular nucleus, dispersed chromatin, prominent nucleoli and moderate amount of cytoplasm. Background showed proliferating capillaries and inflammatory infiltrate. Large areas of necrosis were also present. Immunohistochemistry (IHC) staining was performed. The cells showed strong positivity for CD45, CD 68 and vimentin. Ki67 showed 80% cell positivity. Cells were negative for EMA, Synaptophysin, CD 5, CD 4, CD 56, CD 34, CD 79, CD 8, CD 99, CD 79, Tdt, CD 1a, S100, ALK, CD30 and MPO. Hence based on the morphological and immunohistochemical findings, we suggested the diagnosis of Histiocytic Sarcoma

III. Discussion:

Morphologically the tumor comprises of non-cohesive proliferation of large cells. The individual neoplastic cells are large and round to oval in shape. However few areas with spindle cell like features may also be present. The cytoplasm is generally abundant and eosinophilic. It may be foamy, vacuolated or clear. The nuclei are generally large, round to oval, grooved, indented, convoluted, irregularly folded. Eccentric placement of nuclei is oftenly seen. Occasional bizarre cells with pleomorphic, hyper chromatic nuclei and coarse chromatin may also

be found. Mitotic activity varies from case to case. Necrosis may or may not be present. Reactive inflammatory activity by lymphocytes, plasma cells, polymorphs, eosinophils and benign histiocytes is oftenly found in background.

Based on differential diagnosis, entities like carcinoma, lymphoma (Hodgkins as well as non Hodgkins), Ewings sarcoma, haematopoietic stem cell neoplasm, leukemia, melanoma as well as metastasis were kept in consideration.

EMA ruled out it to be a carcinoma. CD20, CD79, CD 4, CD 5, CD8 and CD56 eliminated B, T and NK cell lymphoma whereas CD 30 ruled out Hodgkins lymphoma. CD 34 disregarded haematopoietic stem cell precursors. CD 99 and S100 excluded Ewings sarcoma and melanoma respectively. Tdt and MPO rejected ALL and Myeloid series neoplasm respectively. CD1a helped in precluding LCH. ALK ruled out

IV. Conclusion:

Histiocytic sarcoma is a rare and aggressive neoplasm with poor response to therapy. There is no standardised treatment yet. Based on its morphology it can mimic other conditions and hence can go unnoticed or misdiagnosed. Amongst various immunohistochemical markers, CD 68 and CD 163 are good, reliable and easily reproducible markers for differentiation of histiocytic sarcoma. However they are not specific or exclusive for this condition. A combined clinicopathological evaluation with molecular studies is necessary for definitive diagnosis of this rare kind of neoplasm.

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