CASE REPORT

Testicular Plasmacytoma

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ABSTRACT

This is the clinical case of a 75-year-old male patient with medical history of Stage III-A immunoglobulin G kappa multiple myeloma with advanced bone disease, who presented right testicular tumor 1 month ago. Scrotal ultrasound showed an heterogeneous 35 × 22 × 52 mm right testis with decreased echogenicity and increased vascularization corresponding to testicular tumor. Tumor markers were normal and computed tomography scan showed no extension of disease. Pathologist confirmed with immunohistochemical study the definitive diagnosis of testicular plasmacytoma. The patient started the third cycle of treatment with monoclonal antibodies, with good initial response.

Key words: Immunohistochemistry, multiple myeloma, orchiectomy, plasmacytoma

CASE REPORT

We present a 75-year-old male patient with the following medical history: Bilateral inguinal herniorrhaphy, hypertension, gastroesophageal reflux, and Stage III-A immunoglobulin G (IgG) kappa multiple myeloma with advanced bone disease. He attended the urology hospital consultation by request from Primary Healthcare Unit due to an increase in the size of the right testicle for 1 month.

Physical examination revealed right scrotum with double size of the left, hard consistency testis with no pain on palpation, and normal left testicle. The patient had not had fever or previous trauma and neither had associated voiding symptoms.

Testicular ultrasound reported the presence of a normal left testicle, heterogeneous 35 × 22 × 52 mm right testis with decreased echogenicity, increased vascularization, no nodular lesions, and more hypoechoic and vascularized solid area that was located in the most caudal part of the testicle and surrounded it laterally, being about 55 mm thick. Both of the epididymis were normal and mild bilateral hydrocele was observed.

With the diagnostic of testicular tumor, blood tests were performed with tumor markers that were within normality (alpha-fetoprotein 0.72 IU/ml and beta-human chorionic gonadotropin 2 IU/l). The thoracoabdominal computed tomography scan showed the absence of extratesticular disease and the presence of multiple bone involvement in relation to his history of multiple myeloma.

With a correct preoperative study, a right inguinal orchiectomy was performed. The patient was discharged after 24 h.

The pathological anatomy study revealed the following findings. The right orchiectomy specimen of 10 × 6 × 4.5 cm length and weighing 131.20 g [Figure 1]. On section, smooth-walled cystic tunica vaginalis was observed and testis was

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replaced in 96% by solid, whitish-pink, and homogeneous tumor, surrounded by tunica albuginea and epididymis without macroscopic changes [Figure 2].

In the microscopic study, proliferation of atypical plasma cells was observed, completely replacing the testicular parenchyma [Figures 3 and 4]. The cells showed eccentric nuclei with prominent nucleoli and abundant cytoplasm [Figure 5]. Sometimes, they were binucleated and multinucleated, with immature appearance. Frequent mitoses appeared [Figure 6].

The immunohistochemical study reported the presence of tumor cells that immunoexpressed CD138, MUM1, ACL, heterogeneous expression of CD79a, CD117, CD56, C-Myc [Figures 7-13], CD10, and focal epithelial membrane antigen. Lack of expression of CD20, PAX5, CD5, CKAE1AE3, Podoplanin, and anti-placental alkaline phosphatase. The cell proliferation index was 18% assessed with Ki67 [Figure 14].

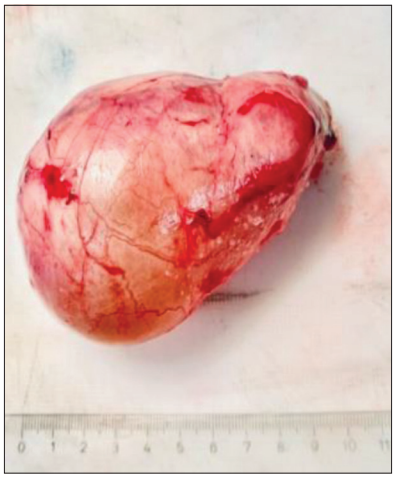

Figure 1: The right orchiectomy specimen of 10 × 6 × 4.5 cm length

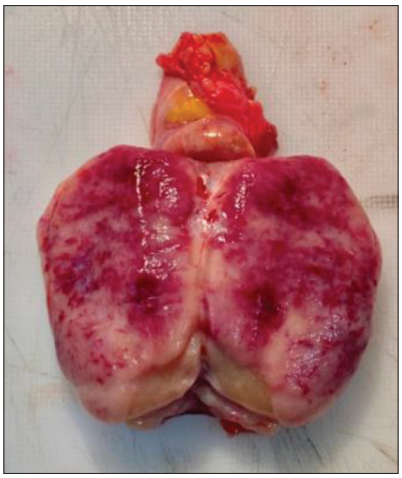

Figure 2: Smooth-walled cystic tunica vaginalis was observed and testis was replaced in 96% by solid, whitish-pink, and homogeneous tumor

The definitive diagnosis was a plasma cell tumor (plasmacytoma) with infiltration of the spermatic cord.

One month after surgery, the patient was visited by hematology, indicating third-line treatment for advanced multiple myeloma with testicular involvement. Two biweekly cycles of daratumumab were prescribed, followed by 4 more cycles monthly. Each cycle of daratumumab was administered intravenously with a 20 ml vial (each vial contains 400 mg of the drug) and requires montelukast 10 mg orally daily 3 days before and dexamethasone 20 mg before and after daratumumab administration. In addition, erythropoietin was prescribed monthly and acetylsalicylic acid 100 mg daily.

The patient is currently awaiting the third dose of daratumumab. He has generalized bone pain that requires analgesic treatment and has had an episode of candidal balanitis that has been treated with clotrimazole topic with good improvement. The analytical response and scrotal physical examination are satisfactory.

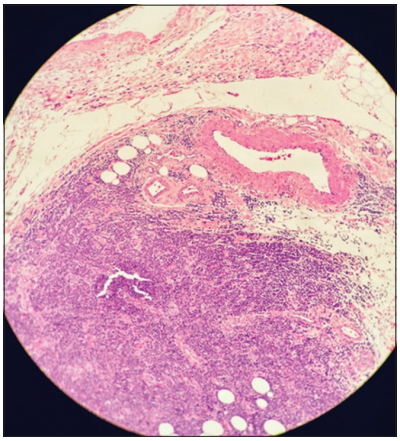

Figure 3: Proliferation of atypical plasma cells (hematoxylin-eosin stain × 10)

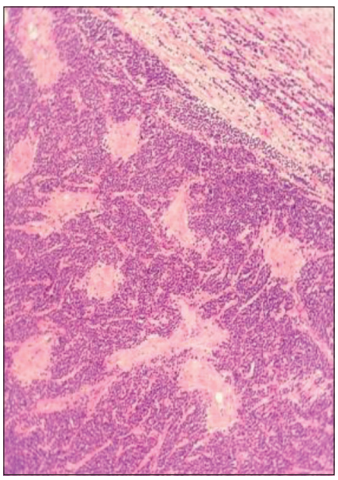

Figure 4: Proliferation of atypical plasma cells (hematoxylin-eosin stain × 20)

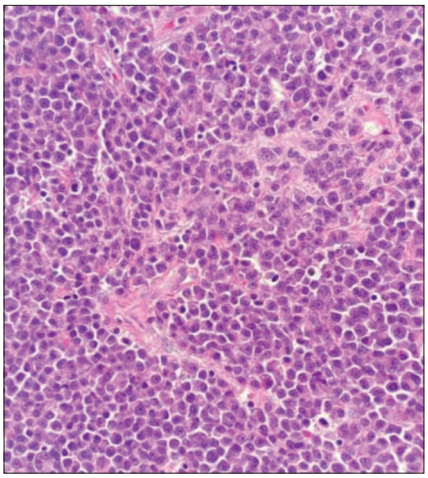

Figure 5: Cells showed eccentric nuclei with prominent nucleoli and abundant cytoplasm (hematoxylin-eosin stain × 40)

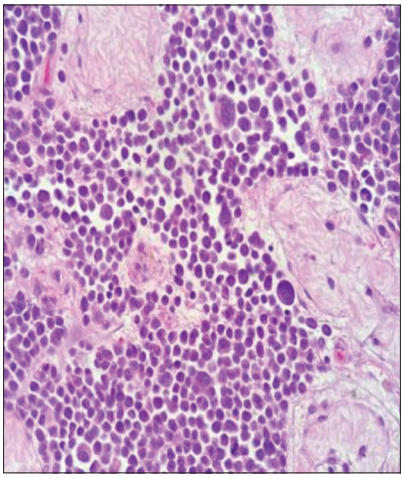

Figure 6: Binucleated and multinucleated cells, with immature appearance (hematoxylin-eosin stain × 40)

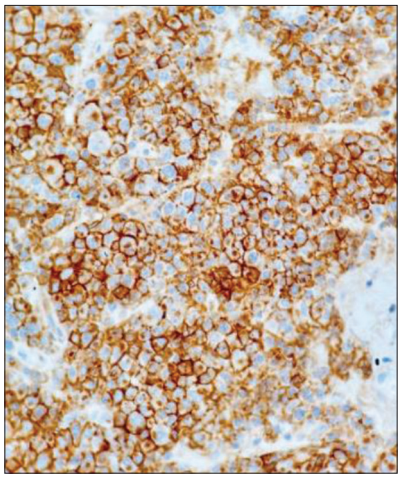

Figure 7: CD138 × 40 +

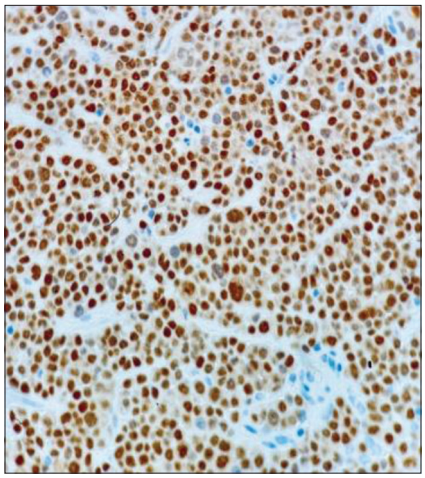

Figure 8: MUM1 × 40 +

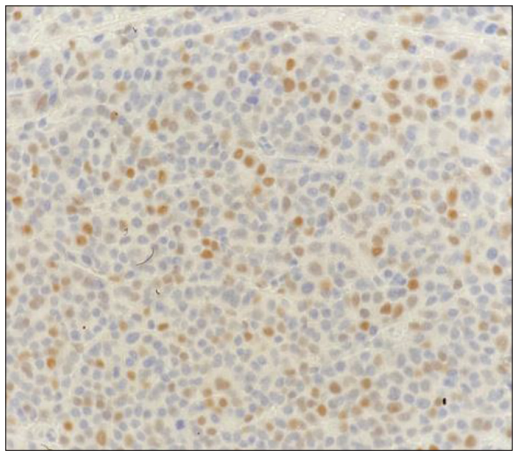

Figure 9: CD79 × 40 +

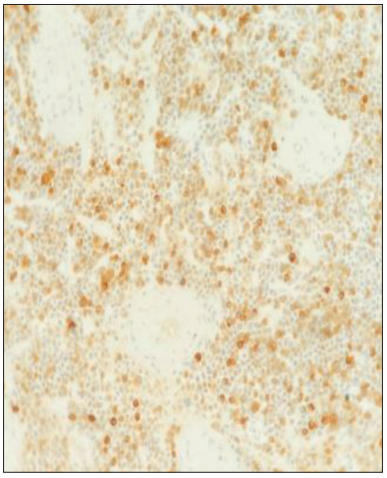

Figure 10: CD117 × 40 +

DISCUSSION

Multiple myeloma is a plasma cell tumor that affects the bone marrow, characterized by the synthesis and excretion of

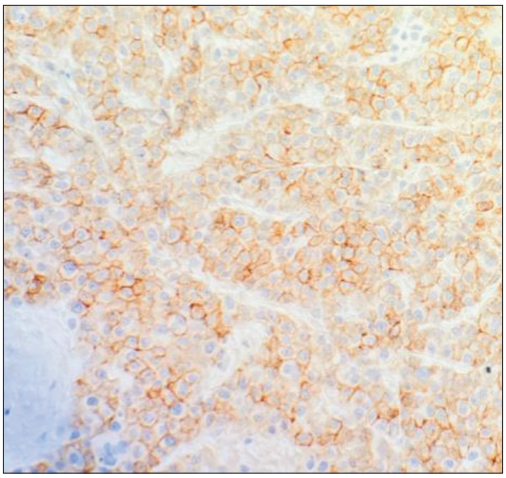

Figure 11: CD46 × 40 +

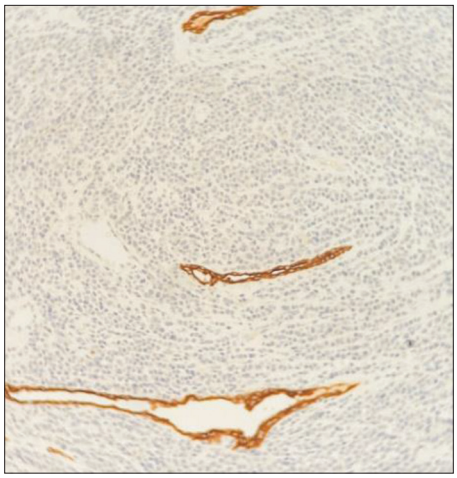

Figure 13: CKAE1AE3 × 20 negative

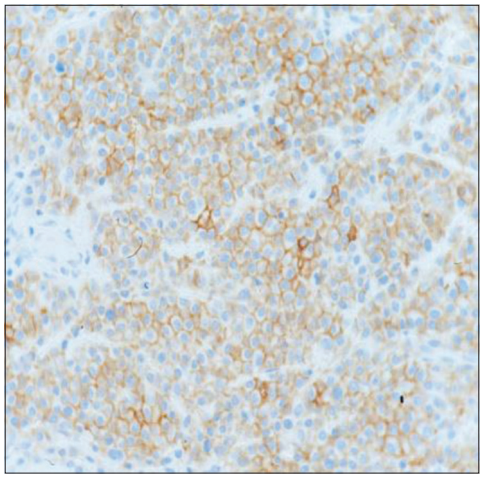

Figure 12: C-Myc × 40 +

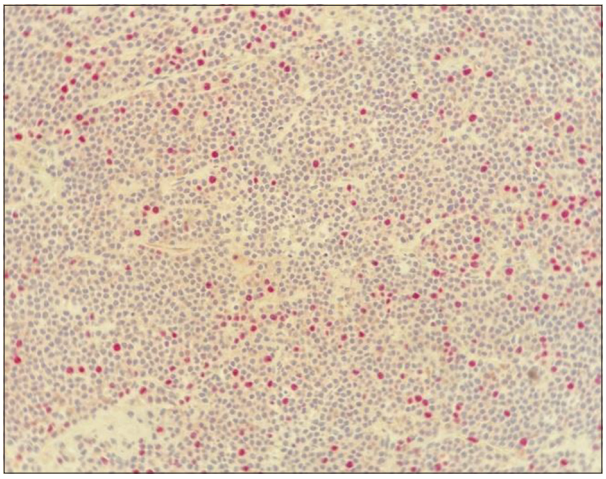

Figure 14: Proliferation index assessed with Ki67 × 20

monoclonal proteins. This tumor originally proliferates in the bone marrow, subsequently invading the adjacent bone with progressive generalized skeletal involvement. Sometimes, it can be located extramedullary.[1]

The most frequent locations of extramedullary plasmacytoma are kidney, submucosal lymphoid tissue of nasopharynx and paranasal sinus, lymph nodes, spleen, lung, adrenal gland, pancreas, pleura, pericardium, skeletal muscle, and testis.[2]

Testicular plasmacytoma has an incidence between 0.03% and 0.1% of all primary and secondary tumors that affect testis, and represents only 2% of all plasma cell neoplasms.[3] It was described for the 1st time in 1939 and cases have been published between 26 and 89 years of age, being more frequent in the 6th decade of life.[4]

Testicular plasmacytoma presents as a painless mass that is similar on physical examination to any other primary or metastatic testicular tumor. It affects the testicular parenchyma, although it can also appear in the epididymis.[5] Histologically, infiltration is demonstrated by the presence of atypical plasma cells. Immunohistochemistry gives us the definitive diagnosis with positivity for CD138, MUM1, ACL, CD79a, CD117, CD10, CD56, and C-Myc. The differential diagnosis must be carried out especially with spermatocytic seminoma and lymphoma.[6]

Plasmacytoma of the testis is treated by radical orchiectomy. When involving advanced stage plasmacytoma or recurrence is present, radiation therapy can be added. In cases involving multiple tumor lesions, systemic chemotherapy or monoclonal antibodies are usually indicated.[7,8]

Daratumumab is a human IgG kappa monoclonal antibody that targets CD38, a cell surface protein that is overexpressed on multiple myeloma cells, and induces direct and indirect antimyeloma activity.[9] It is used from the third line or more of treatment in progressive multiple myeloma, when prior therapy included a proteasome inhibitor and an immunomodulatory drug.

The prognosis of testicular plasmacytoma depends on its association or not with multiple myeloma. In case of the presence of multiple myeloma, most patients show disease progression and have bad prognosis.[2]

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