CASE REPORT

SECONDARY PLASMA CELL LEUKAEMIA

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Plasma cell leukaemia is a clinical condition in which plasma cells circulating in the peripheral blood constitute >20% of white blood cells (WBC) and there is evidence of plasma cell monoclonality. It is important to be diagnosed early for better treatment outcome. Although it is a rare disease, cases have been reported from Pakistan and other countries (including our neighbouring countries), hence making this case report. After taking history of present and past ailments, physical examination was carried out. Blood and bone marrow sampling were done after taking informed written consent from the patient. Blood samples were obtained in plain bottle, anticoagulated bottle and bone marrow was obtained from posterior iliac spine under 2% lignocaine. Plasma cell leukaemia is a rare and aggressive disease, difficult to diagnose and treat, requires early recognition and therapeutic intervention.

Keywords: Plasma cell leukaemia; Multiple myeloma; Monoclonal gammopathy; Plasma cell myeloma; Solitary plasmacytoma

Citation: Idris M, Farid J, Javed N. Secondary plasma cell leukaemia. J Ayub Med Coll Abbottabad 2020;32(2):277–9.

INTRODUCTION

Plasma cell leukaemia is a clinical condition in which plasma cells circulating in the peripheral blood constitute as much as > 20% of white blood cells (WBC) and there is evidence of plasma cell monoclonality.[1,2] It is estimated that PCL constitutes 0.3–0.5% of plasma cell disorders. It is of two types, namely primary plasma cell leukaemia (pPCL) characterized by more aggressive course, affecting relatively younger people with more extra-osseous involvement, and the secondary plasma cell leukaemia (sPCL) in which osseous disease already exists and the involvement of extra-osseous sites is less common.[3] Thirty to 40% plasma cell leukaemias are of secondary type.[4] High level of suspicion is required to diagnose plasma cell leukaemia, especially pPCL, while examining and investigating a patient with clinic-haematological features suggestive of some plasma cell disorder

CASE REPORT

A 52 years old hotel waiter presented to oncology clinic at Ayub Teaching Hospital Abbottabad with pallor, increasing low back pain, fever, weight loss, decreased appetite and difficulty in walking. On examination he had pallor, confusion and armpit temperature of $101F^{o.}$ His pulse rate was 110/min, respiration 18 breaths per min and blood pressure 120/70 mmHg. His liver was palpable 3cm and spleen 5 cm below the costal margin. Rest of his physical examination was unremarkable. His complete blood counts revealed Haemoglobin 7.3 g/dl, platelets $75\times10^3/\mu l$, WBC $3.410^3\times10^3/\mu l$ comprising 50% neutrophils, 45% lymphocytes, 3% Monoctes and 2% Eosinophils Giemsa stained blood film examination revealed as much as 25% plasma cells (Figure 1). His erythrocyte sedimentation rate was 115mm at the end of 1st hr WG. His blood urea was 220 mg/dl, serum calcium 12 mg/dl and creatinine 7.3 mg/dl. His liver functions and coagulation profile were not deranged.

Previously he was treated for multiple myeloma, went into remission, continued treatment for 3 months and was advised regular monthly follow-up with serum beta-2 macroglobulin. He missed follow-up visits for four months and then presented with almost the same picture as was on his 1st presentation (Table-1). Keeping in mind his previous history, suspicion of secondary plasma cell leukaemia was raised. Flowcytometry was performed on his peripheral blood which revealed as much as 35% cells positive for CD138, CD56 and CD38, confirming the diagnosis of plasma cell leukaemia. His bone marrow examination revealed 55% plasma-blasts. Radiological survey of his skeleton revealed multiple lytic lesions in his skull (Figure-1).

His serum protein electrophoresis revealed discrete band with minute but sharp spike in the gamma region. His serum calcium was 12mg/dl, urea 156mg/dl and creatinine 2.2 mg/dl. His plasma albumin was 2.5g/dl. He was hospitalized for supportive care and subsequent chemotherapy.

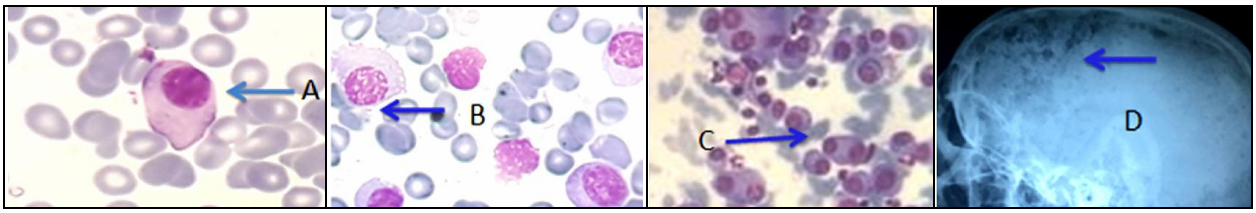

Figure-1: (A, B) Plasma cells in peripheral blood, (C) Plasma cells in Bone marrow, (D) Lytic lesions in skull bone

Table-1: Patient's biochemical, haematological and radiological parameters

Parameter	At 1st presentation	1 month	2 months	3 months	At 2nd presentation (7 months)
Haemoglobin g/dl	8	10	10.5	10	7.3
WBC $\times 10^3/\mu l$	4	5.3	5	4	3.4
Neutrophils %	67	60	62	65	50
Lymphocytes %	28	35	35	29	45
Monocytes %	03	03	02	04	03
Eosinophil%	02	02	01	02	02
Platelets $\times 10^3/\mu l$	79	145	170	190	75
ESR mm 1st hr WG	120	75	70	70	115
Urea mg/dl	197	75	62	60	220
Creatinine mg/dl	9.6	3.5	2.8	2.5	7.3
Sodium mmol/l	129	133	135	134	129
Potassium mmol/l	4.5	4	3.8	4.5	5
LDH IU	650	450	420	430	530
Plasma glucose mg/dl	151	145	135	130	142
Serum Calcium mg/dl	16.4	9.3	10	9.5	12
Serum albumin g/l	2.8	3.2	3.5	3.5	2.6
Serum Kappa mg/l	1244.6	25	Not done	Not done	955
Serum Lambda mg/l	24.9	24	Not done	Not done	15
β_2. Microglobulin mg/l	40	10	11	9	32
Bone marrow Cellularity Erythropoiesis Myelopoiesis Megakaryocytes Plasma-blasts	 45% Depressed Depressed Normal 45%	Not done	Not done	 55% Active Active Normal <5%	 35 Depressed Depressed Decreased 55%
Lytic lesions skull	4	Not done	Not done	Not done	2
Lytic lesions pelvis	3	Not done	Not done	Not done	Nil
Serum paraprotein g/dl	0.2	Not done	Not done	Not done	0.3
Diagnosis	Multiple myeloma				Plasma cell leukaemia

DISCUSSION

Plasma cell leukaemia is rare but very aggressive disease. Tumour burden may be unexpectedly high as is the proliferation index, which may further increase the mortality rate.[4] Novel therapies have improved the overall survival rate of PCL patients from 3 to 5 months to 11 to 13 months.[3] Further improvement in overall survival has been documented when using novel therapies plus Autologous Stem Cell Transplant (ASCT). Cases of PCL have been reported from different countries including Pakistan.[5–10] Our patient is a case of sPCL. Previously he had multiple myeloma and responded to treatment with Vincristine, Adriablastina and Dexamenthasone (VAD). At the time of second presentation as sPCL, he was not taking any treatment and was on follow-up. He failed to come for follow up for four consecutive months and then started having symptoms as mentioned above. The present case is important as, he was a treated case of multiple myeloma presenting at the age of 52 years. This is in accordance with findings of earlier researchers.[10] In the present case, the criteria of >20 % circulating plasma cell was met, but the criteria of $>2\times10^3/\mu l$ circulating plasma cells was not. It has been reported by the other researchers that both of these criteria (>20 % & $>2\times10^3/\mu l$ circulating plasma cells) are difficult to meet in many patients and a revision of the criteria for the diagnosis of PCL is much needed. It is possible that the restrictive nature of these criteria may be one of the reasons why plasma cell leukaemia is under-reported.[11] We agree with this suggestion.

CONCLUSION

Plasma cell leukaemia is a rare entity and needs early recognition for better treatment outcome. Patients diagnosed with multiple myeloma who have taken treatment and have stable disease, must come for regular follow-up.

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Submitted: December 6, 2018	*Revised: --*	*Accepted: January 1, 2020*

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