CORRESPONDENCE

Priapism as the Presenting Manifestation of Multiple Myeloma

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Dear Editor,

Priapism is a surgical emergency. The most common haematological conditions responsible for this condition are sickle cell disease, thalassemia and acute/chronic myeloid leukaemia. Multiple Myeloma (MM) is a plasma cell dyscrasia and presentation with priapism is extremely unusual. The role of plasmapheresis and chemotherapy have a proven benefit however, it is the pace with which the disease is diagnosed and treated which defines the ultimate outcome.

A 44 year old man, nonsmoker with no known comorbidities presented to our emergency services with 3 days history of painful erection and discolouration of phallus. On initial evaluation, patient was restless with excruciating pain. There was swelling at the base of the phallus with blackish discoloration of tip which rapidly progressed to involve entire glans and most of the shaft of the penis (Fig. 1a–c). General examination showed dark bluish-blackish discolouration of distal portion of left index finger and painful gangrenous involvement of right foot (Fig. 2a–c). Doppler study of penis shaft, left hand and right foot was done which didn't show any arterial or venous compromise. He was managed promptly with opioid analgesics and needle aspiration of corpus cavernosa. There was some relieve in priapism, however he continued to have swelling

and gangrene of the involved site. He was catheterised in view of difficulty in micturition. Vasculitis, anti-phospholipid syndrome, leukemias and hyperviscosity related disorders were amongst the differentials which were considered at the prima facie. His complete blood count showed Hb-110 g/L, TLC-42 × 10⁹/L and platelet count-180 × 10⁹/L with rouleaux formation in peripheral blood examination. His biochemical profile were normal except for the albumin: globulin ratio reversal (A: G ratio of 0.2). Considering the abnormal laboratory findings (mild anaemia, rouleaux formation with A: G reversal), possibility of plasma cell dyscrasia with hyperviscosity syndrome was kept. Due to high serum globulin levels, fulminant disease course with impending loss of organ and limbs, his blood tests were sent to confirm the diagnosis and was immediately given plasmapheresis and high dose dexamethasone (40 mg OD × 5 days) to halt the disease progression. Meanwhile, investigations revealed “M protein”-3 g/dL, Serum immunofixation-IgG Lambda monoclonal gammopathy, Bone marrow biopsy—50 % plasma cells, serum free light chain—(Kappa chain-5.5 mg/L, Lambda chain-70.34 mg/L) and LDH-540 U/L (Fig. 3a, b). He was initiated treatment with bortezomib and cyclophosphamide. All these measures arrested the further progression of the disease except for the distal half of penis shaft which underwent auto-amputation (Figs. 1d–f, 2d–f).

MM is a clonal plasma cell disorder of elderly individuals which presents most commonly with nonspecific symptoms such as bone pain and generalised weakness, classically described with the acronym of CRAB. Priapism is a urological emergency which rarely may have underlying haematological disease. Common haematological diseases includes haemoglobinopathies, acute and chronic leukaemias (e.g. CML). MM is a very unusual and rare cause of priapism [1–3]. On PubMed search, we found that

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Fig. 1 Showing sequential images of penile shaft (a–c) with swelling of shaft and blackish discoloration during initial phase (d–f) followed by darkening of colour along with presence of clear cut line of demarcation

only three cases of “Priapism and Multiple myeloma” have been reported so far till date.

Overall, reported thrombo-embolic complications approximately range from 3 to 5 % in MM [1]. Possible explanation of priapism in MM are (1) hyperviscosity related to paraproteinemia (2) Spontaneous platelet aggregation (3) or venous thrombosis secondary to drug effect eg. thalidomide (4) High fibrinogen and VIII levels (5) associated amyloid deposits [1, 2]. In our case, hyperviscosity and spontaneous platelet aggregation might have played some role in causing priapism. Possibly, there might be more than one of these factors interplaying together for

development of priapism in an individual. Thereby leading to large variation in time interval from priapism occurrence to MM detection (17 years in the case reported by Rosenbaum et al. versus 5 days in the index case) [3–5]. Amongst the plasma cell dyscrasias, Waldenstrom’s macroglobulinemia is known for hyperviscosity related symptoms owing to the presence of macroglobulin (IgM type) with increased chances of symptoms when serum monoclonal protein concentration is more than >70 g/l [6, 7]. Contrary to this, all the reported cases of MM associated with priapism till now are of non-IgM subtype with low magnitude para proteinemia (Table 1). This

Table 1 Demographic features of patients of MM presenting with priapism with their outcome

References	Age	Symptoms	Year of diagnosis	Other symptoms	Type	M spike (g/L)	Treatment	Outcome
Rosenbaum et al. [3]	60	Priapism since 1952 (17 years history before diagnosis of MM was made)	1969	C–R–A+B+, no hyperviscosity symptoms	IgG, Kappa	4.3	Plasmapheresis ×7 MP followed by VAC/BCNU	Resolution of priapism. Patient died of progressive disease in 1974
Flanagan et al. [4]	63	Priapism for 3 days	1987	C–R–A–B+, DVT of left leg PTE	IgG, Kappa	4.8	Plasmapheresis	Complete resolution of priapism and venous thrombosis
Bahat et al. [5]	61	Recurrent/refractory priapism for 9 months	2010	C+R+A+B+, no hyperviscosity symptoms	IgA, lambda	1.74	Aspiration/shunt surgery and high dose dexamethasone	NA
Sahu et al. (present case)	44	Priapism for 5 days	2016	C–R–A–B– Gangrene of right foot and left index finger	IgG, Lambda	3	Aspiration, Plasmapheresis ×7. Below knee amputation and CyBorD chemotherapy	Auto amputation of shaft of penis

Fig. 2 Sequential images of (a–c) right foot and left index finger showing gangrene with (d–f) improvement in discolouration following plasmapheresis and chemotherapy

condition is a combination of both medical plus surgical emergency which requires individualised treatment including aspiration, shunting surgeries, combination chemotherapy, high dose dexamethasone therapies etc.

The case emphasises on the need of extensive and prompt evaluation of any patient presenting with priapism and not showing rapid and complete response to surgical measures. It also highlights the efficacy of presently

Fig. 3 **a** Peripheral blood film shows Rouleaux formation along with an occasional circulating plasma cell (May Grunwald Giemsa, $\times 20$). **b** Bone marrow aspirate shows numerous mature plasma cells (May Grunwald Giemsa, $\times 40$)

available measures like plasmapheresis and chemotherapy to treat MM and related complications. This case aims to set the stage to look for other unknown associated factors in MM which precipitates such catastrophic complications.

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