Case Reports

ISCHAEMIC PRIAPISM AS A PRESENTATION OF CHRONIC MYELOID LEUKAEMIA


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Summary.- OBJECTIVE: To describe ischemic priapism as an atypical presentation of chronic myeloid leukaemia.

METHODS / RESULTS: We discuss two patients diagnosed with chronic myeloid leukaemia presenting an episode of priapism, adequately resolved after applying the treatment protocol established in our center.

CONCLUSION: Priapism is defined as a persistent erection that persists despite not having sexual stimulus, without involvement of the spongy tissue of the penis.

INTRODUCTION

Priapism is defined as a persistent and usually painfull erection which is maintained despite a lack of sexual stimulation and without the participation of the spongy tissue of the penis. It is a urological emergency that must be treated in order to avoid future complications such as erectile dysfunction (1).

This situation is associated with a wide variety of factors, amongst them drugs and medication, infectious diseases, immune or metabolic disorders, blood diseases and infiltrative tumour growth, etc (1,2).

According to its etiopathogenesis, it is classified as follows:

1. Ischaemic or veno-occlusive priapism: the most common form of presentation, associated with multiple

Keywords: Ischemic priapism. Chronic myeloid leukaemia. Erectile dysfunction.

Resumen.- OBJETIVO: Describir el priapismo isquémico como forma de presentación atípica de la leucemia mieloide crónica.

MÉTODO/RESULTADO: Comentamos 2 pacientes diagnosticados de leucemia mieloide crónica a partir de un episodio de priapismo, resueltos adecuadamente tras la aplicación del protocolo de tratamiento establecido en nuestro centro.

CONCLUSIÓN: El priapismo se define como una erección persistente, que se mantiene a pesar de no haber estímulo sexual, sin participación del tejido esponjoso del pene.

Su aparición como debut de una discrasia hematológica es un suceso poco frecuente, constituyendo una urgencia urológica que requiere un manejo precoz y multidisciplinar por parte de los servicios de Urología y Hematología, ya que la rapidez en el tratamiento derivará en una función eréctil conservada, y con ello preservar la calidad de vida.

Palabras clave: Priapismo isquémico. Leucemia mieloide crónica. Disfunción eréctil.

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Its debut appearance as a hematologic dyscrasia is a rare event. It is a urological emergency, requiring early multidisciplinary (Urology and Hematology) management, since the speed in treatment will result in good functional results and the preservation of a good quality of life.
triggering factors such as medication, blood diseases, toxins, parenteral nutrition and neoplasia. In a doppler ultrasound scan (2), it presents as a reduction or absence of cavernous blood flow. It is characterised by a full, painful erection and behaves like a compartment syndrome.

2. Non-ischaemic or arterial priapism: its triggering factor is pelvic trauma or intracavernous laceration. It presents as a partial, non-painful erection. In the Doppler ultrasound scan an increase of cavernous blood flow is observed.

3. Recurrent or stuttering priapism: variant of ischemic recurrent painful erections presenting alternating with periods of sagging

This disease may present as an onset form of certain blood dyscrasias, affecting up to 5% of all leukaemia patients (3,4).

**CASE 1**

The first of our cases deals with a 24-year-old male with no relevant history of disease, who came to A&E complaining of an erection lasting 14 hours, not provoked by sexual stimulation. Upon examination of the patient's medical history, the patient notes that it is the second such episode. The first event occurred approximately 4 months before, it lasted 2 hours and subsided spontaneously. Examination showed a full and painful erection, without compromise of the glans. A blood gas analysis is carried out on corpora cavernosa, with the following results: pH of 6.42, PCO2 of 148 mmHg, PO2 of 43 mmHg and HCO3 of 8.9 mmol/L. As such, the patient is diagnosed with ischaemic priapism.

Following this diagnosis, the management protocol established in our unit, based on existing literature, is initiated:

1. Dorsal penile nerve block with dilution of 2% lidocaine
2. Aspiration of corpora cavernosa
3. Irrigation with cold normal saline
4. Intracavernous injection of 1mg/ml dilution of phenylephrine, injecting 0.2 ml every 5 minutes, up to a total dose of 1.6 ml.

This patient's erection subsided with a dose of 1.2 ml of the dilution.

Subsequently, the results of the blood test requested upon admission to A&E are received. They yielded the following results: leukocytes 177.15x 10^9 /L (normal: 5-10x10^9/L); neutrophils 86.4%(40-65%); lymphocytes 3.5%(20-40%); basophils 6.4%(0-1%); monocytes and eosinophils normal(4-8%, 1-3%); haemoglobin: 103 g/L(14-18g/L). In view of these findings, the Haematology Service is contacted and it completes the study of the case through bone marrow puncture with cytogenetic diagnosis, obtaining 100% of p210 rearrangement, compatible with chronic myeloid leukaemia in the chronic phase, and indicating treatment with imatinib, currently being monitored by the haematology department.

**CASE 2**

The second of our cases deals with a 29-year-old patient with no relevant history of disease, who came to A&E with a second episode of an erection initially provoked by sexual stimulation which lasted 6 hours. The first of these episodes occurred less than a month before, and lasted 4 hours, subsiding spontaneously. Though clinical examination we discovered a full and very painful erection not involving the glans. We carried out blood gas analysis on corpora cavernosa, with the following result: pH of 6.7; PCO2 132 mmHg; PO2 38 mmHg and HCO3 9.1 mmol/L, resulting in the diagnosis of venous priapism. We carried out the same management as in the previous case, this time requiring a total dose of a 1 mg/ml dilution of 1.6ml of phenylephrine. The erection then subsided.

From the blood test results, the following values were obtained: leukocytes 402.24x 10^9/L; neutrophils 77.4%; lymphocytes 3.7%; monocytes 2.2%; basophils 12%; eosinophils normal; haemoglobin of 82g/L.

We contacted the Haematology Service, which completed the study with bone marrow puncture aspiration, observing amplification compatible with the presence of bcr-abl p210 [t (9; 22)] rearrangement, with diagnosis of chronic myeloid leukaemia in the full blown phase of the illness.

Treatment with hidroxyurea was started.

**DISCUSSION**

This disease represents a true urological emergency, manifesting as a persistent erection, which is not necessarily related to sexual activity and is full, painful and without compromise of the corpus spongiosum and behaves like a compartment syndrome. It also poses a risk to erectile function, owing to the resulting ischaemic damage, fibrosis and impotence. This likelihood increases in accordance with the duration of each episode and with delay of the treatment.

Three different mechanisms have been described through which haematologic dyscrasia (leukaemia) may produce said phase:

1. Increase of venous congestion of corpora cavernosa owing to the elevation of intra-abdominal pressure by organomegaly.
2. Agglutination of leukaemia cells in the corpora cavernosa and dorsal vein of the penis
3. Infiltration of the sacral nerve leukaemia cells (5).

Of the total blood dyscrasias, 50% of priapisms observed are associated with Chronic Myeloid Leukaemia, presenting a bimodal age distribution, from 5-10 years old and from 20-50 year old (4).

As regards diagnosis, it is necessary to carry out a comprehensive investigation which attempts to shed light on possible precipitating causes of the episode such as use of medication (intracavernous agents, antihypertensive drugs, anticoagulants, hormones, parenteral nutrition, illegal drugs, etc.), infectious diseases (malaria, syphilis, amongst others), immune or metabolic disorders (diabetes, dyslipidaemia, gout, amyloidosis, lupus, deficiency of vitamin C, etc.), blood diseases (polycythaemia, thalassaemia, leukaemia, myeloma), infiltrative tumour growth (1,3,4,6), highlighting the duration of the episode, previous events, baseline erectile function, trauma, drugs consumption or already known illnesses amongst others.

In the physical examination, the discovery of organomegaly or lymphadenopathy may assist diagnosis.

With regard to supplementary tests, it is important to highlight the importance of a complete haemogram, blood gas analysis of corpora cavernosa (the blood obtained being thick, dark, with a partial pressure of oxygen of under 30 mmHg, partial pressure of carbon dioxide of over 60 mmHg and a pH of under 7.25 in the case of ischaemic priapism), toxicology examination (carbon monoxide, alcohol, cocaine, marihuana, scorpion venom, etc.). Furthermore, a PSA test should be carried out to screen for prostate cancer in men over 40 years (1,3,5) (Table I).

The doppler ultrasound scan (2) is useful in the diagnostic process for assessing intracavernous blood flow to differentiate between ischaemic and non-ischaemic priapism.

As regards treatment, it is essential to intervene quickly, with step and multidisciplinary management of the same being necessary.

Initially the approach is conservative, consisting of ensuring the proper hydration of the patient and analgesia through dorsal penile nerve block, sometimes requiring sedation.

Subsequently the aspiration and irrigation of corpora cavernosa is carried out, as well as the application of sympathomimetic drugs, phenylephrine being the most used, for its minimal stimulation of β1 receptors (1,7).

If this were not sufficient, surgical treatment would be required through the creation of caverno-saphenous shunts, with preference for the distal shunts for their lower rate of complication (1,4,5) or ligatures of arteriovenous malformations. As deferred treatment, it is necessary to highlight that a penile prosthesis is also integrated into the latter as a solution to likely secondary erectile dysfunction.

As a systemic multidisciplinary approach, it should be highlighted that chemotherapy directed at the cause of the lesion which is based fundamentally on the use of hydroxycarbamide or tyrosine-kinase inhibitor (8), amongst others, can begin to be administered from the time of diagnosis (4).

**CONCLUSION**

Ischaemic priapism, although a common complication in blood diseases, rarely appears from the outset.

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**Table I. Description of symptoms and findings presents in the non-ischemic and ischemic form of priapism.**

<table>
<thead>
<tr>
<th></th>
<th>Ischemic priapism</th>
<th>Non-ischemic</th>
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</thead>
<tbody>
<tr>
<td><strong>Type of erection</strong></td>
<td>Complete</td>
<td>Incomplete</td>
</tr>
<tr>
<td><strong>Presence of pain</strong></td>
<td>Yes</td>
<td>Rarely</td>
</tr>
<tr>
<td><strong>Altered venous blood gas</strong></td>
<td>Yes</td>
<td>Rarely</td>
</tr>
<tr>
<td><strong>Traumatic history</strong></td>
<td>Rarely</td>
<td>Frequently</td>
</tr>
<tr>
<td><strong>Pharmacological history</strong></td>
<td>Frequently</td>
<td>Sometimes</td>
</tr>
<tr>
<td><strong>Hematologic disorders</strong></td>
<td>Sometimes</td>
<td>Rarely</td>
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The case of chronic myeloid leukaemia requires multidisciplinary treatment, which implies urgent step urological assessment and treatment, since the speed of diagnosis and the beginning of treatment will determine long term the prognosis of erectile function, as well as the subsequent management of the underlying pathology by the Haematology Service.

REFERENCES AND RECOMMENDED READINGS
(*of special interest, **of outstanding interest)


CHORIOCARCINOMA SYNDROME

Ana Medina, Manuel Ramos, Margarita Amenedo and Lorena París.


Summary.- OBJECTIVE: To explore the possibility of choriocarcinoma syndrome developing as a potentially fatal complication in patients with this pathology.

METHOD: Choriocarcinoma syndrome consists of hemorrhagic manifestations of metastases in advanced germ cell cancer containing large elements of choriocarcinoma. It should be suspected in patients with high tumor mass, multiple metastases and elevated tumor markers characteristic of germ cell tumors. It usually occurs before and during the onset of systemic treatment with chemotherapy. Failure to diagnose it can lead to fatal consequences and may