TESTICULAR PLASMACYTOMA AS PRESENTATION OF MULTIPLE MYELOMA: CASE REPORT AND REVIEW OF THE LITERATURE

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Summary.- OBJECTIVE: We present the case of a patient with testicular plasmacytoma as initial presentation of multiple myeloma, and we carry out a literature review of this uncommon pathology.

METHODS: 63 year-old male who consulted for a testicular mass for three months. After clinical and diagnostic studies he underwent radical orchiectomy.

RESULTS: Pathologic study of the specimen revealed the presence of round cells, some with plasmocytic aspect. Immunohistochemical studies gave the final diagnosis of plasmacytoma. Studies on disease extension showed rounded lytic lesions spread over the vault of the skull bones. Bone marrow studies, as well as bone biopsy showed infiltration by plasma cell neoplasia in more than 90%, consistent with the diagnosis of multiple myeloma.

The patient received treatment, developing disease progression and subsequently died from the disease.

CONCLUSIONS: Solitary plasmacytoma represents only 6% of all plasma cell neoplasms. Testicular presentation is an unusual event, representing 2% of cases. Although this is usually an autopsy finding, it may constitute the first manifestation of multiple myeloma or exceptionally be the unique location of a plasma cell neoplasm. To date there are few reports published in the literature. This case constitutes a contribution for the knowledge of testicular plasmacytoma.

Keywords: Solitary plasmacytoma. Testicular plasmacytoma. Plasmatic cell neoplasia. Immunohistochemistry. Treatment.

Resumen.- OBJETIVO: Presentamos el caso de un paciente con diagnóstico de plasmocitoma testicular como presentación inicial de mieloma múltiple y realizamos la revisión de la literatura en relación a lo infrecuente de dicha patología.

MÉTODOS: Paciente de sexo masculino de 63 años por presentador de un tumor testicular, de 3 meses de evolución, de crecimiento progresivo. Una vez evaluado clínicamente y con estudios de ayuda diagnóstica fue sometido a orquitectomía radical.

RESULTADO: La anatomía patológica reveló la presencia de células redondas, algunas de aspecto plasmocitico; el estudio inmunohistoquímico concluyó que era Plasmocitoma. Estudios de extensión de enfermedad mostraron lesiones líticas redondeadas diseminadas en los huesos de la bóveda del cráneo. Los estudios de médula ósea, así como la biopsia de hueso demostraron infiltración por neoplasia de células plasmáticas en más del 90%, consistente con el diagnóstico de Mieloma Múltiple. Paciente recibe el tratamiento respectivo, desarrollando progresión de enfermedad y posteriormente fallece.

CONCLUSIONES: El plasmocitoma solitario es una lesión poco frecuente que representa sólo el 6% de todas las neoplasias de células plasmáticas. El compromiso testicular por esta enfermedad constituye un evento inusual, estimado en el 2% de los casos. Aunque éste es generalmente un hallazgo de autopsia, carente de expresión clínica, en raras ocasiones, como en el presente caso, puede constituir la primera manifestación de un mieloma múltiple o excepcionalmente ser la única localización de una neoplasia de células plasmáticas. Hasta la fecha son pocos los reportes publicados en la literatura y éste constituye un aporte más al conocimiento del mismo.
INTRODUCTION

Plasmatic cell neoplasms are divided in three groups: multiple myeloma, solitary extramedullary or osseous plasmacytoma and the leucemic form. Multiple myeloma is the most frequent form, representing 15% of all hematologic malignancies (1). Extramedullary plasmacytomas represents 6% of plasmatic cell neoplasms, and could compromise a great variety of anatomic sites, of which the respiratory tract, gastrointestinal, linfoid tissue, soft tissue and rarely the gonads are the most representative sites (2-5). Exceptionally the testicle is affected by a plasmatic cell neoplasia (6-15).

We present a case of a patients with primary testicular plasmacytoma with a literature review of this unfrequent pathology.

CASE REPORT

A 63 years old male with no relevant prior history presents with a three months history of asymptomatic progressive volume increase of the right testicle. On physical exam: right testicle 8 x 6 cm, increased consistency surrounded by fluid; left testicle normal. Rest of the physical exam was normal.

Ultrasound reports increase of the right testicular size, diffuse hypoecogenic lesion of the testicular parenchyma, absence of cystic or solid injury and the presence of paratesticular fluid collection.

A diagnosis of orchoepididimitis was suspected receiving treatment with antibiotics and antiinflammatories for four weeks, without improvement. We decided to perform surgery. Tumor markers (βHCG and αFP), blood and chest x-ray tests were normal. A right radical orchiectomy was done. CT scan of the abdomen found no significant findings. Study of the specimen describes a testicle measuring 8.5 x 6.5 x 5.5 cm and weighing 230 g, spermatic cord of 7.5 x 1.0 cm. On section of the specimen the testicular parenchyma is completely replaced by a solid brown whitish tumor formation, firm consistency, with thickened tunica albuginea. The epididymis and the spermatic cord are free of tumor (Figure 1).

Microscopically a lesion with irregular borders constituted by round cells, some with plasmocitic aspect and eccentric nucleus with “car wheel” shape and basophilic cytoplasm, others binucleated, with nuclear atypia and mitosis (CAP 1-5/10). The tumor involves the vascular tunica and albuginea, does not affect epididymis and spermatic cord. Vascular or perineural invasion was not seen. The surgical margin’s are free of neoplasia (Figure 2).

With this histomorphological findings, Immunohistochemistry tests were performed to define the diagnosis, which was testicular Plasmacytoma. The results and images are presented in Figure 3 and table I, respectively.

Tumor extension studies were evaluated. Bone scan revealed litic lesions spread in the vault of the skull bones (Figure 4).

Bone marrow aspiration, as well as a bone biopsy showed infiltration by more than 90% with plasmatic cells, consistent with the diagnosis of multiple myeloma. In the immunofixation we can observe monoclonal band corresponding to immunoglobulin D of lambda light chain (Figure 5).

The patient begins treatment with Thalidomide and dexamethasone, for five months, with apparent good clinical response, until the onset of pain in the left thigh and increase of IgD, New studies where
performed shows progressive of disease. He received two more treatment schemes of chemotherapy without response, developing widespread pain, cauda equine compression, deep venous thrombosis of left lower limb, renal insufficiency with metabolic disorder and multifactorial encephalopathy. On re-evaluation of the disease it was found massive involvement of all systems, metastasis in the abdominal cavity, chest wall, soft tissue and central nervous system (Figure 6). Due to the symptoms described the patient died with multiple organic failure.

Figure 2A – 2B. Hematoxyline-eosin staining: atypical plasmatic cells.

Figure 3. Immunohistochemistry – microscopy: note positive CD38.
DISCUSSION

Plasma cells neoplasia are divided into three groups: multiple myeloma, solitary plasmacytoma, bone or extramedullary, and the leukemic form; multiple myeloma is the most common form, corresponding 15% of all malignant hematological neoplasias that requires criteria diagnostic (1,16) (Table II).

Extramedullary plasmacytoma represents 6% of plasma cells neoplasm’s (1) and can involve a variety of anatomical sites; among them the respiratory and the gastrointestinal tract, lymphoid tissue, soft tissue and very rarely the gonads (2,5).

The testicle is a body that exceptionally is affected by plasmatic cells neoplasia (6-15). From the first description for Ulrich in 1939, to date there are few cases of testicular plasmacytoma reported in the literature (6,7) and their occurrence as solitary injury without systemic disease, is even more unusual (8). In a recent literature review, 71 cases of plasma cells neoplasms with testicular involvement has been identified (9). Levin and Mostofi found 7 cases of myeloma in a study of 6000 testicular and paratesticular tumors which only one was apparently primary (10). However, in spite of this partnership, only 2% of patients with myeloma have testicular involvement which is generally a finding at autopsy (7,11). Patients age ranges between 26 and 83 years, with an average of 55 years, being younger in the case of solitary testicular plasmacytoma (8,12). Clinically occur as a painless, testicular mass of progressive growth (2,7,9).

The concomitant presence of hydrocele, as in our report, has only been described in three publications, in two of which the liquid was aspirated and a cytologic diagnosis was done preoperatively (13).

Since most cases occur in the presence of systemic disease, it is required a complete workup study to discard multiple myeloma or other plasmacytomas. This was the conduct followed in our patient, where diagnostic criteria for multiple myeloma was found (7,12,14,15).

It has been accepted that the follow-up time necessary to determine the primary nature of testicular plasmacytoma is one year post-orchiectomy in at time should not exist bone lesions or immunoglobulins abnormalities; however for other authors, this time period is very short, where subclinical disease may not be excluded (7,14,15).

Our macroscopic and microscopic findings of the specimen are similar with other publications. It is characterized by a tumor that almost completely replaces the testicular parenchyma, with gray-whitish color and firm consistency. Cytological features vary in

Figure 4. Bone scan: round litical lesions on skull.

Figure 5. Immune fixation: monoclonal band corresponding to IgD, lambda chain.
relation to the degree of differentiation of plasma cells. They are usually larger than its normal counterparts with abundant cytoplasm and eccentric nucleus, some with the typical “car wheel” shape; those less differentiated can be binucleated or multinucleated, with bizarre atypical nucleus and a variable mitotic index (1,7,9).

Problems with differential diagnosis due to the rarity of its locations and its histomorphology are common, and should be with lymphoma and seminoma, specially spermatocitic or anaplastic varieties. Immunohistochemistry study helps us in the differential diagnosis of plasmacytoma, which is positive for CD38 and immunoglobulins, and negative for CD3, CD20, CD30 and PLAP (16,17). Other diseases to be considered in the differential diagnosis are Leydig cell tumor, granulocytic sarcoma and chronic inflammatory diseases with increase of reactive plasma cells (idiopathic granulomatous orchitis and malakoplakia) (11,12).

In 99% of patients with multiple myeloma, electrophoretic analysis reveals an increase in levels of immunoglobulin in blood and urine; the most common are IgG and IgA in 50% and 25% of cases, respectively. The elevation of IgM, IgE and IgD has been described in few publications, being less frequent in the testicular presentation. It is worth noting that in the present case increased IgD was found, a characteristic that makes it even rarer (7,13-15).

The prognosis of testicular plasmacytoma depends on its association or not with multiple myeloma. In solitary testicular lesions, radical orchiectomy is the treatment of choice, not existing yet good results with chemotherapy or radiotherapy (1). In cases of systemic disease almost all patients die from disease progression, as shown in the present report (9,17). The combination of high dose chemotherapy with autologous stem cell transplant shows encouraging results (8).

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Figure 6. Follow-up studies: multiple metastasis in abdominal cavity, thoracic wall, soft tissues, and central nervous system.
CONCLUSIONS

Solitary plasmacytoma represent only 6% of all plasmatic cell neoplasms. Testicular involvement by this disease in unusual, representing only 2% of all cases. Even though it is found incidentally at autopsy without clinical expression, rarely as in the present case, may constitute the first manifestation of a multiple myeloma or exceptionally may be the only location of a plasmatic cell neoplasia. At present, there are few cases reported in the literature, and this case represents a contribution to the knowledge of this disease.

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


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