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CASTLEMAN DISEASE – A RARE CASE REPORT OF NON-CLONAL LYMPHOPROLIFERATIVE DISORDER PRESENTING AS A HUGE NECK MASS

Pathology

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ABSTRACT

Castleman disease is an uncommon lymphoproliferative disease also known as angiofollicular lymph node hyperplasia. It can be classified as unicentric or mutlicenteric type based on the clinical presentation. Histological variants can be either hyaline vascular or plasma cell type. Here we present a case Castleman disease occurring as a huge cervical lymphnode swelling in a young man. Surgical excision was done & hyaline vascular variant of Castleman disease was confirmed on histopathological examination. In addition, we discuss the pathogenesis, clinical features and reported comorbidities of unicentric and multicentric type and evaluate effective treatment strategies based on the results of lymph node biopsy and careful staging. The aim of this discussion is to revisit this very rare condition and emphasise the importance of histology to confirm the diagnosis from other lymphoproliferative conditions. Kaposi sarcoma & HIV are closely associated with this disorder and hence require thorough investigation.

KEYWORDS

Castleman disease, Hyaline vascular, Lymphoproliferative disorder

INTRODUCTION

Castleman disease is a rare lymphoproliferative disorder with unknown cause. [1] It may be either unicentric or multicentre. [2] Unicentric Castleman disease (UCD) is localized and carries an excellent prognosis, whereas multicentric Castleman disease (MCD) is a systemic disease occurring most commonly in the setting of HIV infection and is associated with human herpesvirus 8. [2,3] Unicentric Castleman disease (UCD) is rare, and there are no reliable estimates of its incidence in the population. [4] Here we discuss a case of UCD diagnosed in a young male presenting with huge neck mass.

Case Report

A 24-year-old male patient presented with a swelling on left side of the neck since two years which was gradually increasing in size. The swelling was not associated with pain, fever, difficulty in breathing or swallowing. Past & family history did not reveal anything significant. On examination a solitary swelling on the left side of neck about 9x8cm was noted extending from midline to posterior triangle of neck and one cm below the angle of mandible to supraclavicular area. It was firm in consistency and had a smooth surface. (Figure 1).

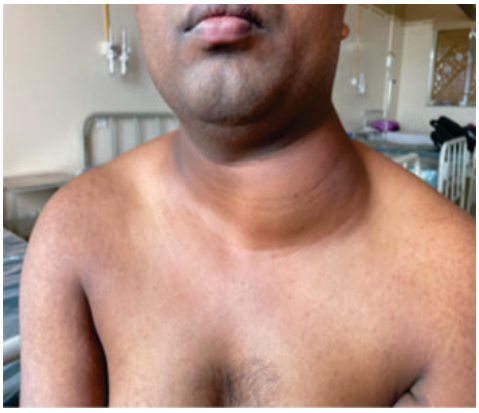

Figure 1: Clinical image of huge neck mass

No other lymphodes were palpable and patient did not have any organomegaly. Systemic examination was normal in the patient. Ultrasonography showed a homogenous lesion with internal vascularity in the lateral aspect of neck and Magnetic imaging resonance (MRI) showed a large well defined vertically oval lesion on the left side of neck with epicentre of the lesion at the junction of anterior and posterior triangle without vascular encasement (Figure 2a & b).

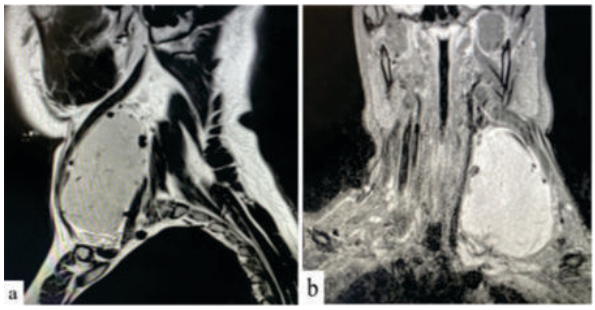

Figure 2a & 2b: Magnetic resonance imaging showing a large well defined oval lesion displaying markedly hyperintense signal intensity on T2W TIRM sequences.

A diagnosis of Schwanomma was made on imaging studies. Patient was negative for HIV & routine blood investigations showed no significant change. Fine Needle Aspiration was attempted but it yielded only hemorrhagic aspirate. Excision biopsy was done eventually. Gross examination showed a well circumscribed soft tissue mass measuring 7.5x6x3.5cm. External surface was nodular. (Figure 3a) Cut surface showed a solid, homogenous, grey white mass. (Figure 3b).

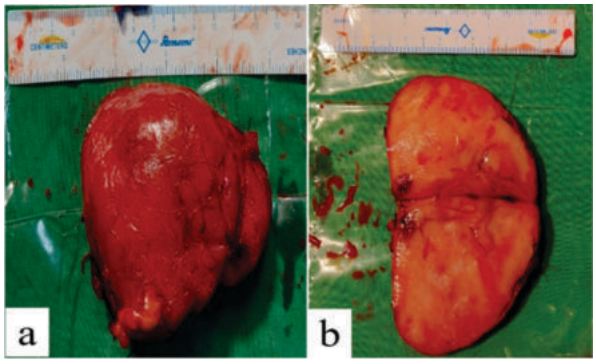

Figure 3a: Gross photograph of enlarged lymphnode **3b**: Cut section showing homogenous solid lesion

Microscopy revealed an enlarged lymph node with atretic germinal center (Figure 4a) showing marked vascular proliferation with hyalinization. Marked expansion of mantle zone consisting of

concentric layers of lymphocytes creating an onion skin appearance (Figure 4b). Penetrating blood vessels in the germinal centers at right angle giving it a lollipop like appearance (Figure 4c). Twinning, i.e. more than one germinal center with in a single mantle was also noted. (Figure 4d). The interfollicular stroma showed extensive vascular proliferation admixed with lymphocytes.

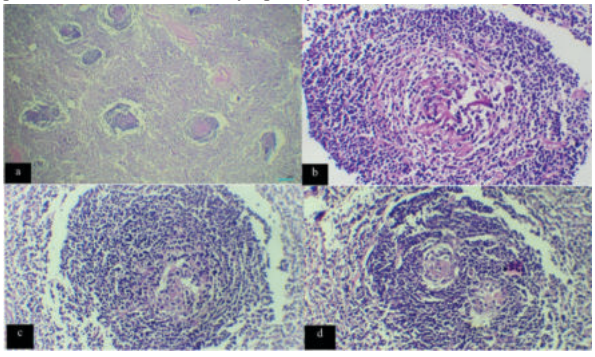

Figure 4 a: Microphotograph showing atretic germinal centre surrounded by broadened mantle zone (H&E x 100) **b**: Microphotograph showing marked expansion of mantle zone with concentric layers of lymphocytes (Onion skin appearance) & hyalinised blood vessels in germinal centre (H&E x 400). **c**: Microphotograph showing blood vessels penetrating the germinal centre at right angle (Lollipop appearance) (H&E x 400). **d**: Microphotograph showing fused mantle zone with more than one germinal centre (Twinning) (H&E x 400)

A final diagnosis of Castleman disease -Hyaline vascular variant was rendered. Patient remains asymptomatic since 6 months of follow-up.

DISCUSSION

Castleman disease is a rare, heterogeneous disorder characterized by atypical proliferation of lymphocytes. [2] Clinically two types of the disease are described i.e. unicentric & multicentric. UCD, first described by Castelman, is a localized disorder with no sex predilection & involving single lymphnode or single extranodal site or Oligocentric type (multiple lymphnodes of single group). The frequent sites involved are abdomen, thorax & peripheral lymphnodes. [1] It is usually asymptomatic or presents with pressure symptoms. MCD, involving more than four group of lymphodes, described by Gaba et al, is a systemic disease occurring most commonly in association with HIV and Human Herpesvirus 8 infection.[2,3] Lymphomas, Follicular dendritic cell sarcomas, Kaposi sarcoma, paraneoplastic pemphigus have also been known to occur with Castleman Disease.[5] MCD has 3 subtypes: human herpes virus-8 (HHV-8)-associated MCD; polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS)-associated MCD; and idiopathic MCD (iMCD).[5,6] iMCD can be further subclassified into iMCD–thrombocytopenia, ascites, reticulin fibrosis, renal dysfunction, organomegaly (iMCD-TAFRO) or iMCD–not otherwise specified (iMCD-NOS).[3] Male subjects are slightly more often affected with MCD than female subjects.[3,7] Systemic symptoms which includes fever, significant weight loss, weakness & hepatosplenomegaly are attributed to interleukin 6 production in these cases. [3,6] They usually have cytopenia, elevated erythrocyte sedimentation rate, C reactive protein, abnormal liver enzymes & renal function tests.[5,8]

On imaging studies Castleman disease can have varied appearance. UCD usually manifests with a hyperenhancing lymph nodal mass and should be considered in the differential diagnosis of lymphoma, metastasis, and infectious and/or inflammatory diseases that result in lymphadenopathy.[9] However in our case it was misdiagnosed as Schwanomma. Histopathological examination becomes very necessary to diagnose these cases. Major histologic subtypes of Castleman disease are Hyaline vascular, Plasma cell and mixed varints.[2,3] The hyaline-vascular histology accounting for most cases of UCD are characterized by central hyalinised vessel surrounded by onionskin pattern of lymphoid proliferation.[1] The spectrum of MCD histopathological features which include patients with regressed germinal centers and prominent vascularization are considered to fall on the hypervascular end of the spectrum, whereas hyperplastic germinal centers with prominent plasmacytosis are considered to fall on the plasmacytic end of the spectrum.[3] Castleman disease shows polyclonal population of lymphocytes on immunohistochemistry, low proliferation index & follicular dendritic cells expressing CD35 & CD21.[10] Immunhistochemistry is needed only to differentiate it from lymphoma or thymoma. Cytological diagnosis varies based on the architectural changes in lymphnode & may not be specific to the disease.[10] Even in our case cytology was not contributory.

UCD cases usually have excellent prognosis only requiring local excision, as was noted in our case. If surgery is contraindicated or lymph node is in inaccessible sites, therapies such as radioablation, embolization have been tried.[5]Treatment protocol for MCD cases are varied, which includes excision, use of preoperative immuno modulators or chemotherapy, antiviral treatment, glucocorticoids, thalidomide, interferon-alpha, and molecular targeted therapies.[5]

CONCLUSION:

Castleman disease is an uncommon lymphoproliferative disorder often misdiagnosed due to atypical or incidental clinical manifestations observed during the workup of other pathological diseases. Histopathological evaluation is diagnostic. Unicentric and multicentric disease are distinct clinical entities that mandate different therapeutic approaches.

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