CLINICAL CASE

Solitary fibrous tumor: report on a case with unusual presentation

A.B. Ticona-Garrón a,*, G. Alfaro-Méndez b, J. Farrera-Torija b and E. Medina-Olivera a

a Urology Service, Hospital Regional de Alta Especialidad Ciudad Salud de Chiapas, Chiapas, Mexico
b Pathologic Anatomy Service, Hospital Regional de Alta Especialidad Ciudad Salud de Chiapas, Chiapas, Mexico

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Abstract  Solitary fibrous tumor is uncommon and generally presents at the level of the pleura. The location of this tumor in the seminal vesicle is very rare. It is difficult to distinguish from other tumors through imaging techniques and 90% of solitary fibrous tumors are benign. Diagnosis is made through immunohistochemical staining to distinguish it from other malignant tumors; the CD34 marker is strongly expressed by this tumor. We report herein the case of a 49-year-old man that presented with this tumor, whose site of origin was the right seminal vesicle. Histopathologic and immunohistochemical studies confirmed the diagnosis of solitary fibrous tumor.

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Keywords  Solitary fibrous tumor; Seminal vesicle; Extrapleural

Tumor fibroso solitario, reporte de un caso con presentación inusual

Resumen  El tumor fibroso solitario es un tumor raro, que generalmente se presenta a nivel de pleura. Su localización en vesícula seminal es muy rara. Por técnicas de imagen es difícil diferenciarlo de otros tumores, y el 90% de ellos tienen características benignas. Para su diagnóstico se utiliza la realización de marcadores de inmunohistoquímica como el CD34 que es fuertemente expresado por este tumor, para diferenciarlo de otros tumores de extirpe maligna.
Introduction

Solitary fibrous tumor (SFT) is considered a neoplasm of mesenchymal origin. In 1870, Wagner was the first to describe SFT and it was distinguished from soft tissue tumor in 1931 by Klemperer and Coleman.\(^1\) Ninety percent of solitary fibrous tumors are benign and their differentiation from other tumors is difficult through imaging techniques. Diagnosis is made by means of immunohistochemical staining and the CD34 marker is strongly expressed by this tumor, differentiating it from other malignant tumors.\(^2\) The present case, after transrectal biopsy, was considered a fusocellular tumor with mild atypia and immunohistochemistry was suggested to the pathology service of our hospital in the suspicion of a possible diagnosis of a phyllodes tumor.

Case presentation

A 49-year-old man sought medical attention for gastrointestinal disorders and weight loss, with mild nonspecific pain in the hypogastrium. He did not complain of urinary symptoms. Upon physical examination no abdominal mass was palpated, digital rectal examination showed a normal prostate and a firm mass was palpated in the anterior rectal wall, proximal to the edge of the base of the prostate. Studies included prostate-specific antigen with a level of 0.47 ng/ml, an ultrasound that identified a tumor mass in the pelvic cavity, and a computed axial tomography scan that revealed a heterogeneous solid tumor with a diameter of approximately 9 x 8 cm in the pelvic cavity consistent with a neoplastic process in the right seminal vesicle that involved the prostate gland; no lymphadenopathy was observed (figs. 1 and 2).

In addition to the abovementioned studies, transrectal biopsy of the lesion was performed that reported a mildly atypical fusocellular tumor and immunohistochemical staining at our pathology service was suggested for the possibility of diagnosing a phyllodes tumor. Given these findings, radical cystoprostatectomy with an ileal conduit diversion was performed. The tumor was not firmly adhered to the rectal wall, but it could not be separated from the bladder, and there was no evidence of regional lymphadenopathy.

The macroscopic findings of the histopathologic analysis were: a light brown, firm tumor situated on the right seminal vesicle, measuring 10.5 x 6 x 8 cm at its largest axes; when sliced it was predominantly solid and light brown, but it had cystic zones that contained a transparent...
fluid and solid zones with a nodular configuration with yellowish hues; the serial slices showed the same characteristics (fig. 3).

The microscopic findings were: neoplasia that was well-delimited by a layer of thick connective tissue surrounding it, made up of cells with fusiform nuclei, blunt edges, uniform chromatin, and eosinophilic cytoplasm with imprecise, but defined edges arranged in interlaced segments with a focal pinwheel pattern, numerous hyalinized vessels in a hemangiopericytoma-like pattern alternating with hypercellular epithelioid zones; no tumor necrosis was observed and the mitotic count was 2 to 3 per 20 high power fields.

The immunohistochemical reactions had the following results:
CD34 and Bcl-2 were positive, whereas actin, CD99, CD117, PS 100, chromogranin, and synaptophysin were negative.

The following diagnosis was made with the abovementioned markers: SFT of the seminal vesicle measuring 10.5 cm at its greatest diameter, with no necrosis and a low mitotic count. At one year from surgery the patient has had no tumor recurrence.

Discussion

SFT is a neoplasia that commonly presents in the pleura but it can manifest at any extrathoracic site. Virtually any point in the body can be affected, including the genitourinary tract. The extrapleural locations include the orbit, paranasal sinuses, nasal cavity, respiratory tract, lung, greater salivary gland, mammary gland, meninges, liver, and urogenital organs.

These tumors can be asymptomatic and cause manifestations due to compressing the neighboring organs, but paraneoplastic syndromes with degenerative osteoarthropathy and hypoglycemia have been described. Around 10-15% of solitary fibrous tumors are malignant. Malignancy criteria include atypical locations, increased mitotic activity (more than 4 mitoses per 10 HPF), necrosis, hemorrhage, infiltrative margins, and p53 expression. However, a histologic prediction of aggressive tumor behavior is difficult. Clinical behavior based on histopathology cannot be adequately suspected; tumors of benign appearance can exhibit aggressive behavior and vice versa. Tumor profile is characteristic: diffuse positivity for CD34, now considered the solitary fibrous tumor marker; 70% of these tumors express CD99 and Bcl-2 and only 20-35% are variably positive for EMA and smooth muscle actin. In general, solitary fibrous tumors are negative for cytokeratin, smooth muscle actin, S-100, and c-Kit.

These tumors can be confused with others, such as sarcomas. Our case was initially diagnosed as a mildly atypical fusocellular tumor with the possibility of being a phyllodes tumor, which was why the radical procedure was performed that identified a large pelvic tumor. It has been reported that even when organ-preservation is possible, the risk for recurrence and positive margins should be contemplated and extensive pelvic surgery performed.

Thus it is important to carry out immunohistochemical tests to define the accurate diagnosis and later patient follow-up.

Conclusions

SFT is rare and its location in the urogenital tract is exceptional and difficult to diagnose preoperatively. In our case, the definitive diagnosis was made from the surgical specimen through confirming immunohistochemistry tests. Tumor locations in the urogenital tract include the seminal vesicle, prostate, kidney, bladder, and spermatic cord.

Ethical responsibilities

Protection of persons and animals. The authors declare that no experiments were performed on humans or animals for this study.

Data confidentiality. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Financial disclosure

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Conflict of interest

The authors declare that there is no conflict of interest.

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