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UNUSUAL PRESENTATION OF PLEOMORPHIC MANTLE CELL LYMPHOMA AND COMPARATIVE STUDY OF CLINICAL FEATURES WITH PREVIOUSLY PUBLISHED CASES

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BACKGROUND

- Pleomorphic Mantle Cell Lymphoma (PMCL) is a rare and aggressive variant of Mantle Cell Lymphoma(MCL). It has distinct cytomorphological features consisting of large irregular lymphoid cells with often bizarre nuclear shapes compared to classical MCL[1].
- It is often difficult to diagnose due to its rarity and varied presentation which warrants a high index of suspicion. There is also a lack of established guidelines for the diagnosis and treatment of this variant which calls for further clinical studies[1].

OBJECTIVE

- This study aims to compile and analyse the baseline characteristics, presentations, and outcomes of PMCL as documented in published case reports. It also includes a case report with imaging findings of a rare presentation of PMCL encountered in clinical practice.

METHOD

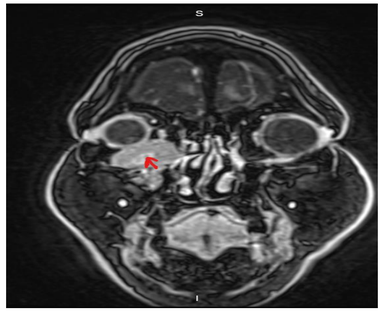
- A systematic search using the keywords “case report” and “ pleomorphic mantle cell lymphoma” was conducted on the following databases**: PubMed, PubMed Central, Google Scholar, Biomed Central, DOAJ, and MEDLINE**. Inclusion criteria encompassed meticulously documented cases showcasing confirmed diagnoses of PMCL.
- Electronic Medical Record was assessed for our single case report included in this study.
- A total of 17 cases were analysed in this comparative study.

CASE REPORT

- A 64-year-old female presented with worsening nasal congestion that had persisted for 5 months despite outpatient treatment for sinusitis. She also experienced right facial numbness and dysphagia .

CASE REPORT CONTD.

- Imaging revealed several enhancing masses in the right maxillary, ethmoid, and sphenoid sinuses as well as the right extraconal space and left masticator space. Additionally, there was an enlarged left parotid lymph node with evidence of perineural invasion through the pterygopalatine fossa and Meckel’s caves. (Lesion shown with red arrow)

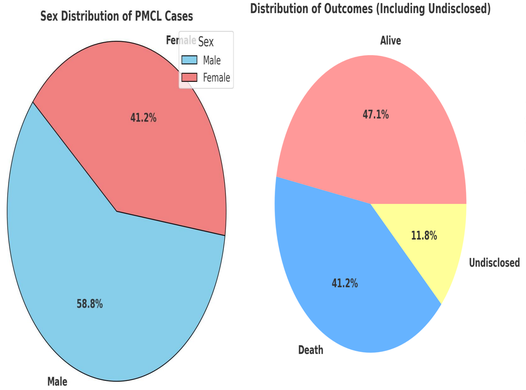


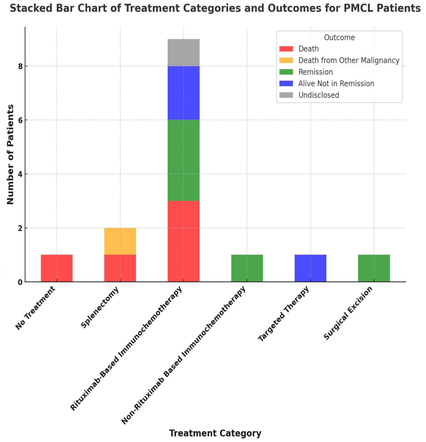
- Immunohistochemistry of the sinonasal mass was deemed most consistent with PMCL based on the studies below:

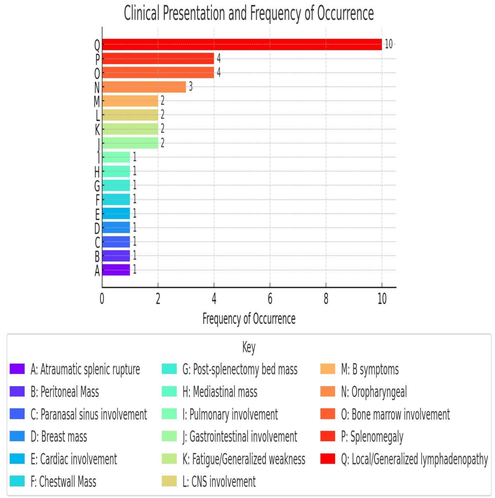
Positive	Negative	Partial positive
CD 5, CD 19, CD 20	CD 10, CD 23, CD 30, CD 56	SOX 11
FMC -7	C-MYC	
Kappa light chain	TdT	
BCL -2	BCL-6	
PAX-5	Cyclin D1	
MUM-1	EBER	
High Ki-67 proliferative index (80%)	Double/triple hit lymphoma	
t(11;14)		

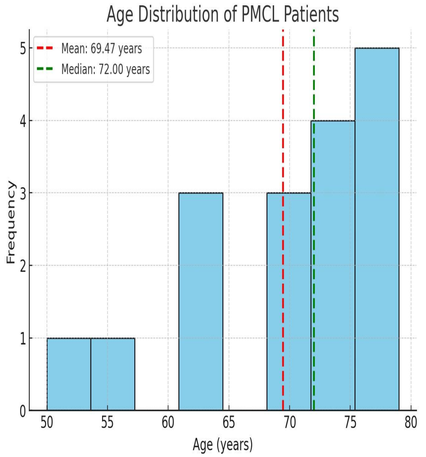
- She received 2 cycles of R-CHOP (Rituximab, Cyclophosphamide , Doxorubicin, Vincristine Prednisone) and appeared to be responding to treatment at the time of this report with improvement in sinonasal lesions

RESULTS









DISCUSSION

- In our analysis of 17 PMCL cases, the mean age at presentation was 69, with a median age of 72. A slight male predominance was observed, with 58.8% (10/17) male and 41.2% (7/17) female. The most common clinical presentations included localized or generalized lymphadenopathy (26.3%), splenomegaly (10.5%), and bone marrow involvement (10.5%).
- Mortality occurred in 41.2% (7/17) of cases, with 6/17 deaths attributed directly to PMCL and 1/17 to a coexisting malignancy.
- Rituximab-based therapies were the most frequently employed treatment regimens, yielding mixed outcomes. Remission was achieved in 4/17 cases: 3/17 after rituximab-based therapy, either alone (1/17) or followed by allogeneic stem cell transplant (1/17) or combined with targeted therapy (1/17). 1/17 achieved remission following surgery alone for localized disease.
- Given the small sample size of this study, larger clinical studies are needed to clarify these findings further,

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