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Editorial

Midline destructive lesions: A diagnostic challenge

Banushree C S ^{1,*}¹Dept. of Pathology, Indira Gandhi Medical College, Puducherry, India

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ABSTRACT

Midline destructive lesions (MDLs) are a diagnostic challenge due to an extensive differential diagnosis and vague presenting signs and symptoms. It may be due to neoplastic, autoimmune, traumatic, infectious, or unknown. The lethal lesions are characterized by ulcerative destruction of midline structures of the face like the nose, paranasal sinus and palate. A spectrum of diseases with myriad clinicopathological features can present as midline destructive lesions. Immunohistochemistry has played a major role in discerning the wide range of diseases into specific categories over the years.

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1. Introduction

Midline destructive lesions were called by various nomenclatures in the past, as lymphomatoid granulomatosis, polymorphic reticulosis, idiopathic midline granuloma, lethal midline granuloma, midline nonhealing granuloma, Stewart's syndrome. The feature usual to all this is destructive ulcerative lesions leading to functional and cosmetic deformity as a result of loss of tissue. The evaluation of these lesions by immunohistochemistry and molecular genetics studies has helped in further categorizing the lesions.^{1,2} A spectrum of diseases is included under MDLs. The most recent entity to be added to the list is IgG4-related disease (IgG4-RD).³ The other entities included in the spectrum can be infectious, neoplastic, autoimmune, trauma and many a times unknown.⁴

2. Discussion

The commonest cause of MLDs is Nasal type T-NK cells Lymphoma, followed by autoimmune vasculitis.⁵ A complete work-up, including clinical history, imaging

studies, pathological, serological and molecular studies, should be carried out to narrow down the differential diagnosis. In many cases, in spite of complete work-up, the cause cannot be discernible.⁶ Nasal type T-NK cells lymphoma is not a common type of Epstein-Barr virus (EBV) associated lymphoma but commonly presents as MDLs. In any MDLs, histopathological examination and ancillary tests are pivotal in establishing the diagnosis. In Nasal type T-Natural Killer cells Lymphoma, a mixed population of atypical lymphocytes is seen with Angiocentric and Angio-invasive features. On Immunohistochemistry, cytoplasmic CD3ε+, CD56+ and germline T-cell receptor (TCR) positivity and on In-situ hybridization, EBV encoded RNA can be demonstrated.^{7,8} Wegener's Granulomatosis (WG) is a multi-system disease characterized by necrotizing granulomatous inflammation which is immune mediated, and can present as MLDs. Histologically, necrotizing vasculitis along with noncaseating multinucleated giant cell granulomas in an inflammatory background helps in the diagnosis of WG. The presence of granular diffuse cytoplasmic staining for antineutrophil cytoplasmic antibodies on serology further substantiates the diagnosis of WG.^{9,10}

* Corresponding author.

E-mail address: drbanushree15@hotmail.com (Banushree C S).

The other diseases which can present as MDLs are cocaine-induced midline lesions, IgG4-related disease, leishmaniasis and fungal infections.¹¹ Cocaine-induced midline lesion (CIML) can show extensive necrosis and antineutrophil cytoplasmic antibodies on serology mimicking WG, but multinucleated giant cells, fibrinoid necrosis, and perivascular inflammatory infiltrate are not seen in CIML.¹²

A rare manifestation of cutaneous leishmaniasis can be MLD, especially in the endemic region. The dermis can show a spectrum of changes ranging from necrotizing inflammatory lesions to granuloma. The molecular confirmation by a polymerase chain reaction to demonstrate leishmania may be required in such cases.¹³ A rare type of systemic fibro-inflammatory condition like IgG4-related disease can present as MDL. The IgG4-related disease will have characteristic histology of obliterative phlebitis with storiform fibrosis along with elevated serum IgG4 levels. The plasma cells present in the lesion will also show positivity for IgG4.¹⁴

3. Conclusion

Midline destructive lesions (MDLs) are a diagnostic dilemma due to exhaustive list of differential diagnosis and non-specific presenting signs and symptoms. A comprehensive laboratory investigation will aid in the specific diagnosis. The histopathological, immunohistochemical and serological examination is very crucial in establishing the diagnosis.

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Author biography

Banushree C S, Professor & HOD  <https://orcid.org/0000-0003-4637-8837>

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