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RESEARCH ARTICLE

PLASMABLASTIC MYELOMA: CASE REPORT

Sami Zriba[1], Karima Kacem[2], Hela Ghédira[1], Raoudha Doghri[3], Ben Hamida Marwa[1], Hela Stambouli[1] and Fehmi M'Sadek[1]

1. Clinical Haematology Department - Military Hospital - Tunis.
2. Clinical Haematology Department - Aziza Othmana Hospital - Tunis.
3. Department of Pathology - Institut Salah Azaiez - Tunis.

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Abstract

Plasmablastic myeloma is an atypical variant of multiple myeloma with a poor prognosis. We report a case of a 59-year-old-male, who was diagnosed to have plasmablastic myeloma. We emphasize the diagnostic difficulty of this type of myeloma and the therapeutic options.

Introduction:-

Case report:

We report a case of a 59-year-old man without significant past medical history. He consulted the 1st time in July 2012 for a poor general condition.

Investigations revealed that he had pancytopenia (hemoglobin: 6 g/dL; MCV: 90 fL; leukocyte count: 1900/cumm; Platelets count: 29,000/cumm), serum protein electrophoresis found a peaking at narrow base in gamma globulin. Serum immunofixation (SIF) showed immunoglobulin G lambda monoclonal immunoglobulin at 36 g/L. Urine immunofixation (UIF) showed monoclonal light chain lambda at 0,1 g/24H.

Bone marrow biopsy showed dense and diffuse plasma cells proliferation (figure 1) which was CD38 and CD138 positive with lambda restriction (figure 2). Some cells showed plasmablastic morphology (large cells with abundant cytoplasm, large nucleus highly nucleoli).

Evolution was marked by improved cytopenias after transfusion of 4 blood units (hemoglobin: 11 g/dL; leukocyte count: 4200/cumm; platelets count: 250,000/cumm).

Cytopenias would be linked to a vitamin B12 deficiency that was prevalent in patients with plasma cell dyscrasias and which was corrected by blood transfusion [1].

The patient was considered to have an asymptomatic multiple myeloma (calcemia: 2,2mmol/L; serum creatinine: 98 µmol/L; hemoglobin: 11 g/dL; skeletal survey did not show any evidence of lytic lesions). The patient was regularly monitored.

In January 2013, appearance of Menelaus and worsening of cytopenias (hemoglobin:7 g/dL; leukocyte count: 3000/cumm, platelets count: 6000/cumm). SIF: IgG lambda at 26 g/L and UIF: lambda at 8 g/24H.

Corresponding Author:-Sami Zriba
Address:-Clinical Haematology Department - Military Hospital - Tunis.

Gastroscopy objectified the presence of congestive polypoid lesions with substance loss and bleeding stigmata (Forrest IIc). These lesions affected the whole stomach and duodenum.

Pathological examination showed large cells with clear plasma cell differentiation morphologically corresponding to plasmablasts (figure 3) CD38 + and IgG lambda restriction (figure 4).

Computed tomography scans revealed mediastinal and left gastric lymph nodes and heterogeneous splenomegaly.

The diagnosis of plasmablastic myeloma (PBM) was retained and the patient developed lethal hemophagocytic syndrome before initiation of chemotherapy.

Discussion:-

Plasmablastic myeloma (PBM) is a morphologic subset of myeloma. It may represent the plasmablastic transformation of a known multiple myeloma or be de novo PBM. The prevalence of plasmablastic morphology in patients with newly diagnosed myeloma is approximately 10% [2].

Under the criteria of Greipp and colleagues, PBM cells are defined as having a large, centrally placed nucleus (>10 μm in diameter) or a nucleolus (>2 μm in diameter) and a high nuclear/cytoplasmic ratio; the cytoplasm is scant and lacks a prominent perinuclearhof. To be classified as PBM, plasmablasts must comprise 2% or more of nucleated cells in the bone marrow (BM) aspirate. By contrast, in the grading scheme of Bartl et al, designation as PBM requires that plasmablasts represent the predominant cell type in the BM biopsy specimen [3].

The main differential diagnosis considered was plasmablastic lymphoma (PBL) [4,5]. Although difficult, it is clinically important and critical to differentiate between these two entities, as treatment for each one is significantly different. The distinction between PBL and PBM frequently depends on the clinical presentation given that morphologic distinction is not always possible and immunophenotypic features showed a virtually identical profile for PBM and PBL (Table 1). Colomo et al found CD56 to be specific for plasma cell myelomas [6]. Peroxiredoxin I, expressed in the terminally differentiated plasma cells, but not in B-lymphocytes, has been identified as specific for plasma cell neoplasms [7].

Plasmablastic morphology conferred a very poor prognosis. For PBM, this independent prognostic impact persists even after autologous stem cell transplantation [3]. Median overall survival of myeloma patients with PB morphology and non-PB morphology was 1.9 versus 3.7 years; whereas, median event free survival of patients with PB morphology and non-PB morphology was 1.1 versus 2.7 years [2].

The outcome of untreated patients with PBL is dismal, with a median OS of 3 months for HIV-positive patients and 4 months for HIV-negative patients [8]. In a retrospective analysis of 53 patients diagnosed with PBL, the five year OS of the overall population was 21.4%.

PBM patients should be considered as having high-risk myeloma particularly forms with extramedullary disease and treated accordingly [9].

The recommendations for first-line treatment of PBL is 6 cycles of dose-adjusted EPOCH (with or withoutBortezomib) with intrathecal prophylaxis with each cycle of EPOCH and consideration of consolidative high-dose chemotherapy followed by ASCT in first remission for appropriate candidates [8].

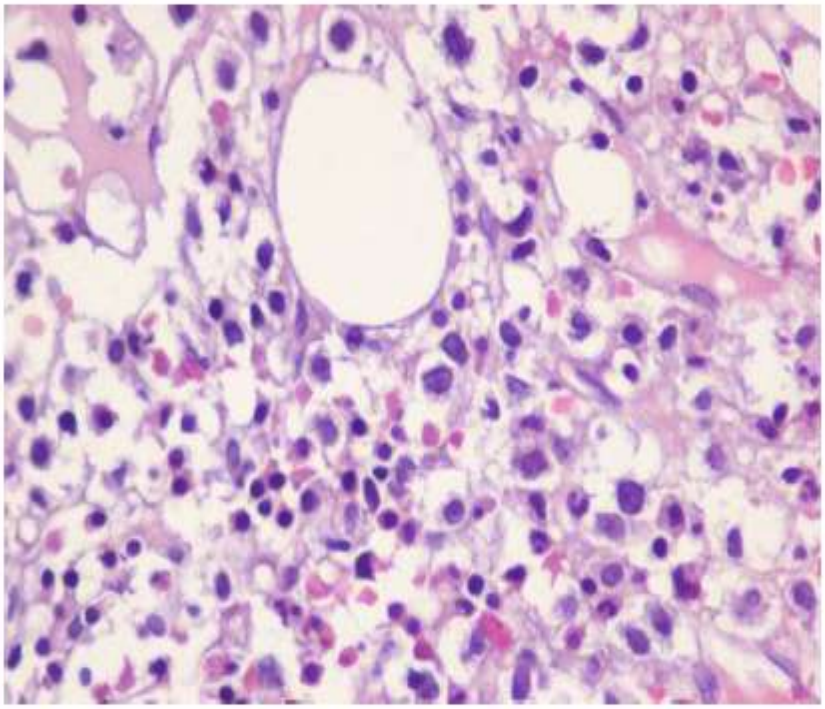

Figure 1:- The bonemarrowwasinfiltrated by large lymphoidcellswithprominentnucleoli and numerous mature plasma cells.

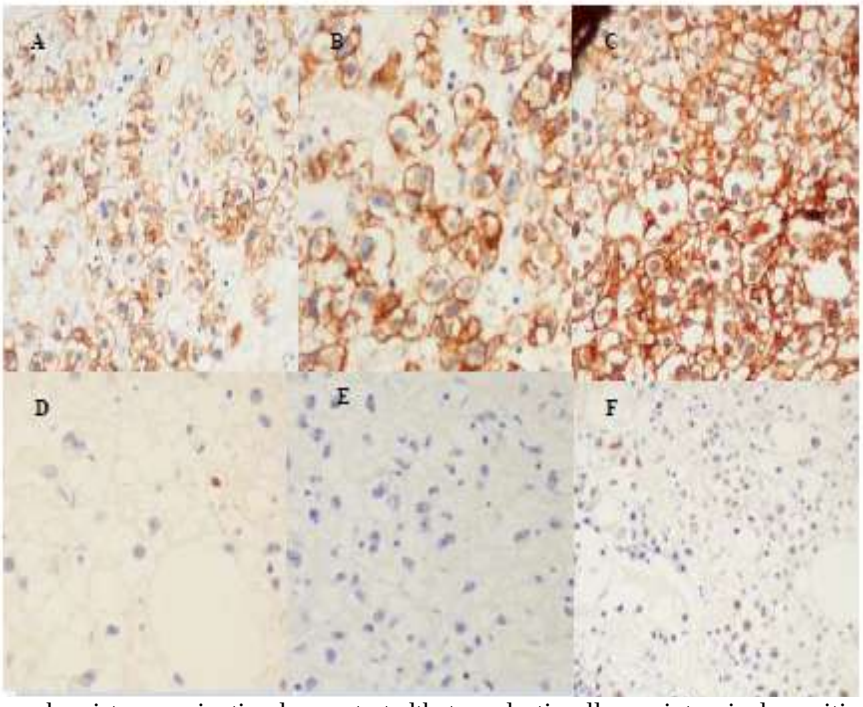

Figure 2:- Immunochemistryexaminationdemonstratedthatneoplasticcellswereintensively positive for CD38 (A), CD138 (B), monotypic light chain λ (C) and negative for CD20 (D) and CD56 (E) (x40). Immunochemistryshowed Ki-67 (F) expressed in the nuclei of 30% of neoplasticcells (x40).

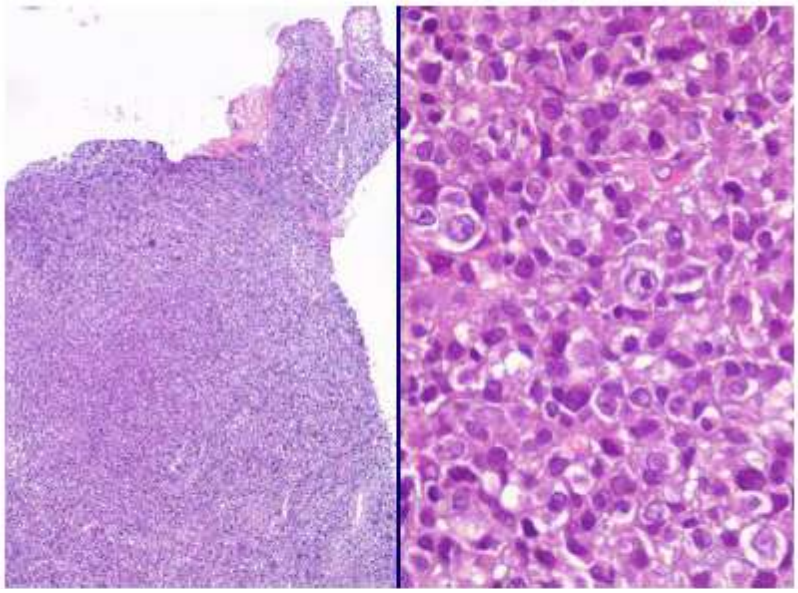

Figure 3:- The gastricmucosawasextensivelyinfiltrated by medium and large lymphoidcellswithprominentnucleoli and amphophiliccytoplasm. The tumorcellsalsohadsomeevidence of plasmacyticdifferentiation (H &E).

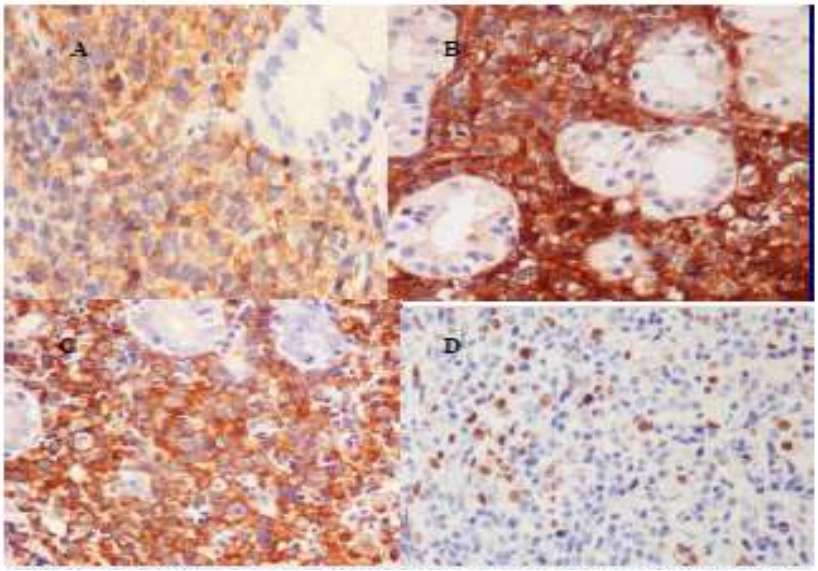

Figure 4:- Immunohistochemicalstains of CD38 (A), IgG (B) and light chain λ (C). Ki-67 (D) expressed in the nuclei of 20% of neoplasticcells (x40).

Table 1:- Comparison of clinical features, morphology and immunophenotype between PBM and PBL.

	PBM	PBL
Location	BM and extranodal	extranodal
Immune status	Competent	Suppressed, HIV
EBV infection	Absent	Present (60-75%)
Serum protein electrophoresis	M-spike (IgG, IgA, IgM, IgD)	No proteinemia
Bence-Jones protein	Present	Absent
Bone lytic lesions	Common, but may be absent	Rare, in widely disseminated disease
Morphology	Plasmablastes and frequent plasmacytic cells	Plasmablastes and/or immunoblasts
Ki-67	High (5-60%)	Very high (>85%)
Plasma cell differentiation (CD38,	Positive	Positive

CD138, MUM1)		
Kappa/Lambda restriction	80-90%	40-50%
CD20	Negative	Negative
CD79a	-/+, 50-85% positive	-/+
CD4, CD10	-/+	-/+
CD56	Usually positive	Usually negative
PAX-5	Negative	-/+ Weak
Peroxiredoxin I	Positive	Negative
BCL-2	-/+	-/+
BCL-6	Negative	Negative
ALK-1	Negative	Negative

References:-

1. Baz R, Alemany C, Green R, Hussein MA. Prevalence of vitamine B12 deficiency in patients with plasma cell dyscrasias. Cancer 2004; 101(4):790-5
2. Shrikrishna PA, Anant RN, Rameshrao AA, Ranjitsingh SK. Bone marrow aspiration of plasmablastic multiple myeloma. J ApplHematol 2013;4:121-2
3. Lorsbach RB, His ED, Dogan A, Fend F. Plasma cell myeloma and related neoplasms. Am J ClinPathol 2011;136:168-82
4. Vega F, Chang CC, Medeiros LJ, Udden MM, Cho-vega JH, Lau CC, Finch CJ, Vilchez RA, McGregor D, Jorgensen JL. Plasmablastic lymphomas and plasmablastic plasma cell myelomas have nearly identical immunophenotypic profiles. Modern Pathology 2005;18:806-15
5. Choi Q, Kim HK. Plasmablastic plasma cell myeloma mimicking plasmablastic lymphoma. K J Hematol 2010;45(4):223
6. Colomo L, Loong F, Rives S, Pittaluga S, Martinez A, Lopez-Guillermo A, Ojanguren J, Romagosa V, Jaffe ES, Campo E. Diffuse large B-cell lymphomas with plasmablastic differentiation represent a heterogeneous groupe of disease entities. Am J SurgPathol 2004;28:736-47.
7. Thakral C, Thomas L, Gajra A, Hutchison RE, Ravizzini GC, Vajpayee N. Plasmablastic lymphoma in an immunocompetent patient. JCO 2009;27:e78-e81
8. Castillo JJ, Bibas M, Miranda RN. The biology and treatment of plasmablastic lymphoma. Blood 2015;125(15):2323-2330
9. Appleby N, Cullen R, Painter D, Smith A, Roman E, Barrans S, O'Connor SJM, Burton CH, Jack AS, Owen RG, Tooze RM. Poor outcomes in HIV negative and HIV positive patients with plasmablastic lymphoma in a population based patient study. Blood 2015;126:3924.