Scrotal mesothelioma

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A 67-year-old male patient presented with right scrotal swelling and underwent a right hydrocelectomy. A 1 cm paratesticular lesion was found within the hydrocele sac after entering the tunica vaginalis. Local excision grossly removed this in its entirety. Pathology returned as well differentiated papillary mesothelioma of the tunica vaginalis. Pathologic features and management options are discussed.

Key Words: paratesticular tumor, mesothelioma, scrotal mass

Introduction

A 67-year-old male presented to the emergency department for evaluation of 3 weeks of right-sided scrotal discomfort.

Case report

The patient's pain was associated with scrotal swelling and had developed after horseback riding. He denied a prior history of similar problems, urinary symptoms, fevers, or changes in bowel habits. His past medical history included well-controlled diabetes, coronary artery disease status post remote myocardial infarction, and a 15 pack-year history of smoking, which he quit 20 years ago. His family history is non-contributory. He is retired from managing a motel in Costa Rica. There is no documented asbestos exposure. Physical examination demonstrated a 6 cm x 10 cm right scrotal swelling consistent with a hydrocele. The right testis could not be palpated. His left testis measured 15 mL, the epididymis was normal. There was no evidence of varicocele. The patient was circumcised. Laboratory data were within normal limits. Scrotal ultrasound found normal appearing testicular architecture bilaterally. A right hydrocele was present, in confirmation of his physical exam. In addition, a discrete solid appearing mass was seen arising from the inner wall of the scrotum. This mass contained microcalcifications and blood flow on Doppler evaluation.

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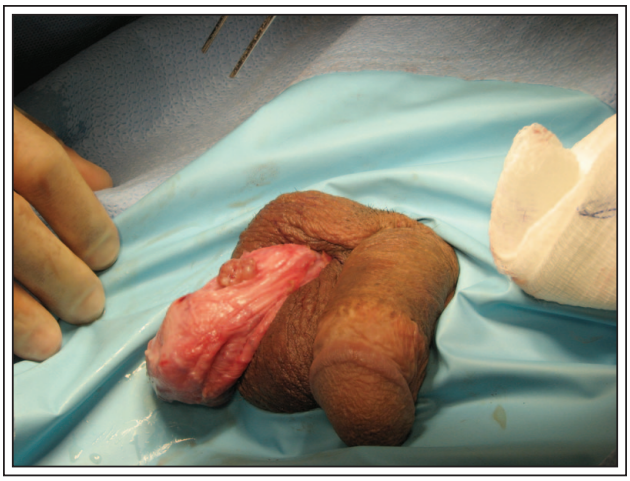

Figure 1. Gross image of paratesticular lesion along the parietal surface of the tunica vaginalis.

The patient underwent a right hydrocelectomy. At the time of surgery, the surgical team encou ntered a 1 cm paratesticular lesion within the hydrocele sac after entering the tunica vaginalis. The lesion was located on the parietal surface of the tunica vaginalis and did not appear to be invasive, Figure 1. Local excision grossly removed this in its entirety. Both the lesion and the hydrocele sac were sent for pathologic analysis.

Pathological examination of the two submitted specimens revealed: 1) a paratesticular lesion consisting of a tan, firm fibrous tissue fragment measuring 1.0 cm x 0.8 cm x 0.7 cm; 2) two fragments of membranous hydrocele sac measuring 4.0 cm x 1.5 cm x 0.2 cm. Histologically the paratesticular lesion consisted of a papillary and tubular proliferation with a focally sclerotic stroma that in some areas entrapped segments of the tubular proliferation. The papillae were lined by a single layer of cuboidal cells lacking cytologic atypia. Mitotic figures were inconspicuous and necrosis was absent. Immunohistochemical stains demonstrated that the cuboidal cells were strongly positive for calretinin and WT-1 supporting a mesothelial origin of this tumor, Figure 2. Sections of the hydrocele sac exhibited a fibrotic cyst wall with inflammation but without evidence of invasion or infiltration by the tumor. A diagnosis of well-differentiated papillary mesothelioma of the tunica vaginalis was made. As such, the patient ultimately underwent right inguinal orchiectomy.

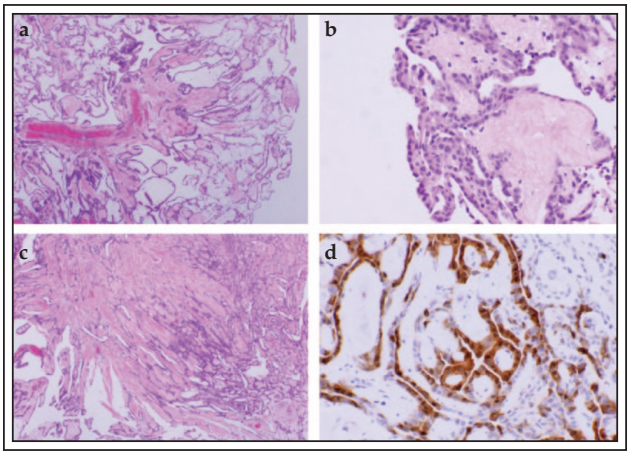

Figure 2. (a) 10x, low power magnification demonstrating tubulo-papillary architecture of paratesticular mass; **(b)** 40X, high power magnification of the single layer of cuboidal epithelial lining cells with bland cytology; **(c)** 10x low power magnification demonstrating papillary fronds with focal sclerosis; **(d)** 40x high power magnification of immunohistochemical stain for calretinin demonstrating positivity in the mesothelial lining cells.

Discussion

The etiology of well-differentiated papillary mesothelioma (WDPM) of the tunica vaginalis remains unknown. It has been suggested that some papillary proliferative lesions may be related to local trauma or inflammation.[1] WDPM exists within the spectrum of proliferative changes of the mesothelium that encompass reactive hyperplasia at one end to malignant mesothelioma at the other. Reactive hyperplasia displays small proliferations of mesothelial cells without forming the papillary fronds with fibrovascular cores or the tubular structures seen in WDPM. It is often a focal process and may be associated with a history of trauma. Importantly formation of a gross mass rules out reactive mesothelial hyperplasia.[2] WDPM is differentiated from malignant mesotheliomas of the tunica vaginalis by observing a well-defined, sharply demarcated zone of proliferation near the luminal surface with more fibrotic areas beneath, but with no evidence of true invasion.[3] It is important to note that the stroma of WDPM may be sclerotic with entrapment of mesothelial cells, producing a pseudoinfiltrative pattern.[4] Distinction from true invasion is essential in making the proper diagnosis. Microscopic features of WDPM exhibit well defined stout papillary structures with myxoid cores, lined by a bland, sometimes flattened, single layer of cuboidal or columnar mesothelial cells. Subnuclear vacuolization may be noted but mitotic figures are rare.[4] In contrast, features favoring malignancy include large size, cellular pleomorphism, significant mitotic activity and necrosis.[1] The defining feature

of malignant mesothelioma of the tunica vaginalis is true invasion into underlying stroma. There are recent case descriptions of "borderline lesions" or lesions of uncertain malignant potential that do not fit into the well-differentiated or diffuse malignant mesothelioma categories. It has been proposed that the designation WDPM be restricted only to WDPM of tunica vaginalis lacking any complex or adverse pathology, while cases with more complex morphology that do not show overt signs of malignancy should be designated "mesothelioma of uncertain malignant potential".[5] Lesions falling into this category exhibit a more complex architecture than that of WDPM, with cribriform, syncytial and solid areas.[2] Invasion is not present in borderline lesions, excluding a diagnosis of malignant mesothelioma. Due to the paucity of reports this concept continues to evolve and is best considered on a case-by-case basis.

Mesothelioma of the tunica vaginalis is a rare entity, occurring in less than 1% of all mesothelioma cases and may be an aggressive, malignant lesion or WDPM, a benign entity. The benign variant is typically described in the peritoneum of young women.[5,6] A recent report notes 223 cases described in the literature, generally in adult and elderly patients. Usually this presents as a firm mass and recurrent hydrocele. It is most commonly caused by asbestos exposure, although only 30% to 40% of these cases document this association.[7] Mesothelial tumors form along the serosa of the pleura, pericardium, and peritoneum. The tunica vaginalis develops from a fold of peritoneum, thus explaining how these paratesticular neoplasms may arise.[8]

Radical orchiectomy is considered first line treatment for malignant paratesticular mesothelioma. In malignant cases, patients undergoing local resection of the hydrocele were more likely to recur and progress than those undergoing radical orchiectomy. Radiation therapy may be of benefit. Regardless of treatment, malignant disease is aggressive with a mean overall survival of 23 months.[8] In contrast, clear guidelines on treatment of WDPM are not available. Most case reports describe patients undergoing radical orchiectomy or simple hydrocelectomy. One report found no recurrent disease or progression after surgical excision (both radical orchiectomy and hydrocelectomy) in 10 patients, although follow up was short with a median of 21 months.[6] WDPM is believed to be benign in nature; however, there has been evidence of rare progression to malignant mesothelioma. As such, some advocate for radical orchiectomy, and may discourage scrotal violation.[8] □

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