TESTICULAR HEMANGIOPERICYTOMA, SOLITARY FIBROUS TUMOR: A VERY RARE CASE

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Summary.- OBJECTIVE: To describe the case of a patient with paratesticular hemangiopericytoma and to perform a bibliographic review.

METHODS: We describe the case of a patient with paratesticular hemangiopericytoma. We conducted an exhaustive literature review in different databases to support the case discussion.

RESULTS: We present the case of a 64 year old male with a progressively growing mass in the right scrotum that underwent complete resection of the mass. The result of
Resumen.- OBJETIVO: Describir el caso de un paciente con un hemangiopericitoma paratesticular y realizar una revisión de la literatura.

MÉTODOS: Se describió el caso clínico de un paciente con hemangiopericitoma paratesticular. Se realizó una búsqueda exhaustiva de la literatura en diferentes bases de datos con las que se sustentó la discusión del caso.

RESULTADOS: Se presentó el caso de un varón de 64 años con una masa de crecimiento progresivo en el escroto derecho, quien fue sometido a resección completa de la masa. El resultado de la patología reportó hemangiopericitoma/tumor fibroso solitario y se presenta el manejo instaurado.

CONCLUSIONES: Se presentó un caso poco frecuente de hemangiopericitoma paratesticular. La mayoría de éstos tienen un comportamiento benigno, sin embargo se debe tener en cuenta los criterios de malignidad para tomar decisiones con respecto a su manejo.


INTRODUCTION

Hemangiopericytoma/solitary fibrous tumors are rare neoplasms classified as perivascular tissue sarcomas. They are very uncommon in genitourinary system and when present, they usually localize in kidneys. These tumours are often present for several years before diagnosis as they do not cause symptoms, are slow growing and they are frequently incidentally discovered. Its clinical behavior is uncertain because of the lack of defined histological features. We present the case of a patient with a paratesticular fibrous solitary tumour and a review of literature.

CASE REPORT

A 64 year old man presented to surgery department of our clinic with a scrotal mass progressively growing over 2 months. During initial assessment, a diagnosis of inguinoscrotal hernia was made. The herniorrhapsy was performed and a large paratesticular mass was found, approximately 20 cm in diameter that was biopsied. The pathology report informed an intermediate cell tumour, with hemangiopericytoid features and uncertain clinical behavior.

Pathology report described a mass of 20x15x15 cm in diameter, with irregular shape and 6 mitosis in 10 high power fields (HPF), 10% of the tissue was necrotic and vascular invasion was present; tumor section borders and 8 inguinal nodes resected were negative. Immunohistochemistry was positive for CD34 (predominantly perivascular) (Figure 3), BCL2 and CD99. It was negative for ALK, CD117, AML, calretinin, EMA CK AE1AE3 and inhibin. The proliferation rate Ki67 was 5%; with these results the diagnosis of fibrous solitary tumor was made.

Because of the low proliferation rate and absence of metastasis, we decided not to administrate adjuvant therapy. Three months later patient was asymptomatic, with...
normal wound scarring and without local lesions. Chest X-ray and computed tomography of abdomen and pelvis were negative for metastasis.

DISCUSSION

Hemangiopericytoma (HPC) is a soft tissue neoplasm thought to arise from pericytes; current trend is to classify it as fibrous solitary tumors although World Health Organization (WHO) still classifies it as hemangiopericytoma/solitary fibrous tumors. Principal argument to abandon this term is that clinical, morphologic and immunohistochemical profile lack specificity; that is why current trend is to classify it as fibrous solitary tumor-cellular variant (1).

Patients are usually asymptomatic and the mass is incidentally discovered. The tumor is a mesenchymal neoplasm, probably derived from myofibroblasts (2). HPC primarily affects adults with median age of appearance in the sixth and seventh decade of life. Men and women are similarly affected. Findings that are consistent with the clinical presentation of our patient, who was taken to surgery thinking that the scrotal mass was a inguinoscrotal hernia.

The most commonly affected anatomic sites for HPC include pleura (60%), lower extremities, retroperitoneum/pelvic fossa, head and neck, meningeal, orbital and superior respiratory tract. This tumor has also been described in other locations including lung, breast, peritoneum, liver, stomach, uterus, ovary, vagina and pericardium. In the genitourinary tract, it has been described in seminal vesicles, prostate, spermatic cord and testicle vaginal layer (2-4).

Most of these tumors are well circumscribed and it is possible to find internal hemorrhagic content, cystic mixoid degeneration and necrosis or focal calcification. Classic histologic appearance includes a well developed branching ‘staghorn’ thick-walled vessels surrounded by a connective tissue sheath with moderate-to-high cellularity. These histologic features are nonspecific (2). Immunohistochemistry show positivity to CD34 (positive in 44-95%), CD 99 (positive in 64-91%) and BCL2 (positive in 50%). HPC is usually negative for smooth muscle actin, desmin, citokeratin, S100 protein, and CD3 (1,2,5-7).

In our patient immunohistochemistry was important to establish diagnosis, because the three more common positive markers were confirmed.

A malignant behavior is found in 23% of cases, especially in extrathoracic locations (2,8). There have been described malignant features as size, incomplete resection, high cellularity, high mitotic activity (more than 4 mitosis per HPF), diffuse positivity for p53, high expression of Ki67 and CD31, nuclear pleomorphism and necrosis (2). Our patient showed some factors for malignant behavior that can determine relapse.

Differential diagnosis are: lipomatosus hemangiopericytoma, dendritic fibromixolipoma, big cells angiofibroma, spindle cell lipoma, metastatic carcinoma, desmoplastic mesothelioma and undifferentiated liposarcoma (2,9).

Treatment of choice is complete surgical excision and negative surgical margin (principal determinant of recurrence) (2). Strict follow-up is absolutely necessary and use of radiotherapy and chemotherapy are suggested by some authors although there is no consensus about this issue (2,10).

CONCLUSIONS

We present a rare case of a patient with paratesticular hemangiopericytoma, with difficult clinical and patholo-
gic diagnosis, treated surgically and in satisfactory condition during follow-up. Most of solitary fibrous tumors with hemangiopericytic variant show benign behavior but it is important to consider possible malignant features in order to make treatment decisions, although there is no consensus about this issue because of few cases. Strict and periodical follow-up is a main point.

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